

All-Ireland Lymphoedema Guidelines 2022

for the Diagnosis, Assessment and
Management of Lymphoedema



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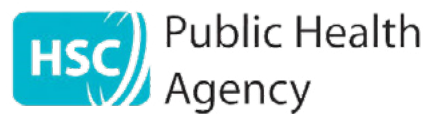
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Note that there are variations across the UK in terminology and training requirements.

All-Ireland Lymphoedema Guidelines for the Diagnosis, Assessment and Management of Lymphoedema 2022

This document is a Clinical Guideline

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Foreword

Dr David Hanlon, HSE National Clinical Advisor for Primary Care

It is with great pleasure that I introduce the All Ireland National Guidelines for the diagnosis, assessment and management of lymphoedema (2022) developed in partnership with our colleagues in Northern Ireland. This guideline updates the 2008 CREST guidelines and provides a standardised, evidence based approach and expert opinion for lymphoedema management. It aims to improve patient outcomes by supporting the clinical decision making process in the management of lymphoedema.

Lymphoedema is a progressive, chronic disease that effects an estimated 45,000 people in Ireland and Northern Ireland combined. It has major personal, social, and economic impact. Healthcare is an ever changing science where advances and new developments in lymphoedema continue to take place. Evidence based knowledge and skills related to advancing lymphoedema management are of crucial importance in delivering care safely, effectively and efficiently. This revision of the 2008 CREST guidelines for lymphoedema ensures that the most up-to-date evidence is available to support the standardisation of care and encourage best clinical practice and patient outcomes.

The guideline is applicable for both hospitals and healthcare organisations, to ensure that patients with lymphoedema wherever they are benefit from high standards of care and quality of interventions. Health Care Professionals with clinical competence in lymphoedema management play a vital role by promoting quality and continuity of care that enables patients/clients to receive treatment effectively and efficiently in the healthcare setting most appropriate to their needs while enabling self-management and individual agency. The availability of these guidelines will support the implementation of good lymphoedema care in all settings and improve the experience and safety of people in our care.

On behalf of the HSE, I wish to acknowledge and express gratitude for the effort and commitment of all those involved in revising the guideline. The service users who have been integral to this process have been invaluable in providing their expertise and experience for which we would like to express special thanks. Our thanks are extended to the project team members in both jurisdictions for their time, commitment and expertise in updating this pivotal guideline.

Go raibh maith agaibh go léir,

Dr David Hanlon MICGP
HSE National Clinical Advisor for Primary Care



Ms Michelle Tennyson, Deputy Director, Public Health Agency, Northern Ireland

The All Ireland Guideline for the Diagnosis, Assessment and Management of Lymphoedema has been developed to replace the 2008 Clinical Resource Efficiency Support Team (CREST) Lymphoedema guideline. The updated evidence review and recognition of changes in pathways, healthcare practices and technology are welcomed to support the increasing number of people living with lymphoedema on the Island of Ireland. In particular, the move to shared care must be celebrated with its increased recognition of the chronic conditions model, and need to improve service user empowerment to enable lifelong self-management and both physical and psychological coping. Similarly, the introduction of cancer prehabilitation, and the lymphoedema screening and surveillance model, will help with early identification of sub-clinical lymphoedema for known at risk cancer-related sub groups, thereby reducing the risk of developing this long term condition for many. Additionally, new service challenges, such as, the increasing number

of children with lymphoedema, those living with palliative lymphoedema needs, and the impact of global bariatric issues have been addressed in independent sections, and will support wider care. The new evidence base provides a thorough platform to ensure care pathways continue to be modernised and provide effective and efficient care.

This integrated work programme with the Health Service Executive exemplifies strategic cross border working, and provides an evidence base and recommendations for practice across Ireland. The Public Health Agency and Department of Health (Northern Ireland) would like to thank the project leads and all members of the Guideline Development Group, Working Groups and External Review Group for their contribution to this work, despite the challenges associated with Covid 19. In particular, I would like to extend special thanks to the service user representatives who have been integral to this process, and invaluable in providing their experience and expertise.

Ms Michelle Tennyson
Chair, Lymphoedema Network Northern Ireland (LNNI)
Deputy Director (Lead AHP, PPI & PCE)
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Executive Summary

The All-Ireland Lymphoedema Guideline was developed as a clinical resource to promote evidence-based practice and as a guide for managers and commissioners to support modernisation of pathways of lymphoedema care, education and research. The objective is to ensure that all citizens will have access to local, evidence based lymphoedema resources.

There has been a plethora of lymphatic associated research in the past decade, which necessitated the replacement of the CREST Diagnosis, Assessment and Management of Lymphoedema Guideline (Clinical Resource Efficiency Support Team, 2008). The partnership approach by the Health Service Executive (HSE), Republic of Ireland, and Health and Social Care (HSC) Northern Ireland, facilitated a cross-border initiative to address this resource gap. The Guideline Development Group membership ensured multi-professional, service user, cross sector and pan-Ireland representation and engagement. The creation of Working Sub Groups again widened participation. Further engagement from two external reviews ensured that colleagues throughout the United Kingdom and Ireland were included in agreeing the content and structure design.

The terms Lymphoedema and Chronic Oedema can be used interchangeably, and for the purposes of this resource, the term lymphoedema is used. This document aims to ensure that all involved in health delivery recognise the extending role of lymphoedema management. Lymphoedema can affect people at all stages of life: from birth where people are born with damaged or absent lymphatics due to genetics, through to vascular associated lymphoedema in later life and palliative lymphoedema management.

General population health changes have also altered recent lymphoedema referral patterns with increasing numbers of frail elderly citizens, and a substantial increase in those living with obesity; lymphoedema incidence increases with age and with weight. There is also an overall increase in numbers of co-morbidities, such as diabetes and arthritis-related reduced mobility, which affect management and life-long care. Recurrent cellulitis can be a both a cause and a sign of un-managed lymphoedema. Recognising antimicrobial stewardship as best clinical practice, timely referrals and successful lymphoedema management will reduce the incidence of lymphoedema-associated cellulitis thus improving health outcomes and reducing antibiotic usage with related savings.

The impact of these observations has changed the complexity of lymphoedema care provision, with a greater need for social prescribing, third sector liaison and widening of funded professional

support to lymphoedema services. Complexity requires an holistic approach to achieve reduction in symptoms and improve quality of life; modern lymphoedema services therefore require funded access to psychology and dietetics and, bariatric services. This document provides the evidence, recommendations and pathways to support a more effective and prudent use of resources as part of wider multi-professional teams.

The guideline will be available on the [HSE](#) and [LNNI](#) websites, and will be shared with national and international stakeholders.

Disclaimer

The All-Ireland Lymphoedema Diagnosis, Assessment and Management Guideline (2022) should be used in conjunction with clinical judgement, training and scope of practice. Recommendations may not be appropriate in all circumstances, and decisions to adopt specific recommendations should be made by the clinician taking into account the circumstances presented by individual patients, available resources and specific settings.

Section A. Guideline Recommendations

Section A includes the clinical topics which are broken into sub-sections. The sub-sections are cross referenced/linked to improve readability, however the general section should be referenced alongside each of the other sub-sections as it has many recommendations relevant to more than one other sub-section.

The colour-coded sub sections are:

1. **General Section:**
 - Introduction
 - Risk reduction and awareness
 - Diagnosis and assessment
 - Management
2. Chronic Oedema
3. Primary Lymphoedema
4. Surgery and lymphoedema
5. Oncology related lymphoedema
6. Lymphoedema in children and young people
7. Lymphoedema in people living with obesity
8. Lymphoedema in palliative care patients
9. Lymphoedema education

Section B. Guideline Development

Section B includes details relating to the process of developing this guideline. Areas covered include initiation, development, governance and approval, communication and dissemination, implementation, monitoring, audit and evaluation and revision/update, as per guidance by the HSE National Framework for Developing Policies Procedures Protocols and Guidance (2016).

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PART A: Guideline Recommendations

Introduction

Lymphoedema can be defined as “The progressive swelling of a body part, usually an extremity, following developmental (**Primary Lymphoedema**) or acquired (**Secondary Lymphoedema**) disruption of the lymphatic system resulting in lymph (a protein-rich fluid) accumulating in the interstitial space. The extremities are most commonly involved, followed by the genitalia. In general, lower limb lymphoedema is significantly more common than upper limb lymphoedema (Tiwari et al., 2003).

‘Chronic Oedema’ is often used interchangeably with the term ‘Lymphoedema’. Oedema results from an imbalance between capillary filtration into and lymphatic drainage from the interstitial space. In every case of chronic oedema there will be some impairment of lymphatic drainage, either through an underlying abnormality (‘primary’) or as an acquired lymphatic failure (‘secondary’) with a result of the capacity of the lymphatics being overloaded. Where there is an impairment of lymphatic drainage, over time, the fluid component of oedema may become replaced by fibrosis and/or adipose tissue (National Lymphoedema Partnership, 2015).

Of note, even with acute causes, there may be an impact on the lymphatic system which then predisposes to a more chronic condition. In other situations, the clinical picture may indicate that long term management will be needed even within the first 3 months after presentation. According to the National Lymphoedema Partnership (NLP), the term ‘chronic’ should therefore not be seen as restrictive (2015).

Lymphoedema has been characterised by Professor Peter Mortimer as a failure of the lymphatic system. This can manifest as swelling, skin and tissue changes and predisposition to infection (Mortimer and Gordon, 2016). As per the National Lymphoedema Partnership (NLP), the term ‘Chronic oedema’ refers to a condition caused by various factors such as venous disease, trauma, infection and surgery (National Lymphoedema Partnership, 2015).

Therefore, for the purpose of this document the term lymphoedema will be used as an umbrella term and will include all chronic oedema as they have been shown to be the same entity (Moffatt et al., 2019). In table 1, the lymphoedema column indicates a classification describing dysfunction to lymphatic drainage, either genetically or by secondary causes. Ideally these conditions which lead or contribute to poor lymph drainage should be managed, which may in turn reduce the oedema. The chronic oedema column shows examples of the many conditions which may progress to lymphatic overload and failure of drainage, thus resulting in oedema formation. The treatment options for the management of simple chronic oedema and lymphoedema are therefore similar. It is expected that simple chronic oedema can be managed by non-specialists as part of a wider pathway, with timely access to specialist lymphoedema services for more complex presentations.

Table 1. Diagnosis of chronic oedema and lymphoedema

Chronic oedema	Lymphoedema
• Heart Failure	• Primary: genetic • Secondary: trauma, cancer-related, obesity, venous disease, infection
• Venous disease	
• Renal disease	
• Obesity	

Secondary causes of lymphoedema

The aetiology of lymphoedema can be related to many secondary elements, see classification below for common causes of secondary lymphoedema (Table 2).

Table 2. Classification of causes of Secondary Lymphoedema

Classification	Example
Trauma and tissue damage	<ul style="list-style-type: none"> • Lymph node excision • Radiotherapy • Burns • Varicose vein surgery/harvesting • Large/circumferential wounds • Scarring • Self-harm
Malignancy	<ul style="list-style-type: none"> • Lymph node metastases • Infiltrative carcinoma • Lymphoma • Pressure from large tumours
Venous disease	<ul style="list-style-type: none"> • Chronic venous insufficiency • Venous ulceration • Post-thrombotic syndrome (DVT) • Intravenous drug use
Infection	<ul style="list-style-type: none"> • Cellulitis/erysipelas • Lymphadenitis • Filariasis • Tuberculosis
Inflammation	<ul style="list-style-type: none"> • Rheumatoid arthritis • Psoriatic arthritis • Dermatitis/eczema • Sarcoidosis and orofacial granulomatosis • Podoconiosis • Pretibial Myxoedema
Immobility and Dependency	<ul style="list-style-type: none"> • Dependency oedema • Paralysis • Sleep Apnoea
Obesity	<ul style="list-style-type: none"> • Increased BMI >30Kg/m²

(Adapted from Lymphoedema Framework, Best Practice for the management of lymphoedema, International Consensus. London MEP Ltd, 2006)

Structure and Function of the Lymphatic System

The lymphatic system is a network of vessels, fluid (lymph), cells and organs that has many functions including maintaining body fluid homeostasis, transporting and facilitating cells of the immune system, and facilitating the absorption of dietary lipids (Ozdowski and Gupta, 2021). The formation of lymph occurs when interstitial fluid (that has leaked/extravasated from the vascular system into the interstitial space) enters the lymphatic capillaries (Scallan and Huxley, 2011). The lymph is transported through the lymphatic system via both intrinsic mechanisms (contraction of lymphatic vessels and intraluminal valves within vessels) and external mechanisms (surrounding tissues - contraction of skeletal muscle, breathing, bowel movements etc.) (Breslin, 2014). The lymph drains to the lymphatic vessels (capillaries and then to collecting vessels), through lymph nodes, to lymphatic trunks, and finally to ducts where the lymph then re-enters the vascular system via the subclavian veins (Ozdowski and Gupta, 2021). Lymph functions to clear pathogens by preventing their entry in to the bloodstream and transporting them to the lymph nodes where they are engulfed by phagocytic immune cells (Liao and von der Weid, 2015).

The term “lymphoedema” has historically been defined as either primary or secondary lymphoedema. Primary lymphoedema occurs as a result of lymphatic development failure and secondary lymphoedema is caused lymphatic injury most commonly due to trauma, cancer treatment or parasitic infection (filariasis) (Sleigh and Manna, 2019).

Primary Lymphoedema

Primary lymphoedema is an inherited or congenital condition that causes a malformation of the lymphatic system, most often because of genetic mutation, with swelling resulting from failure of the lymphatic system. Primary lymphoedema is uncommon and consists of malformation of the lymphatic vessels and valves (Brouillard et al., 2014) and most often affects the lower limbs but can also occur in the upper limbs and genitalia (Ross et al., 1998). Congenital lymphoedema is the term used for primary lymphoedema in the new-born (Kitsiou-Tzeli et al., 2010). The more recent St George’s Algorithm (Gordon et al., 2021) classifies congenital as occurring within the first year of life.

Secondary Lymphoedema

Secondary lymphoedema is acquired and is relatively common compared to primary lymphoedema with estimated prevalence of 1 in 1,000 versus 1 in 100,000 for primary lymphoedema (Butler et al., 2009). The mechanism by which secondary lymphoedema develops is obstruction or damage to the lymphatic vessels (Grada and Phillips, 2017, Cueni and Detmar, 2008) due to various different aetiologies including trauma, malignancy and its treatment, surgery, radiotherapy and infection (Grada and Phillips, 2017), chronic venous insufficiency, lipoedema, immobility and chronic underlying systemic diseases (Borman, 2018).

Filariasis

Filariasis is a parasitic infection and is the most common cause of secondary lymphoedema worldwide (Grada and Phillips, 2017). The vast majority of infections are due to the mosquito-borne roundworm *Wuchereria bancrofti* (Melrose, 2002). This tropical disease is endemic in parts of sub-Saharan Africa and India where a bite by an affected mosquito can result in migration of larvae to the lymphatics where they mature into adult nematodes and cause subsequent lymphatic dysfunction by damaging lymphatic valves, resulting in lymph stasis and dilatation of vessels (Grada and Phillips, 2017).

Filarial nematodes are transmitted by insects which consume human blood. These nematodes suppress the immune system of infected individuals leading to chronic infection. Inflammation is

triggered by the death of the parasite, resulting in hydrocoele, lymphoedema, and elephantiasis. Filarial infection may be diagnosed by detection of microfilariae in blood taken at night (owing to the nocturnal nature of *W. bancrofti*). Treatment of lymphoedema secondary filariasis typically consists of a combination of physiotherapy and pharmacotherapy. Drugs to treat filariasis include diethylcarbamazine, ivermectin, and albendazole, which are used mostly in combination to reduce microfilariae in blood (Taylor et al., 2010).

Patients at risk of lymphoedema

In some recognised situations, (e.g. after some cancer treatments, post deep vein thrombosis, obesity, severe immobility) an impairment of the lymphatic system may be present even before the outward signs of oedema are observable or measurable. In these situations, the affected person is at risk of developing oedema and risk reduction strategies may prevent or delay the onset of symptoms or signs.

Other causes of secondary lymphoedema

Cellulitis is a bacterial skin infection involving the skin and subcutaneous tissue that can also cause secondary lymphoedema by inflicting damage to the lymphatic system (Al-Niami, 2009). The relationship between lymphoedema and cellulitis is reciprocal in that lymphoedema is also a significant risk factor for cellulitis (Al-Niami, 2009). The impeded flow of lymph provides an ideal medium for bacterial proliferation, while reduced lymphatic drainage enable the pathogens to evade the local immune response (Al-Niami, 2009).

Inflammatory conditions

Lymphoedema has been documented as a rare consequence of rheumatological diseases, with rheumatoid arthritis being the most commonly associated disorder, followed by psoriatic arthritis, ankylosing spondylitis, systemic sclerosis and juvenile rheumatic disease (Eyigör, 2013). The pathophysiology of the lymphoedema is uncertain but presents as extra-articular disease and may be attributed to inflammatory processes and subsequent injury to the lymphatics (Eyigör, 2013). Though not strictly ‘oedema’ (i.e. an accumulation of interstitial fluid), other conditions (e.g. lipoedema) that cause swelling of the limbs, can, in themselves lead to longer term lymphatic changes and are therefore included in the group of lymphoedema from the perspective of assessment and management.

Table 3. Data from the 2018 and 2020 LNNI referral audit/HSE 2021 audit

Type of lymphoedema	LNNI 2018 % referral type	LNNI 2020 % referral Type	HSE 2021 % referral type
Non-cancer-related secondary lymphoedema	52 %	53.51 %	48%
Cancer related secondary lymphoedema	40 %	39.4 %	45%
Primary lymphoedema	6 %	7.1 %	7%

Lipoedema

The remit of this guideline is lymphoedema (risk reduction, diagnosis and management) and does not cover lipoedema. The UK Lipoedema guideline was published in 2017, and the British Lymphology Society has produced a 2021 lipoedema Fact Sheet to support the 2020 European Lipoedema Consensus document. These lipoedema documents, alongside charity (Lipoedema UK) publications, can be found on the website link <https://lnni.org/content/lipoedema>.

Epidemiology

The accuracy of the prevalence of lymphoedema is evolving. In 2003, (Moffatt et al.) completed a study in the UK which suggested that 1.33 per 1,000 people of all ages are affected with lymphoedema, increasing to 5.4 per 1,000 after the age of 65. It is well documented that the incidence of lymphoedema increases with age due to immobility, poor circulation and other comorbidities. This figure was used to calculate anticipated regional caseload in the 2004 Department of Health, Social Services and Public Safety (DHSSPS) 'Lymphoedema Services: Report of the Lymphoedema Services Review Group' and also the 2008 Clinical Resource, Efficiency and Support Team (CREST) 'Guidelines for the assessment and management of lymphoedema'.

This has been proven to be an underestimation by several studies (Moffat, 2012) showing a rate of 3.99 per 1,000 across all ages. In 2017 a prevalence of 3.93 per 1,000 was recorded (Moffatt et al., 2017). This study also identified that nearly one third of all in-patients had chronic oedema which dispels the myth that chronic oedema is confined to those seen by community-based health services. This in-patient result was also noted in a similar prevalence study in Denmark which also noted the complexity of this in-patient group regarding co-morbidities (Nørregaard et al., 2019). Another 2019 international paper (Quérel et al., 2019) also reported the same one third prevalence of an in-patient caseload; risk factors also reflect those of the other studies: age, obesity, heart failure, immobility and neurological deficits.

A 2019 UK community nursing services study reported that 56.7% of patients in the district nursing caseload had chronic oedema (Moffatt et al., 2019c). Compared to those without chronic oedema, patients with chronic oedema had a higher incidence of diabetes, heart failure and peripheral arterial occlusive disease. In a 2019 study, cellulitis affected 24.7%, 71.6% had reduced mobility and 61.9% were people living with obesity highlighting the significance of co-morbidities (Moffatt et al., 2019b).

Regarding the location of potential patients and prevalence, a 2019 Australian study (Gordon et al., 2019) covered 4 sites, all of which presented varied prevalence figures for chronic oedema:

- Site 1 residential care facilities: 54% of residents had swelling
- Site 2 community-delivered aged care services – 24% of patients had swelling
- Site 3 hospital – 28% of patients had swelling
- Site 4 wound treatment centre – 100% of patients had swelling

Lymphoedema Network Northern Ireland (LNNI) referral data collected from 2010 to 2018 suggests an average of 1,348 referrals per year. The total annual referral figure has however increased year on year; a more relevant figure for current activity would therefore be from 2019 which had 2050 referrals. This could reflect an incidence figure, but the changing picture of chronic oedema populations makes this challenging to confirm, and would still be considered an underestimate. These figures suggest a prevalence of 6.78 per 1,000 (Table 4). Wales has had a lymphoedema service for longer than Northern Ireland (N.I.); their 2017 data suggested a prevalence of 5.49 per 1,000 which has been increased in 2019 to 6.4 per 1,000 (Underwood et al., 2019).

LNNI has identified an increasing number of referrals for those without a primary lymphoedema diagnosis or direct secondary trauma. This new population is associated with fluid dependency and obesity, and reflects other factors which apply pressure to the natural system of lymphatic drainage. Paediatric lymphoedema is also an evolving referral group. In Northern Ireland in 2008 there were no paediatric cases recorded regionally. The network has engaged with neo-natal, paediatric, Health Visitor and School Nurse services, and has noted a gradual increase in numbers

with a 2021 primary caseload of thirty-five children (aged 0 – 18) (plus forty-eight children with a vascular anomaly). The Welsh Lymphoedema Network has a dedicated paediatric oedema post and 2019 extrapolated data from this work has suggested that Northern Ireland should have approx. 150 children/young adults with lymphoedema and 405 in the republic (includes vascular anomalies, cancer-related and secondary non-cancer cases).

Oedema in palliative care is a significant problem. In non-cancer patients an oedema prevalence of 85% is reported near the end of life which may occur months prior to death, and may be amenable to management during that time. The International Lymphoedema Framework (Framework, 2010) states that palliative oedema is thought to be approximately 5-10% of all lymphoedema referrals, but this is considered an underestimate. In Northern Ireland, a 2016 regional audit found that palliative lymphoedema referrals were 4.42 - 6.5% of the dedicated lymphoedema team referrals, and additionally 19.96 - 26.93% of the specialist palliative care physiotherapy referrals (Public Health Agency, 2018). A study in the Republic of Ireland (Real et al., 2016) found an incidence of 10.5% of lymphoedema/oedema at end of life. Taking an average of 10%, this would equate to approximately 550 patients annually in Northern Ireland and 825 patients annually in the Republic of Ireland, based on the current populations.

Table 4. Estimated overall prevalence of primary and secondary lymphoedema using research and current regional data (Northern Ireland)

Population in N.I. (mid-year 2018 regional population data (NISRA))	Northern Ireland Prevalence rates			
	1.3 in 1,000 5.4 in 1,000 (> 65) (Moffatt, 2003)	3.99 in 1,000 (Moffatt, 2012)	3.93 in 1000 (Moffatt, 2017)	6.78 in 1000 (N.I., 2018)
Total - 1,881,600	2,446	7,507	7,394	12,550
Age Related				
< 65- 1,573,400	2,045			
> 65- 308,200	1664			
Total	3,709	7,507	7,394	12, 550

It is important to note that this accounts only for the population growth; given the higher incidence of Lymphoedema/Chronic Oedema in older patients, the ageing population will also have a big impact on increasing prevalence. The 2018 LNNI audit of referral cause reported 52.2% of the recorded caseload was related to secondary non-cancer-related lymphoedema /chronic oedema; many of these referrals are chronic oedema with causes linked to venous changes, dependency and obesity.

Table 5. Estimated overall prevalence of primary and secondary lymphoedema using research and current regional data (Republic of Ireland)

Population in Ireland 2018	Ireland Prevalence rates			
	1.3 in 1,000 5.4 in 1,000 (> 65) (Moffatt, 2003)	3.99 in 1,000 (Moffatt, 2012)	3.93 in 1000 (Moffatt, 2017)	6.78 in 1000 (N.I., 2018)
Total - 4,761,865	6,190	12,380	19,000	32,285
Age Related				
< 65- 1,573,400	5,014			
> 65- 308,200	3,443			
Total	8,457	12,380	19,000	32,285

Prevalence of Primary Lymphoedema

Prevalence rates of primary lymphoedema vary widely however it is considered rare, with estimates ranging from 1 in 6,000 (Lymphoedema – NHS www.nhs.uk) up to 1 in 100,000 (Sleigh and Manna, 2019). Several sources (HSE, 2018) have used international data to estimate primary lymphoedema, and estimate that 10% of all lymphoedema patients have primary lymphoedema; this may be congenital and present at birth, or may develop later in life. Using this percentage, this would give an estimated 1,255 patients in N.I. with primary lymphoedema and 3229 in the ROI.

The impact of lymphoedema

Lymphoedema has a significant impact on the individual, society and on the wider health and social care systems. A dedicated funding stream for lymphoedema/lipoedema services would ensure sustainability. A national health economic assessment of the value of prevention in lymphoedema would be eagerly welcomed in the scientific literature. Early detection and prevention play an important part in reducing the disease burden in this condition for which prevalence is expected to increase due to the growing and aging population, increased cancer incidence, cancer survivorship and obesity.

Personal impact

Lymphoedema/chronic oedema can have a devastating impact on the individual (Moffatt et al., 2003) A qualitative study (n = 228) carried out in South West London found that as a direct result of lymphoedema:

- 80% of patients had to take time off work with 8% having to stop work completely.
- 50% of patients had recurrent episodes of cellulitis with 27% requiring hospital admission for IV antibiotics with a mean stay of 12 days.
- 50% of patients reported uncontrolled pain
- 33% of patients had not been told they had lymphoedema
- 36% had received no treatment.

This poor level of diagnosis and care is further supported by the Lymphoedema Support Network members' survey 2015 (Lymphoedema Support Network, 2015):

'I feel as though one has to battle all the way along the line – the discomfort, the fatigue, the disfigurement and no-one to help – it leaves one feeling alone and helpless'.

'I stayed positive for so long during my cancer journey but now I am alone and having to deal with lymphoedema – I just want to be normal, buy clothes that fit, enjoy my family, go on holidays but without help how can I do that – how can I face the future – I am no longer positive'.

This psychosocial impact requires specialist management which is beyond the scope of most HCPs. Each service should ideally have access to funded and ring-fenced psychological resources.

Economic impact

Cellulitis is a complication of lymphoedema but can also be a cause. Recurrent cellulitis can lead to lymphoedema due to a chronic cycle of infection causing prolonged swelling and subsequent damage to the lymphatics (Keeley, 2008). In a study of patients admitted to acute services for cellulitis, lymphoedema was a major factor for 18% (Moffatt, 2016). A Canadian study reported that 72.06% of patients attending a wound management clinic had suffered cellulitis (Keast et al, 2019).

In NI the SEHSCT cellulitis data and costing at £400 per bed day equates to £2,126,400 per annum for one trust. The SEHSCT is responsible for 19% of the regional population. The regional cost extrapolated to Northern Ireland figures equates to £11,191,578.95 for acute cellulitis management. Using the Moffatt (2016) findings, 18% of this (£2,014,484.21) relates to lymphoedema-related cellulitis management.

In the HSE (2016) there were over 8,000 admissions for cellulitis recorded in HSE HIPE data. Using the Moffatt data of 18% prevalence rate, 1,451 of these 8,000 patients could have significant lymphoedema and a high risk of re-occurrence. The 2015/2016 HIPE data showed an average LOS of 11 days for major complexity cellulitis which would corroborate the research data of 10-12 days for cellulitis patients with lymphoedema. This would equate to a cost of approximately €13.6m for hospital admissions with cellulitis related to lymphoedema/chronic oedema (HSE Lymphoedema Model of Care).

If a patient has had more than one episode of cellulitis in a limb, there is almost certainly some failure of lymphatic drainage. In 2013-2014, there were 104,598 recorded cases of cellulitis treated in secondary care in the UK, of which 69,229 hospital admissions involved a mean and median bed stay of 6.2 and 3 days, respectively reflecting the current SET mean figures. In England, it is currently estimated that over £178 million is spent on acute hospital admissions for lymphoedema related cellulitis (Health London Partnership 2017; National Lymphoedema Partnership, 2019).

The correct management of chronic oedema/lymphoedema significantly reduces the risk of cellulitis development. The introduction of a new chronic oedema/lymphoedema service to a London borough CCG has shown a decrease of 94% in cellulitis episodes and an 87% reduction in cellulitis admissions to hospital (Healthy London Partnership, 2017). Timely and appropriate management of lymphoedema/ chronic oedema will save both admission costs, and personal cost to the patient.

Data from the Lymphoedema Network Wales, Enfield Community Services and the Accelerate CIC lymphoedema service in London have all shown the significant financial benefits from the investment in specialist lymphoedema care (Healthy London Partnership, 2017). A recent economic analysis from Swansea Centre for Health Economics on the value of Lymphoedema Network Wales demonstrated that implementation of the service resulted in reductions in waste, harm and variation.

Data showed statistically significant reductions in GP surgery and home visits, community nurse

care and hospital admissions due to cellulitis. Savings were also highlighted in dressing and bandaging costs as well as significant improvements in quality of life (Thomas et al., 2017). Hill and Davies reported that, although the Enfield service has seen a considerable rise in referrals over recent years, patients are now referred at earlier stages of their condition, reducing the need for intensive treatment and reducing hospital admissions for cellulitis (NHS Transforming Cancer Services Team for London, 2016). Accelerate CIC reported that in a study of 496 patients treated in the first year following introduction of a new community based service for City & Hackney CCG, 30% had a history of cellulitis in the year prior to treatment. They demonstrated a 94% decrease in cellulitis episodes for the same group following commencement of treatment, with an 87% reduction in cellulitis-related hospital admissions.

The National Lymphoedema Partnership (2019) commissioning guidance for lymphoedema services for adults in the UK states the economic impact for England, which can be replicated regionally. The key findings of the report were as follows:

- Lymphoedema has a significant impact on the individual, society and on the wider health and social care system. England currently spends more than £178 million on admissions due to lymphoedema (National Lymphoedema Partnership, 2019).
- Patients with lymphoedema have a significant risk of developing cellulitis and of resulting hospitalisation. In the Moffatt (2001) study of 228 patients, 65 (29%) had at least one episode of cellulitis and 16 of those required hospital admission with a mean length of stay of 12 days.
- In a study of patients admitted to acute services for cellulitis, lymphoedema was a major factor for 18% of patients (Moffatt, 2016), which equates regionally to £2,014,484.21 for lymphoedema-related cellulitis management (using 2018-19 figures) with an average length of stay of 6.06 bed-days (for cellulitis of limbs only).
- Compression is core to the management of lymphoedema, and £500,000 was spent by regional lymphoedema services in the 2015 LNNI audit (not including vascular/Tissue Viability Nursing or other community spend). The audit also demonstrated that a lack of specialist expertise in processing prescriptions for these garments, led to mistakes, delayed appropriate management and consequential waste.

The 2019 Northern Ireland pharmacy expenditure for all prescription compression (across all professional groups) regionally was £2,168,123 (an increase of £168,950 on 2018).

Economic Impact key points (LNNI Population needs assessment, 2020):

- Lymphoedema impacts physically, psychosocially, and economically.
- The lymphoedema services do not have access to ring-fenced psychological support.
- Cellulitis related to unmanaged oedema is responsible for extensive hospital bed stays and prescription costs; a reduction in cellulitis risk will occur with good oedema management.
- Improved multi-professional differential diagnosis is needed to distinguish between cellulitis and other dermatological conditions
- Investment in lymphoedema specialists to work jointly with general community, nursing home and general practice staff is effective in both clinical outcomes and resource savings.
- Prescribing compression garments needs to be completed by a skilled HCP. Mistakes can be harmful to the patient and wasteful of resources. Efficient and timely prescribing will ensure correct lifelong management.
- There has been an annual increase in prescribed compression spend.

Chronic disease is categorised into four levels which relate condition severity to level of

intervention required. As with all chronic diseases it is essential that the risk of lymphoedema is reduced where possible, and the condition be detected and treated early. The at-risk population, the largest proportion of the management triangle, must be actively targeted through awareness, education and prevention programmes. Lymphoedema can be maintained and controlled effectively, improving quality of life for patients with reduced costs. Appropriate intervention, at the right time reduces the requirement for more intensive and expensive treatment further down the disease pathway. The 2012 LNNI management figures are consistent with the Kaiser Permanente hierarchy of need triangle model (Table 6). The referred population also shows a shift in care provision in relation to the change in referral type from 2012 to 2018.

Table 6. The Kaiser Permanente, LNNI (2012) and Lymphdat (2018) report findings

Kaiser Permanente Triangle of Management/ hierarchy of need Levels	Kaiser Permanente %	2012 LNNI Lymphdat %	2018 LNNI Lymphdat %
At risk population	-	-	-
Level 1 : self-care/ support management	70	64.86 (other)	64.22 (other)
Level 2: Long Term management	20	16.67 (modified CDT)	29.47 (modified CDT)
Level 3: complex management	10	11.75 (CDT)	6.43 (CDT)

Management of Lymphoedema

While lymphoedema is not curable it can be successfully managed with the correct treatment, self-management and support. The assessment, treatment and risk factors for lymphoedema are comprehensively covered in this section.

Lymphoedema Stages

There are many internationally recognised classification of stages of lymphoedema and for this guideline the following classification will be used.

Stage 0 (Latency stage)

The patient is considered “at-risk” for lymphoedema development due to injury to the lymphatic vessels but does not present with outward signs of oedema. Includes patients with cancer who have undergone nodal surgery, sentinel lymph node biopsy and/or radiation but have not yet developed swelling. Lymphatic transport capacity has been reduced, which predisposes the patient to lymphatic overload and resultant oedema.

Stage 1 (Spontaneous)

- Reversible
- Pitting oedema
- Swelling at this stage is soft, and may respond to elevation

Stage 2 (Spontaneously irreversible)

- Tissue fibrosis/induration
- Swelling does not respond to elevation
- Skin and tissue thickening occur as the limb volume increases
- Pitting may be present, but may be difficult to assess due to tissue and or skin fibrosis

Stage 3 (Lymphostatic elephantiasis)

- Pitting oedema
- Fibrosis
- Skin changes: papillomata, cellulitis, xerosis (drying of the skin)

Complications of lymphoedema

Skin problems are common in patients with lymphoedema. Swelling may produce deep skin folds where fungal and bacterial infections (cellulitis) can develop. Chronic inflammation causes deposition of fibrin and collagen, contributing to skin thickening and firm tissue consistency. Reduced tissue compliance may further compromise lymph flow and increase the tendency to infection. Lymphorrhoea is also a common complication. There are well documented psychosocial complications including but not limited to altered body image and reduced independence.

Lymphoedema Investigations

Lymphoedema diagnosis utilises symptoms and clinical signs alongside radiological evidence. At present lymphoscintigraphy is the most common and traditionally utilised modality of imaging. The visualisation of the lymphatics can inform the clinician of the likely cause of the oedema.

Table 7. Lymphoedema Imaging Modalities (Bernas et al., 2018)

Imaging	Method	Details
Lymphoscintigraphy	Nuclear Medicine	The current gold standard which can assess from injection site in hand or foot to the entry in the venous system.
Lymphography	X-ray	Classic method providing anatomical details.
ICG	Fluorescence	Increasing in use, but contrast agents limit use. Primarily superficial imaging requiring multiple injections.
Ultrasound	Sonography	Useful in some populations, e.g. asymptomatic filariasis. Also useful for superficial tissue composition for diagnosis and treatment monitoring.
MRI	Magnetic Resonance	Both contrast and non-contrast methods are used. High spatial resolution MR provides greater detail useful for surgical planning or interventional radiology procedures.
CT	Computed tomography	Rarely used due to ionising radiation, but can be used for tissue analysis and volume rendering.
Others	Experimental	There is a variety of emerging imaging techniques e.g. photo-acoustics, specific tracers targeting the lymphatic system and new contrast agents.

Treatment of Lymphoedema

After systemic causes have been excluded and/or treated, the goals of lymphoedema management should be focused on improving venous and lymph drainage. Compression, elevation, physical activity and skincare should be included as part of the management of lymphoedema. Compression may include bandaging and/or garments. American Venous Forum (Gloviczki, 2016) recommend that at least 6 months of conservative therapy be carried out in patients with lymphoedema before any surgical interventions are attempted.

Core prevention measures:

- Physical activity
- Weight management
- Skin care
- Lymphoedema awareness course

Management can be split into two phases:

1. Intensive phase

In this phase of management, the short-term aims are to:

- Restore maximal functional independence and postural balance
- Provide psychological support
- Reduce risk of infection
- Reduce the volume of swelling until it becomes stable
- Improve limb shape where possible
- Improve skin condition
- Meet subjective patient goals
- Educate patients in understanding their condition and rationale for treatment
- Collaborative work with other services if wounds/Lymphorrhoea are present

Initial treatment phase components may include:

- Lymphoedema education to enable self-management
- Strength training and physical activity
- Skin care
- Weight management
- Compression Garments/Wraps
- Compression bandaging
- MLD/SLD
- Intermittent pneumatic compression (IPC), pressotherapy (in combination with other modalities)
- Mobilising tissue, fascia release technique
- Other modalities e.g. Kinesio taping

2. Maintenance phase

In this phase of management, the long-term aims are to:

- Maintain reduction in swelling
- Maintain improvement in limb shape
- Maintain skin integrity

For both phases, there are four principles of care that should be applied to achieve the management goals. These are:

- Build service user skills and confidence to enable self-management
- Skin and wound care
- Exercise
- Compression therapy

Maintenance phase components may include:

- Lymphoedema education to further develop self-management skills
- Simple/Self Lymphatic Drainage (SLD)
- Self-bandaging (potentially also nocturnal)
- Compression garments (potentially also nocturnal)
- IPC / pressotherapy
- Exercise
- Weight management
- Monitoring and self-application skin care
- Other modalities suggested by a HCP
- Referral to local and/or external resources for lifestyle/behavioural support

Compression Therapy

Introduction to compression therapy

Compression garments are indicated in the treatment of lymphoedema, vascular insufficiency and associated disease, lipoedema and wound healing. Many terms are in use such as: graduated compression, hosiery, graduated support hosiery, stockings. The term compression garments is used in this guideline to include all types of compression garments used for upper/lower limb, head and trunk. Thromboembolic disease stockings (TEDS) are not included in this guideline as they are for short term use (3 weeks as per manufacturer guideline) to prevent DVT in immobile patients in the acute setting.

Compression garments are used to promote circulatory and lymphatic function, reduce pain associated with fluid build-up and prevent accumulation of oedema. Compression garments are available for varying compression levels. These range from low to very high compression and are divided into classes I-4. Unfortunately, the standard defining the level of compression and class varies from country to country and leads to confusion depending on where the garment is made (Table 8).

Table 8: Current standards for compression garments*

	British (mmHg ^{**})	French (mmHg)	German ^{***} (mmHg)
	BS 6612:1985	ASQUAL	RAL-GZ 387:2000
Testing method	HATRA	IFTH	HOSY
Class 1	14-17	10-15	18-21
Class 2	18-24	15-20	23-32
Class 3	25-35	20-36	34-46
Class 4	Not reported	> 36	> 49

* The standards are based on lower limb pressures not upper

**The mmHg ranges refer to the pressures applied at ankle circumference (at smallest girth) by the compression garments.

*** The German standard is the only to include custom made garments

To avoid confusion, a new document ‘List of classes and pressures for Lower Limb Compression Garments in the UK’ has been written by an expert group in the UK and Ireland with the aim of proposing a simple new product grouping and classification. The document proposes the use of the terms “Group” and the sub-tier term of “Category”. Through the use of these two terms, it is hoped that confusion seen with Classes across the UK and Europe is avoided.

The new groups proposed are:

Group 1: Anti-embolism garments – garments which, when worn on the supine leg, exert graduated compression to the leg surface and are intended to reduce the incidence of deep venous thrombosis in non-ambulant patients.

Group 2: Graduated support garments – support garments which, when worn on the leg, exert support pressure to the leg surface for sub-clinical indications such as relief of aching legs. The pressure progressively reduces towards the upper leg. Such garments can be purchased from high street pharmacies and stores.

Group 3: Graduated compression garments – compression garments which, when worn on the leg, exert compression to the leg surface which reduces progressively towards the upper leg, and are used for management and prevention of lymphoedema and venous disease. Such garments are prescribed for management of clinical conditions as outlined below. This group 3, “Graduated Compression Garments” are divided into compression categories and pressure ranges as outlined in table 9.

Table 9. New categories and pressure ranges for graduated compression garments

Category	Pressure Range
Category IA	14-17 mmHg
Category IB	18-21 mmHg
Category II	22-30 mmHg
Category III	31-40 mmHg
Category IV	41-50 mmHg
Category IV Super	51+ mmHg

This guideline supports the use of this new terminology to re-categorise pressure ranges. Please see appendix III.II for the indications for use for each category. Please see the HSE National Guideline for ordering Compression Garments for the Prevention and Management of Chronic Oedema/Lymphoedema 2022, available at [hse.ie/lymphoedema](https://www.hse.ie/lymphoedema).

Types of compression garments

Compression garments can be off the shelf (OTS) or made to measure (MTM) depending on the diagnosis and patient needs, see criteria below.

Off the shelf

Patients requiring low level compression for non-complex lymphoedema with a normal limb shape will usually require an OTS garment and will not always need to be seen by a lymphoedema therapist but can be assessed by a HCP with training in measuring for OTS garments and who is able to give appropriate advice, exercise and skin care for the patient. Made to measure garments should be considered if OTS are uncomfortable or not effective.

Made to Measure

If a patient requires a MTM garment and there is limb shape distortion due to oedema, the patient should be seen by a lymphoedema therapist for a full assessment. Measurements for the garment should be taken and appropriate treatment prescribed. If lymphoedema treatment is effective and the limb shape normalises then the patient should be reassessed for suitability for an OTS garment if appropriate.

Some patients may require MTM due to irregular shape of the leg which is not due to oedema, in this case a lymphoedema therapist assessment is not required and measuring for the MTM garment can be completed by a HCP with appropriate training.

The fabric types in compression garments include:

Circular knit:

Circular knit garments are seamless, thinner and better tolerated by some patients than others, but are more likely to cause problems where there are skin folds. Some circular knit garments, have characteristics of both circular knit and flat knit garments. These may be suitable for some patients with minimal shape distortion and may improve tolerance of application.

Flat knit

Flat knit garments tend to be thicker and firmer with increased rigidity than circular knit and are more suitable when there are skin folds and more significant distortion of the limb.

Neoprene wraps

Neoprene wraps are available with Velcro fastening which allows ease of application, however their bulkiness limits practicality for daily use. They can also provide varying levels of compression and can be useful if there is a dressing on the limb which can be disrupted with donning and doffing of compression garments. Wraps can be ordered by lymphoedema clinicians and HCPs with the appropriate competencies in their practice.

Night compression systems

Night Compression systems are available for the management of fluid build-up overnight. Patients that require night compression systems should be assessed by a lymphoedema therapist.

Fitting and evaluation

A lymphoedema therapist or trained HCP should:

- Check the fit of a newly prescribed compression garment and ensure patients have the information they need to use their garments effectively and appropriately including the manufacturer laundering instructions; in general, garments should be washed daily.
- Demonstrate garment application and removal, and check that the patient or carer can perform these tasks.
- Educate patients regarding application aids and strategies to prevent garment slippage.
- Review new patients (face to face or by telephone) three weeks after fitting, and then after three to six months, at the practitioner's discretion, if fit and response to compression are satisfactory.
- During reassessment, seek to understand the patient's perspective of their progress and ensure that the level of compression is adequate. They should evaluate the patient/carer's ability to manage the garment and care for the affected limb.
- Pay particular attention to the presence of pain, which may indicate a problem such as ischemia, infection, or deep vein thrombosis. Patients should be evaluated to ensure that the garment does not cause damage to the foot or ankle.
- Reassess the patient if deterioration occurs to determine whether there is a change in overall health status and to explore whether there are issues affecting the patient's ability to self-manage e.g. their access to carer support.

Practitioners should consider whether an adaptation of the compression garment is required or whether a period of intensive treatment involving manual lymphatic drainage and multi-layer lymphoedema bandaging (MLLB) would improve the patient's clinical condition. The need for replacement garments should be reviewed every three to six months, or when they lose elasticity. Very active patients may need replacement garments more frequently.

Compression bandaging

Compression bandaging is used to reduce oedema and is mainly used in the intensive phase of treatment for lymphoedema. These bandages are multi-layered inelastic bandages which usually have a liner padding layer and bandages.

Members of the lymphoedema MDT

Involvement of the various members of the lymphoedema MDT depends on clinical presentation and may include: dermatologists, physiotherapists, occupational therapists, nurses, dietitians, podiatrists and psychologists.

General Lymphoedema

Risk Reduction and Awareness

Over the last decade there has been a plethora of research addressing risk reduction strategies for lymphoedema. This is mostly directed at cancer-related lymphoedema, however, newer approaches assess the increasing impact of obesity and dependency. Each Lymphoedema service should include risk reduction education (HCP and service user) as core activities. Cancer Prehabilitation is newly recognised as a core component of the cancer pathway and facilitates not only healthy living behaviour changes, but also a multi-professional opportunity to share risk reduction strategies with those at risk of developing lymphoedema.

Studies have shown that lymphoedema awareness is lacking in settings such as care homes, despite the prevalence of lymphoedema in care home populations (Thomas et al., 2020). The latest research addresses specifics of cancer management alongside BMI and this is reviewed in this section and additionally linked to in the [oncology section](#).

There has also been a recent update in NICE guidance on the topic of lymphoedema risk after breast cancer treatment (National Institute for Health and Care Excellence [NICE], 2018). This new guidance has challenged the more traditional 'risk reduction' content, such as blood taking and flying. This is also addressed and should guide local and regional patient information and education.

It is important that clinicians and patients are aware of their risk of developing lymphoedema so that risk reduction education can be tailored for the patients to ensure relevance and not overburdening patients. There are tools that clinicians can use to calculate the level of risk e.g. [Bundred \(2020\)](#).

Early detection of lymphoedema

Screening and surveillance pathways have been widely used to detect subclinical lymphoedema. This involves equipment that can detect early volume changes and should also include patient reported symptoms and clinical examination to ensure that early lymphoedema is detected (Figure 1 below). Patients treated in the subclinical stage may not develop lymphoedema with short-term interventions.

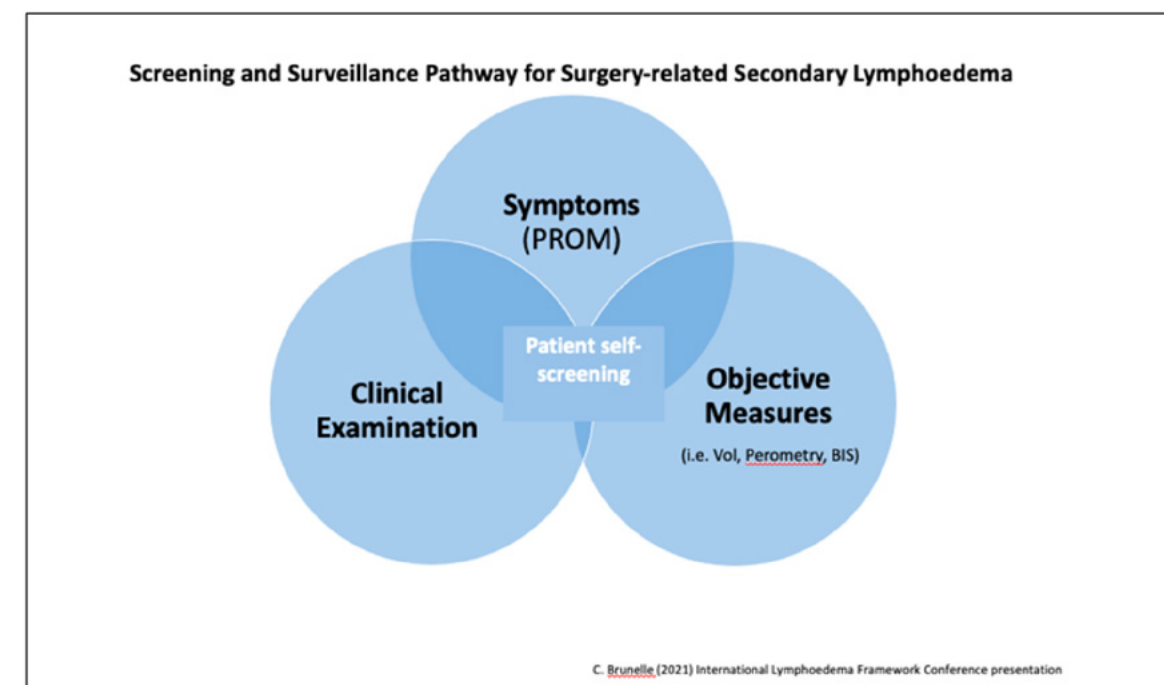


Figure 1. Screening and Surveillance Pathway for Surgery-related Secondary Lymphoedema

1.1 Risk Reduction

Risk factors for lymphoedema development

Research into risk factors associated with lymphoedema primarily pertains to breast cancer populations and almost exclusively pertains to patients with or treated for cancer. There are patient-related risk factors and treatment-related risk factors associated with the development of oncology related lymphoedema. There is also emerging evidence that increased BMI is a significant risk factor for lymphoedema development.

Patient-related risk factors

The following patient-related factors have been identified as placing oncology patients at risk of developing lymphoedema:

- Living with obesity
- Fluctuations in weight
- Dependency/Immobility
- A history of cellulitis or inflammation in the ipsilateral limb (Asdourian et al., 2016a)

Disease-related risk factors

The following disease-related factors have been identified as placing oncology patients at risk of developing lymphoedema:

- Number of positive lymph nodes at diagnosis
- Negative oestrogen receptor status (breast cancer patients)

Treatment-related risk factors

The following treatment-related factors have been identified as placing oncology patients at risk of developing lymphoedema:

- Degree of lymph node dissection (number of nodes removed)
- Radiotherapy
- Chemotherapy
- Biologic treatment
- Axillary cording
- Seroma formation

The main treatment-related risk factors for BCRL as per the literature, include axillary lymph node dissection (ALND) and regional lymph node radiation (RLNR). There is strong evidence that both ALND and RLNR are independent risk factors for BCRL. A large cohort study of breast cancer patients concluded that risk of developing lymphoedema is primarily related to the multimodal treatments chemotherapy and radiation (and not surgery alone), and disease stage. BCRL rates are higher in patients receiving chemotherapy, radiation, ALND, more advanced disease stage, and those with a higher body mass index at diagnosis (Nguygn, 2017).

GQ1: What are the risk factors for developing non-cancer related secondary lymphoedema?

Evidence Summary

Several studies have examined the risk factors for developing non-cancer related secondary lymphoedema (Byun et al., 2021). Evidence has shown the following factors are considered to increase risk of lymphoedema :

- Obesity (BMI >30 kg/m²)
- Age
- Dependency
- Paralysis
- Recurrent Cellulitis
- Dermatitis/Eczema
- Venous Thromboembolism (VTE)
- Chronic Venous Insufficiency/Venous Ulceration
- Venous Surgeries
- Hereditary risk
- Scarring
- Large circumferential wounds

Recommendations

GQ1.1 Patient information (written/online) should be developed and continuously refined to ensure those at risk of developing secondary non-cancer related lymphoedema are aware of the risk, and how to reduce their individual risk.

Evidence Grade: D

Strength of recommendation: Strong

GQ1.2 HCPs and students who are working with patients at risk of developing secondary non-cancer related lymphoedema should receive evidence based education to be able to provide awareness and risk reduction advice, taking into consideration benefit versus burden on the patient.

Evidence Grade: D

Strength of recommendation: Strong

GQ1.3 There should be partnership working between potential referring services and lymphoedema teams. Some services, e.g. bariatric management, should have staff with specialist lymphoedema skills (see [obesity section](#)).

Evidence Grade: D

Strength of recommendation: Strong



Education Need:

HCPs and students who are working with patients at risk of developing secondary non-cancer-related lymphoedema should receive education to be able to provide awareness and risk reduction advice. Refer to the education section for further guidance.

GQ2: Is increased BMI (>30) associated with an increased risk of developing lymphoedema?

Evidence Summary

A large prospective cohort study (n=787) examined the impact of pre-operative BMI on the risk of developing lymphoedema in patients being treated for breast cancer (Jammallo et al., 2013). The study found that pre-operative BMI of ≥ 30 was an independent risk factor for lymphoedema development, whereas a BMI of $25 < \text{BMI} < 30$ was not. The study also found that large post-operative weight fluctuations also increased the risk of developing lymphoedema. Patients with a pre-operative BMI ≥ 30 and those who experience large weight fluctuations during and after breast cancer therapy should be considered at higher-risk of developing lymphoedema.

A study by Helyer (2010) examined risk factors for developing lymphoedema after treatment for breast cancer involving SLND with or without an ALND. This study of 137 patients found that the risk of developing lymphoedema post-surgery was significantly related to the patients' BMI ($p = 0.003$). Multivariate analysis revealed patients with a BMI >30 (obese) had an odds ratio of 2.93 (95% CI 1.03-8.31) of developing lymphoedema, compared with those with a BMI of <25 . A longitudinal study (Ridner et al., 2011b) examining the relationship between BMI and BCRL found that pre-treatment, BMI may be a risk factor for developing BCRL. The study found that Breast Cancer patients with a BMI ≥ 30 at the time of treatment had 3.6 times greater risk of developing BCRL compared to those with a BMI under 30. The same effect was not seen in those with an increase in BMI or those with a rise in BMI greater than 30 after treatment. These findings were echoed in a 10 year follow up study which found patients who were obese at the time of surgery had an increased risk of BCRL (HR: 1.52) (Ribeiro Pereira et al., 2017). Even in early stage breast cancer, pre-op obesity has been linked to subclinical lymphoedema (Lyigun et al., 2018). A study examining lymphoedema development in gynaecological cancers also found the risk of lymphoedema was greater with increasing BMI at time of surgery (Hayes et al., 2017).

In a prospective cohort study (n = 486) of women who developed lymphoedema after cancer treatment, pre-morbid obesity was significantly associated with lymphoedema symptoms (Mehrra and Greene, 2014). The oedema component in obesity is associated with increased lymph production due to the increased ultrafiltration and overburdening of the lymphatic system, rather than a structural impairment.

People living with obesity (PWO) may be at risk of developing lymphoedema because they have compromised lymphatic function at baseline, abnormal inflammatory responses which can negatively impact the lymphatic system, and have impaired ability to regenerate damaged lymphatics after injury. Recent studies suggest that the interaction between obesity and lymphoedema is reciprocal. That is, not only is it now clear that obesity can lead to impaired lymphatic function, it is also evident that impaired lymphatic function can lead to adipose deposition and obesity.

Recommendations

GQ2.1. Clinicians should be aware that increased BMI >30 is associated with an increased risk of developing lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ2.2 Patients and/or their carers should be made aware that increased BMI >30 is associated with an increased risk of developing lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ3: What is the impact of weight gain or weight fluctuation on developing lymphoedema?

Evidence Summary

A longitudinal cohort study of patients after breast cancer surgery answered this question (Jammallo et al., 2013). A post-op weight fluctuation of 10 pounds (gained or lost) per month was associated with a significantly greater risk of developing lymphoedema (HR: 1.97). The authors concluded that a pre-op BMI ≥ 30 and large fluctuations in BMI post-op were associated with an increased risk of lymphoedema development. A study of breast cancer patients undergoing neoadjuvant chemotherapy found that while BMI was an independent risk factor of the development of lymphoedema, change in body weight was not a significant factor in the incidence of lymphoedema (Park et al., 2018).

Recommendations

GQ3.1 Clinicians should be aware that a reduction in BMI to under 30, and avoiding significant fluctuating weight patterns (10 pounds or more lost or gained) can reduce the risk of developing lymphoedema post operatively.

Evidence Grade: C

Strength of recommendation: Strong

GQ3.2 Patients at high risk of lymphoedema should be educated that reducing their BMI to under 30 and avoiding fluctuating weight patterns can reduce the risk of developing lymphoedema. Clinicians should consider referral to a dietitian or local weight management service if clinically indicated.

Evidence Grade: D

Strength of recommendation: Strong

GQ4: Does exercising increase the risk of developing lymphoedema in at-risk patients?

Evidence Summary

A 2015 Cochrane Systematic Review (Stuiver et al., 2015) examined conservative interventions for preventing clinically detectable upper-limb lymphoedema in patients at risk of BCRL. The review examined ten trials (n = 1,205) and concluded that the quality of the evidence generated by these trials was low, due to risk of bias in the included trials and inconsistency in the results. The existing evidence does not indicate a higher risk of lymphoedema when starting shoulder-mobilising exercises early after surgery compared to a delayed start (i.e. seven days after surgery). Shoulder mobility (that is, lateral arm movements and forward flexion) is better in the short term when starting shoulder exercises earlier compared to later. The evidence suggests that progressive resistance exercise therapy does not increase the risk of developing lymphoedema, provided that symptoms are closely monitored and adequately treated if they occur. These findings were echoed in a recent large systematic review of 23 papers examining the impact of resistance exercise on BCRL. The authors of this review concluded that resistance exercise appears safe and does not increase the risk of developing lymphoedema in at-risk patients (Hasenoehrl et al., 2020).

These findings were mirrored in recommendations by the expert panel convened for the American Society of Breast Surgeons (McLaughlin et al., 2017a), who made the following recommendation: "The Panel agrees that clinicians should encourage at-risk and affected lymphoedema patients to exercise. Resistance and aerobic exercise is safe. Patients with BCRL should work with a trained lymphoedema professional to learn to exercise safely". The 2018 NICE guidelines on Early and locally advanced Breast Cancer Management also recommend "When informing people with breast cancer about the risk of developing lymphoedema, advise them that they do not need to restrict their physical activity".

Recommendations

GQ4.1 At-risk patients should be encouraged to exercise and be advised that gradual, progressive resistance and aerobic exercise is safe.

Evidence Grade: A

Strength of recommendation: Strong

GQ4.2 In the acute phase, any immediate post-operative exercise undertaken should be under the guidance of a physiotherapist and should be in keeping with the treating surgeons' protocol.

Evidence Grade: D

Strength of recommendation: Strong

GQ4.3 At-risk patients should be advised they are not required to limit their physical activity or limit use of their at-risk limb.

Evidence Grade: D

Strength of recommendation: Strong

GQ4.4 At-risk patients should be advised to increase their activity levels gradually.

Evidence Grade: D

Strength of recommendation: Strong

GQ4.5 Advice should be given that when doing physical activity, the affected limb or body part should be closely monitored for changes and patients should be encouraged to seek advice from a lymphoedema clinician as soon as possible if they notice a change in the limb or body part.

Evidence Grade: D

Strength of recommendation: Strong

GQ5: Are there any tools available which risk stratify lymphoedema risk factors?

Evidence Summary

Byun (2022) constructed a nomogram for predicting the risk of arm lymphoedema following typical breast cancer treatment. This nomogram should be validated in patients with different background characteristics before use. In total, 15.6% (n=1377) of patients developed lymphoedema. The median time from surgery to lymphoedema development was 11.4 months. Characteristics identified more frequently in patients who developed lymphoedema had significantly higher BMI (median, 24.1 kg/m² vs. 23.4 kg/m²), a greater number of removed nodes (median, 17 vs. 6) and more frequently underwent taxane-based chemotherapy (85.7% vs. 41.9%), total mastectomy (73.1% vs. 52.1%), conventionally fractionated radiotherapy (71.9% vs. 54.2%), and regional nodal irradiation (70.7% vs 22.4%) than those who did not develop lymphoedema (all P < 0.001). This nomogram was validated in a large multi-institutional cohort and statistically predicted lymphoedema risks were well correlated with the actual lymphoedema rates.

A study by Bundred et al. (2020) examined the risk of developing lymphoedema after axillary lymph node clearance as part of breast cancer treatment. This paper describes a scoring system, stratifying patients as high (76.7% risk of developing LO), moderate (32% risk of developing LO) or low risk (12 % risk of developing LO).

A number of other nomograms have recently been developed which may assist in assessing and predicting risk of lymphoedema in patients treated for cancer (Liu et al., 2021, Li et al., 2017, Gross et al., 2019).

Recommendation

GQ5.1 Clinicians should use a validated tool to assess patient's individual risk of developing lymphoedema.

Evidence Grade: C

Strength of evidence: Strong

GQ6: Is lymph node dissection associated with a greater risk of lymphoedema than radiation therapy?

Evidence Summary

The risk factors for developing lymphoedema following treatment for cancer have been identified as the number of nodes removed, radiotherapy and chemotherapy. Nguyen et al 2017 showed that the groups with highest risk (>25% at 5 years) involved ALND with nodal radiotherapy and/or anthracycline/cytosine + taxane chemotherapy. They concluded that factors significantly associated with BCRL were ALND, chemotherapy, radiation, and obesity. Bundred et al (2020) developed a scoring model for risk to create a total (0 to 5) of sub scores. Sub scores were allocated for RAVI at one month post-op (0–2), BMI (0–0.5) with highest score for BMI >30, ER status (0–0.5) with highest score for ER -, number of positive nodes involved (0–1) with highest score for ≥10 positive nodes, and chemotherapy (0–1) with highest score for chemotherapy with taxane. Based upon this population, using the model scores, potentially 66% of patients could be reassured regarding their low lymphoedema risk (11.6%), and resources concentrated on the moderate (30% risk) and high (4% risk) groups for lymphoedema surveillance.

Armer et al (2019) concluded that a higher risk of lymphoedema was associated with a BMI>30, Neo Adjuvant Chemotherapy for 144 days or longer, >30 nodes removed, increasing number of positive nodes and poor levels of activity. Naoum et al (2019) demonstrated a risk of lymphoedema due to SLNB alone (7.7%), SLNB+RLNR (regional lymph node radiation) (10.8%), ALNB alone (29%) and ALND+RLNR (38.7%). A trial comparing radiotherapy to ALND in breast cancer patients with positive sentinel lymph nodes found that lymphoedema was noted significantly more often after ALND (13% had objective evidence and 23% subjective symptoms) than after axillary radiotherapy (5% objective and 11% subjective) at 1 year, 3 years, and 5 years (Donker et al., 2014). Cormier et al (2010) reported an increase in risk associated with extended radiotherapy: to breast or chest wall (14.5%), to breast or chest wall and supra clavicular area (31.5%) and to breast or chest wall, supra clavicular area plus posterior axillary boost (41.4%). In conclusion, breast cancer patients with >30 nodes removed, NAC for >144 days, chemo with taxane, and RT to supraclavicular area and/or posterior axillary boost appear to be at high risk of developing lymphoedema.

A prospective study examining risk associated with treatment of gynaecologic cancer reported that limb volume change decreased with age greater than 65, but increased with a lymph node removal count greater than 8 in a cohort of endometrial cancer patients. There was no association with radiation or other risk factors (Carlson, 2020). The incidence of limb volume change of ≥10% was 34% (n = 247) in endometrial cancer, 35% (n = 48) in cervical, and 43% (n = 18) vulval cancer patients.

Recommendations

GQ6.1: Clinicians should be aware that there is an increased risk of developing lymphoedema associated with lymph node dissection and less of a risk with radiation or chemotherapy.

Evidence: D

Strength of Evidence: Strong

GQ6.2: Clinicians should use a validated tool to assess patient's individual risk of developing lymphoedema.

Evidence: C

Strength of Evidence: Strong

GQ7: Does the number of nodes removed during nodal dissection increase the risk of developing lymphoedema?

Evidence Summary

A prospective study of patients who underwent axillary lymph node dissection for the treatment of breast cancer, showed neoadjuvant chemotherapy and adjuvant chemotherapy using docetaxel and cyclophosphamide increased the risk of developing lymphoedema post-surgery (Akezaki 2019). Findings of a large prospective trial have suggested that the time course for lymphoedema development may depend on the breast cancer treatment received. This study found ALND was associated with early-onset lymphoedema, and RLNR was associated with late-onset (McDuff et al., 2019).

A prospective study examining risk associated with treatment of gynaecologic cancer reported that limb volume change decreased with age greater than 65, but increased with a lymph node removal count greater than 8 in a cohort of endometrial cancer patients. There was no association with radiation or other risk factors (Carlson 2020). The incidence of limb volume change of ≥10% was 34% (n = 247) in endometrial cancer, 35% (n = 48) in cervical, and 43% (n = 18) vulval cancer patients.

A trial comparing radiotherapy to ALND in breast cancer patients with positive sentinel lymph nodes found that lymphoedema was noted significantly more often after ALND (13% had objective evidence and 23% subjective symptoms) than after axillary radiotherapy (5% objective and 11% subjective) at 1 year, 3 years, and 5 years (Donker et al., 2014).

Recommendations

GQ7.1. Clinicians should be aware that increased number of nodes dissected is associated with an increased risk of developing lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ7.2 Patients and/or their carers should be made aware that increased number of nodes dissected is associated with an increased risk of developing lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ8: Do patients with axillary cording have an increased risk of developing lymphoedema?

Evidence Summary

Axillary cording has also been identified as a risk for developing BRCL. Patients who reported cording had 2.4 times the odds of developing BCRL compared to those who did not. Patients affected with cording most frequently reported these symptoms: tenderness (61.2%), aching (60.7%), and firmness/tightness (59.8%) (Brunelle 2020).

Recommendations

GQ8.1. Clinicians should be aware of that axillary cording is associated with an increased risk of developing lymphoedema.

Evidence Grade: D
Strength of recommendation: Strong

GQ8.2 Patients and/or their carers should be made aware that axillary cording associated with an increased risk of developing lymphoedema.

Evidence Grade: D
Strength of recommendation: Strong

GQ9: What is the impact of scar formation on lymphoedema development?

Evidence Summary

One small study (Warren and Slavin, 2007) examined 11 patients with localized swelling along with linear or curvilinear scars. Radiocolloid lymphoscintigraphy was performed on patients by single or multiple injections, into the site of the scar corresponding to the local oedema. In 8 patients there was no evidence of lymphatic drainage traversing or bridging the scar. In 2 patients with multiple prior Z-plasty revisions there was no visualization of lymph channels across the Z-plasty flaps. Overall, lymphoedema in the area adjacent to or enclosed within the scar was diagnosed in 8 out of the 11 patients.

The authors concluded that these findings suggest that undrained lymphatic fluid contributes to the pathogenesis of the raised and swollen tissues in scar formation. Furthermore, lymphatic pathways do not appear to re-establish themselves across scars, therefore attempting to improve lymphatic flow with Z-plasty revisions may not be successful in certain scar deformities.

Depending upon the type of surgery and other related factors, scar tissue can extend from the skin, down to bones and viscera. Sometimes these scars become hard and inflexible and obstruct lymphatic flow. Post-surgical exercises help to mobilise the area and stretch the scar, but there can still be issues with scar tethering and additional therapeutic massage may be required.

Recommendations

GQ9.1 Clinicians should be aware of the impact of scarring on lymphatic tethering.

Evidence Grade: D
Strength of recommendation: Strong

GQ9.2 Clinicians treating patients with or at risk of lymphoedema should be trained in simple scar management.

Evidence Grade: D
Strength of recommendation: Strong

GQ9.3 Patients should be taught simple scar massage after surgery.

Evidence Grade: D
Strength of recommendation: Strong

GQ9.4 Symptomatic scars should be referred to physiotherapy for specialist management.

Evidence Grade: D
Strength of recommendation: Strong

GQ10: What is the impact of poor posture on lymphoedema development?

Evidence Summary

Changes to the distribution of body mass and centre of gravity may impact normal posture. These can include surgery e.g. mastectomy, increase in weight or joint injury. Swelling will also affect the ability of a joint to correctly articulate, and chronic swelling may have a significant impact on joints, gait and general independence. Immediately after surgery, a patient may restrict movement and alter posture as a form of protection, for example, a forward lean and curved upper body posture after breast surgery. Over time however, this alteration can cause some tissues to tighten and constrict, and others to remain stretched and weaken.

Celenay et al. (2020) found an increase in the sagittal thoracic curvature and the frontal inclination angle towards the unaffected side in women post-modified radical mastectomy with unilateral lymphoedema.

Angin et al. (2014) suggest that unilateral upper extremity lymphoedema may have challenging effects on postural balance. Any deviation from the normal centre of gravity by leaning towards any direction on the supporting surface is described as postural sway, which is an active process to find the best possible position at a given moment. In their study the lymphoedema group showed a significant increase in postural sway velocity in the unilateral stance test on the ipsilateral leg with eyes open ($p = 0.02$) and eyes closed ($p = 0.005$), as well as on the contralateral leg with eyes open ($p = 0.004$) and eyes closed ($p = 0.0001$).

Recommendations

GQ10.1 Clinicians should be aware of the impact of poor posture on lymphoedema, particularly after operations which may lead to poor posture e.g. breast surgery.

Evidence Grade: D
Strength of recommendation: Strong

GQ10.2 Clinicians treating patients with or at risk of lymphoedema should encourage good postural alignment to facilitate range of movement, aid lymphatic flow and overall balance and alignment. Referral to a physiotherapist to discuss physical activity e.g. aerobic or resistance exercise as appropriate may be helpful in supporting improved posture.

Evidence Grade: D
Strength of recommendation: Strong

GQ10.3 Clinicians should consider referral to physiotherapy and/or podiatry if they observe abnormal gait to prevent further joint damage.

Evidence Grade: D
Strength of recommendation: Strong

GQ11: Should patients be given advice on risk reduction practices if they are at risk of developing lymphoedema?

Evidence Summary

The only consistent precautionary advice recommended for at-risk patients appears to be against excess weight gain and infection prevention. NICE recommends that patients with early breast cancer should be informed of the risk of developing lymphoedema and should be provided with relevant written information prior to surgery or radiotherapy. NICE also recommends advising patients in relation to preventing infection and trauma, which may exacerbate or cause breast cancer-related lymphoedema in at-risk patients (NICE, 2018).

Most of the more recent evidence and international guidance appears to call into question the rationale for risk reduction advice and highlights the lack of scientific evidence to support much of the long-standing precautionary guidance.

Much of the historical evidence advocating to provide at-risk patients with risk reduction behaviours originates from small retrospective studies, case series or expert opinion based on physiology and a “common sense approach”. The American Society of Breast Surgeons convened an expert panel to review current data and guidelines on all aspects of lymphoedema (McLaughlin et al., 2017b). In 2017, the panel published the following recommendation: “The Panel agrees that within the context of an early detection/surveillance program incorporating baseline and follow-up assessments, the routine application of many risk-reducing behaviours is not supported.....Personalized risk-reduction strategies are more appropriate than blanket application of behaviours.” In their 2016 Consensus document, the International Society of Lymphology issued similar guidance, by cautioning standard use of risk reduction advice: “the risk of secondary lymphoedema is much less with conservative breast cancer treatments (i.e. SLNB) such that the standard use of some of these “don’ts” for the risk reduction of lymphoedema may not be appropriate and possibly subject patients to therapies that are unsupported.”

The largest prospective cohort study to date on this topic was carried out on patients (n = 632; totalling 760 at-risk limbs) undergoing breast cancer treatment at one institution from 2005-2014 (Ferguson et al., 2016). They found there was no significant association between relative volume change or weight-adjusted change increase, and undergoing one or more blood draws (P = .62), injections (P = .77), number of flights (one or two [P = .77] or three or more [P = .91] v none), or duration of flights (1 to 12 hours [P = .43] and 12 hours or more [P = .54] v none). However, the authors cautioned that they: “cannot affirmatively state that risk reduction practices have no effect on arm swelling”. Other authors went on to extrapolate these findings and claimed this study debunked conventional guidance about lymphoedema prevention, stating that only weight gain and infection are proven to cause Breast Cancer-Related Lymphoedema (BCRL) and therefore counsels providers to give less cautionary advice to at-risk patients (Ahn and Port, 2016).

It is important to note that the majority of research is based on studies examining the breast cancer population. As there is a lack of research in other types of cancer at this time, it is not possible to draw firm conclusions regarding risk reduction advice and its role in other populations.

Recommendations

GQ11.1 General advice about lymphoedema should be offered to all patients at risk of lymphoedema, including skin care, weight management, and the importance of sustainable physical activity.

Evidence Grade: D

Strength of recommendation: Strong

GQ11.2 Tailored risk reduction advice should be given and discussed with each patient depending on their individual risk.

Evidence Grade: D

Strength of recommendation: Strong

GQ11.3 Risk reduction advice should include provision of written patient information leaflets or information regarding appropriate websites or applications, ensuring the patient understands the contents of same through open discussion.

Evidence Grade: D

Strength of recommendation: Strong

GQ12: Which skin care precautions assist in the prevention of lymphoedema?

Evidence Summary

Much of the evidence in relation to skin care in lymphoedema pertains to established lymphoedema, and the avoidance of factors which may exacerbate it, such as extreme temperature changes and prevention of skin infections. Several studies, mostly small and retrospective in nature, have demonstrated a link between skin infection and lymphoedema such that the evidence appears to support the avoidance of infection as potentially protective against lymphoedema (Asdourian et al., 2016a). The impact of extreme temperature on lymphoedema is discussed in [GQ16](#) and [17](#).

NICE guidelines on managing Early Breast Cancer recommend giving at-risk patients advice on how to prevent infection or trauma that may cause or worsen lymphoedema (NICE, 2018). In their 2016 consensus document on the treatment of peripheral lymphoedema, the International Society of Lymphology also recommend that patients are educated to understand the importance of skin hygiene and meticulous skin care. They recommend that patients be educated on cleansing, using low pH lotions and appropriate emollients. Similarly, the International Union of Phlebology recommend skin care as a fundamental part of the treatment of lymphoedema (Lee et al., 2013a).

The National Lymphoedema Network recommends that those at risk of lymphoedema are told skin infections require urgent medical care, due to the risk of development of cellulitis (Damstra and Halk, 2017). The Dutch Lymphoedema guidelines recommend instructing patients on skin care at all stages of treating lymphoedema, including secondary prevention. It is important that patients are able to notice skin changes and know when to report them to their treating clinician. Particularly in the maintenance phase, skin care is very important, as patients will have less interaction with healthcare providers who may have otherwise picked up on changes. The Dutch guidelines recommend patients avoid trauma or any activity which may result in a break in the skin. When defects do occur, they recommend using antiseptic treatment. They also recommend patients wear gloves or protective creams if they are partaking in an activity which poses a risk of skin trauma or infection.

Recommendations

GQ12.1 All patients at risk of lymphoedema should be informed, by the treating HCP, of the importance of meticulous daily skin care and avoid trauma or any activity which may result in a break of the skin. Refer to questions GQ 41-44 for further advice on skin care.

Evidence Grade: D

Strength of recommendation: Strong

GQ12.2 Patients should be informed that signs of skin infections should be given immediate medical attention.

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Signs and symptoms of skin infection

- Fever
- Pus or fluid leaking from a cut
- Red skin around an injury
- Sores that look like blisters
- Progressive pain
- Progressive swelling

GQ13: Should needle insertion be avoided in the at-risk limb?

Evidence Summary

The vast majority of evidence that opposes or supports venepuncture in at-risk arms is from small studies, case reports, retrospective questionnaires or expert opinion. The main evidence for needlestick avoidance originates in a single study of 114 patients by Britton and Nelson (Britton and Nelson, 1962), which found that in patients who developed lymphoedema after radical mastectomy, 53% had a history of recurrent cellulitis following either an insect bite, cat scratch, needle or thorn prick. A comprehensive review of the literature on needle stick avoidance (Cheng et al., 2014a) found no rigorous evidence-based support for the risk-reduction behaviours of avoiding blood pressure monitoring and venepuncture in the affected arm in the prevention of lymphoedema after axillary procedures. These findings are in congruence with several studies (Ren et al., 2019, Ferguson et al., 2016, Showalter et al., 2013), all of which failed to find an association between venepuncture and BCRL.

However, two cohort studies did find an association between needle stick procedures. A large prospective study (Kilbreath et al., 2016) examined patients who underwent axillary surgery and assessed a number of post-surgical events in a sub-group of women with > 5 axillary nodes removed and who maintained weekly diaries and found only blood drawn from the ‘at-risk’ arm was identified as a potential risk (OR 2.0; 0.8, 5.2; p=0.17), there were other practices not associated with lymphoedema including injection to the arm (OR 1.0; 0.3, 2.7; p=0.92). One study found hospital skin puncture (vs. none) (RR 2.44, 95%CI 1.33–4.47) was associated with BCRL with 44% patients having a history of a needle stick procedure developing lymphoedema, compared to 18% of those without (Clark et al., 2005).

NICE guidance (NICE, 2018) states that there is “no consistent evidence of increased risk of lymphoedema associated with medical procedures (for example, blood tests, injections, intravenous medicines and blood pressure measurement) on the treated side, and the decision to perform medical procedures using the arm on the treated side should depend on clinical need and the possibility of alternatives.” In contrast to NICE, Dutch Guidelines recommend that the medical procedures such as venepuncture be taken from the healthy arm. If this is unavoidable they recommend strict antiseptic precaution to be taken on the affected arm.

Refer to GQ20 and GQ91 for advice on acupuncture.

Recommendations

GQ13.1 Although the evidence against needle insertion on the affected side is poor, it is recommended to avoid using the at risk limb and use the non-affected limb for needle insertion where possible.

Evidence Grade: D

Strength of recommendation: Strong

GQ13.2. Personalised risk-reduction strategies are more appropriate than blanket application of behaviours.

Evidence Grade: D

Strength of recommendation: Strong

GQ13.3 In patients who undergo a second mastectomy without axillary surgery, this arm is generally not at risk of developing lymphoedema and needle insertion may be carried out on this arm.

Evidence Grade: D

Strength of recommendation: Strong

GQ14: Is there evidence to suggest that taking blood pressure on an affected limb affects outcome in patients at risk of developing lymphoedema?

Evidence Summary

Two thorough reviews of studies examining whether medical procedures increase risk of lymphoedema found no rigorous evidence-based support for the risk-reduction behaviours of avoiding blood pressure monitoring in the at risk arm after an axillary procedure for breast cancer (Asdourian et al., 2016b, Cheng et al., 2014b). Multiple studies in several settings have failed to find an association between BP monitoring in the ipsilateral arm and an increased risk of BCRL (Ferguson et al., 2016, Kilbreath et al., 2016, Showalter et al., 2013, Mak et al., 2009, Hayes et al., 2005).

The American Society of Breast Surgeons expert panel reviewed current data and guidelines on all aspects of lymphoedema. In 2017, the panel published the following recommendation: “The Panel agrees that ... Use of the ipsilateral arm for IVs or blood pressures is not contraindicated, although most patients prefer to use the contralateral arm” (McLaughlin et al., 2017c). NICE offer similar guidance in their guidance regarding complications of early breast cancer (NICE, 2018). They state there is “no consistent evidence of increased risk of lymphoedema associated with medical procedures (for example...blood pressure measurement) on the treated side, and the decision to perform medical procedures using the arm on the treated side should depend on clinical need and the possibility of alternatives.

It should be noted that all of the available evidence examines once-off blood pressure monitoring in the at-risk limb and does not include sustained blood pressure monitoring which occurs in cases such as during surgical procedures, in intensive care and while wearing ambulatory blood pressure monitoring devices. Therefore conclusions regarding the risk of developing lymphoedema in these circumstances cannot be reached, hence caution should be exercised in these circumstances.

Recommendations

GQ14.1 Although the evidence against blood pressure monitoring on the affected side is poor, patients should be advised to try avoid doing so and use the alternative limb instead, where possible, particularly in situations requiring sustained blood pressure monitoring (e.g. during surgery)

Evidence Grade: D

Strength of recommendation: Strong

GQ14.2 It is recommended that personalised risk-reduction strategies are more appropriate than blanket application of behaviours.

Evidence Grade: D

Strength of recommendation: Strong

GQ14.3 In patients who undergo a second mastectomy without axillary surgery, this arm is generally not at risk of developing lymphoedema and may be used to assess blood pressure.

Evidence Grade: D

Strength of recommendation: Strong

GQ14.4 In patients who undergo bilateral treatment for breast cancer, both arms may be at risk so the lower limb may be used to assess blood pressure where possible.

Evidence Grade: D

Strength of recommendation: Weak



Research Idea:

The impact of continuous blood pressure monitoring in limbs at risk of developing lymphoedema should be explored.

GQ15: Are alert bracelets effective in reducing the risk of lymphoedema in at-risk patients?

Evidence Summary

While there is no evidence examining the use of alert bracelets in patients at risk of lymphoedema, their efficacy has been shown in other patient populations, for example in preserving veins for future dialysis in patients with chronic kidney disease (Vachharajani, 2009). Therefore further research into the use of alert bracelets in patients at risk of developing lymphoedema should be carried out.

Recommendation

GQ15.1 Prior to making any recommendation, further research needs to be carried out to assess the effectiveness of alert bracelets in lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ16: Can extremes of temperature increase the risk of developing lymphoedema or worsen lymphoedema?

Evidence Summary

There is little evidence available to answer this question. The National Lymphoedema Network, the International Lymphoedema Framework, and many other guidance and best practice documents, have in the past recommended avoiding extremes of temperature in patients at risk of lymphoedema. This advice was based on a “common sense approach” and is not backed up by scientific evidence. The rationale for such recommendations is based on the theory that being exposed to extreme cold may cause a rebound increase in circulation, causing vasodilation, which in turn leads to swelling secondary to increased lymphatic load (Dell and Doll, 2006; Cemal et al., 2011).

Recommendations

GQ16.1 Patients should be advised to avoid exposure to extreme heat or cold to the extent that tissue injury could occur such as burns, sunburn or frostbite i.e. any temperature change that causes the skin to become severely red.

Evidence Grade: D

Strength of recommendation: Strong

GQ16.2 When informing patients at risk of developing lymphoedema, advise them that there is no consistent evidence of increased risk of lymphoedema associated with travel to hot countries.

Evidence Grade: D

Strength of recommendation: Strong

GQ16.3 Patients at high risk of developing lymphoedema should be advised to discuss their individual risk with their clinician.

Evidence Grade: D

Strength of recommendation: Strong

GQ17: Does the use of saunas, steam rooms or Jacuzzis increase the risk of developing lymphoedema in at-risk patients?

Evidence Summary

There is a paucity of evidence available on this subject. A sub-analysis of a large trial examining physical activity and lymphoedema (Showalter et al., 2013) was the first and apparently only evidence, to show that sauna use increases the risk of BCRL. The authors advocated for advising patients against the use of saunas. Contrastingly, prospective evidence on other sources of heat such as exercise, sunlight, sunburn and hot baths has failed to demonstrate an increase the risk of BCRL (Cemal et al., 2011).

NICE guidance (NICE, 2018) on breast cancer management recommends:

“When informing people with breast cancer about the risk of developing lymphoedema, advise them that: there is no consistent evidence of increased risk of lymphoedema associated with air travel, travel to hot countries, manicures, hot-tub use ...”

Recommendations

GQ17.1 At-risk patients should be advised that there is no consistent evidence of increased risk of lymphoedema associated with prolonged use of whole body heat environments however they should be used with caution and patients should be educated to self-monitor their response to concentrated heat.

Evidence Grade: D

Strength of recommendation: Strong

GQ17.2 Patients at high risk of developing lymphoedema should be advised to discuss their individual risk with their clinician.

Evidence Grade: D

Strength of recommendation: Strong

GQ17.3 Patients at-risk of, or with lymphoedema, should be advised to avoid hot-tubs, Jacuzzis and saunas if they have any cuts or open wounds as there is a risk of developing cellulitis.

Evidence Grade: D

Strength of recommendation: Strong

GQ18: Can air travel increase the risk of developing lymphoedema?

Evidence Summary

Advice against air travel to prevent BCRL is largely derived from one study (Casley-Smith and Casley-Smith, 1996) which stated that low cabin pressure experienced when flying causes lymphoedema. A number of studies published since have refuted this claim (Graham, 2002, Mak et al., 2009, Swenson et al., 2009, Showalter et al., 2013, Kilbreath et al., 2016, Ferguson et al., 2016, Asdourian et al., 2017) and international guidance does not advocate for or against air travel in patients with or at risk of lymphoedema.

Recommendations

GQ18.1 When informing people at risk of developing lymphoedema, advise them that there is no consistent evidence of increased risk of lymphoedema associated with air travel. Governments and airlines recommend all travellers (both at risk and not at risk of lymphoedema) keep hydrated and mobilise as much as possible during air travel.

Evidence Grade: D

Strength of recommendation: Strong

GQ18.2 Patients deemed at high risk of lymphoedema should seek travel advice from their treating HCP before going on long haul flights (i.e. 4 hours or more) and, if possible, an assessment for subclinical lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ19: Does wearing compression garments when flying reduce the risk of developing lymphoedema in at-risk patients?

Evidence Summary

A large review of risk reduction behaviours for lymphoedema prevention published in the Lancet Oncology (Asdourian et al., 2016a) provides a thorough review of compression garments and flying. Upon reviewing the existing evidence this review concludes that no consensus on the use of compression garments while flying exists within the literature and there are very few studies published examining the utility of compression garments as prophylaxis while flying. One small study questions the safety of compression garments when flying as it found an association between garment wearing and increased rates of lymphoedema.

However many clinicians still recommend the use of compression garments during flights, and several authors report that in their clinical experience, patients anecdotally report on the helpfulness and increased comfort provided by compression garments during long periods of air travel. In summary there insufficient evidence to suggest whether prophylactic compression garment use while flying is or is not of benefit to patients at risk of lymphoedema.

Recommendations

GQ19.1 Compression garments are not recommended prophylactically for flying in patients at risk of developing lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ19.2 Patients deemed at high risk of lymphoedema should seek travel advice from their treating HCP before going on long haul flights (i.e. 4 hours or more) and, if possible, an assessment for subclinical lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ19.3 Patients with established lymphoedema should wear their prescribed compression garments when flying.

Evidence Grade: D

Strength of recommendation: Strong

GQ20: Does acupuncture increase the risk of developing lymphoedema in at-risk patients?

Evidence Summary

There is no direct evidence examining the impact of acupuncture and lymphoedema occurrence in at-risk patients.

Expert opinion advises against the use of acupuncture in the affected quadrant due to the risk of infection from multiple needles over an extended period. Refer to questions GQ13 and GQ91 for further advice on needle use.

Recommendation

GQ20.1 While there is a lack of evidence regarding the risk of acupuncture in at-risk patients, where possible acupuncture should be avoided in the affected quadrant in these patients. (Refer to question GQ13 and GQ91 for further advice on needle use).

Evidence Grade: D

Strength of recommendation: Strong

GQ21: Does hair removal increase the risk of developing lymphoedema in at-risk patients?

Evidence Summary

There were no studies available to answer this question. Multiple documents and online resources advocate for using electric razors over blade razors or avoiding hair removal entirely, however there is no scientific evidence available to support these statements. There seems no reason to advise at-risk patients to avoid shaving.

Recommendation

GQ21.1 As there is no evidence to suggest hair removal increases risk of lymphoedema, patients may carry out hair removal as per their preferred method, ensuring caution and strict hygiene practices around the affected area.

Evidence Grade: D

Strength of recommendation: Strong

1.2 Diagnosis and Assessment

Lymphoedema can be diagnosed as either sub-clinical or clinical using a selection of techniques which are discussed in this section.

Measurements should be both subjective and objective using standardised formats, and should ensure that patient self-assessment is supported by education of the at risk patient and also their key HCP (where appropriate). Early detection ensures that timely referral will produce the best management outcomes with the least negative impact on the patient, and best use of clinical resources. Standardised assessment forms for HSE staff are available on [hse.ie/lymphoedema](https://www.hse.ie/lymphoedema).

A thorough assessment is crucial to ensure an holistic management plan is agreed by both HCP and patient, and to address all aspects of care associated with the patient, and associated onward referral.

GQ22: How should lymphoedema be diagnosed?

Evidence Summary

The general consensus from the reviewed literature is that lymphoedema is primarily a clinical diagnosis based on a thorough history and physical examination including the measurement of limb volume (Cormier et al., 2010, Grada and Phillips, 2017, Kayıran et al., 2017, Wanchai et al., 2016, Fu, 2014, Bernas, 2013, Paskett et al., 2012, International Society Of Lymphology, 2016, Damstra et al., 2017, Levenhagen et al., 2017, New South Wales Agency for Clinical Innovation, 2018). However, a number of specific diagnostic tests are available and may be useful in certain circumstances or if the diagnosis is unclear (Grada and Phillips, 2017, Kayıran et al., 2017, Wanchai et al., 2016, Bernas, 2013, International Society Of Lymphology, 2016, New South Wales Agency for Clinical Innovation, 2018). The criteria for the diagnosis and assessment of lymphoedema remain undefined (Cormier et al., 2010, Kayıran et al., 2017, Wanchai et al., 2016, Fu, 2014) however, a ≥ 2 cm increase in limb circumference and/or a ≥ 200 mL increase in limb volume are common parameters used for diagnosis in unilateral upper limb swelling in a suspected case of BCRL (Wanchai et al., 2016, Fu, 2014, Lymphology, 2016).

Both subjective and objective measures have a role in the diagnosis of lymphoedema (McLaughlin et al., 2017c, Cormier et al., 2010, Levenhagen et al., 2017). The patient should be encouraged to self-report any lymphoedema-related symptoms. Identification of swelling, heaviness and numbness may assist in the diagnosis of subclinical or early stage lymphoedema (Levenhagen et al., 2017). The presence of such symptoms should prompt the use of subjective assessment methods, and subsequently, may facilitate early diagnosis (Levenhagen et al., 2017). Subjective measures alone may not suffice for the diagnosis of lymphoedema as one systematic review highlighted that the use of objective measures resulted in increased identification of patients with lymphoedema compared to subjective measures (Cormier et al., 2010). Thus, a combination of both subjective and objective assessment measures may be the most appropriate method for the diagnosis of lymphoedema.

There is conflicting evidence regarding the best methods for lymphoedema diagnosis. However, there is a consensus amongst some of the literature that there is no one ideal method for lymphoedema diagnosis (Cormier et al., 2010, DiSipio et al., 2013a, Sierla et al., 2018, McLaughlin et al., 2017c, Levenhagen et al., 2017). One systematic review suggests that the use of multiple methods of lymphoedema diagnosis would be more suitable than the use of one method alone, as the incidence of lymphoedema was highest when assessed using > 1 method (DiSipio et al., 2013a).

Some of the literature reported lymphoscintigraphy (LSG) as the gold standard measure for lymphoedema to confirm diagnosis (Abbaci et al., 2019, Burnier et al., 2017, Grada and Phillips, 2017) and useful for depicting a specific lymphatic abnormality (International Society Of Lymphology, 2016). However these studies have also highlighted the limitations of LSG including the required time, cost, nuclear medicine availability, invasiveness and also the lack of image resolution (Abbaci et al., 2019, Burnier et al., 2017). One guideline recommends the use of LSG for the functional assessment of lymphoedema pre-treatment, while also recognising its limitations (Lee et al., 2015). Another guideline deemed LSG and lymphography useful in full assessment of the lymphatic system impairment (Levenhagen et al., 2017). The potential role of ICG lymphography over LSG for lymphoedema diagnosis has also been reviewed (Abbaci et al., 2019, Burnier et al., 2017).

Overall, the evidence appears to suggest that the best method for lymphoedema diagnosis depends on various factors. Firstly, different methods are useful for a particular ISL stage of lymphoedema. ICG lymphography (Abbaci et al., 2019, Burnier et al., 2017), bio impedance analysis (Hidding et al., 2016, Shah et al., 2016a, DiSipio et al., 2013a, Levenhagen et al., 2017), tissue dielectric constant (Hidding et al., 2016), optoelectronic perometry (Shah et al., 2016a, Levenhagen et al., 2017) and DEXA (Shah et al., 2016a) have been noted as useful for the diagnosis of early/subclinical lymphoedema. In the moderate/late stages of lymphoedema, other diagnostic methods have been recommended such as: circumferential tape measurement (Levenhagen et al., 2017), water displacement (Levenhagen et al., 2017), perometry/volumetry (Levenhagen et al., 2017, Hidding et al., 2016), ultrasound (Levenhagen et al., 2017) and tonometry (Hidding et al., 2016).

Secondly, the location and the cause/type of lymphoedema can impact what diagnostic tool should be used. Approximately half of the relevant evidence was that of diagnostic methods for upper limb/breast cancer-related lymphoedema (Forte et al., 2020, Abbaci et al., 2019, Sierla et al., 2018, Shah et al., 2016a, DiSipio et al., 2013a, McLaughlin et al., 2017a, McLaughlin et al., 2017c, Levenhagen et al., 2017) but there was inconclusive evidence to determine the best method for diagnosis. One systematic review summarised the use of diagnostic methods in both upper and lower limbs and suggested the use of water volumetry and tape measurement for the assessment of upper limb lymphoedema and calculated good inter-rater ICCs for Bioimpedance Spectroscopy (BIS) use in the lower limbs, but, noted that more research is needed with regards to assessment of midline and lower limb lymphoedema (Hidding et al., 2016).

In summary, there does not appear to be one gold standard method for lymphoedema diagnosis. Each diagnostic method has advantages and disadvantages, along with its own applicability which depends on various different factors.

Recommendations

GQ22.1 Lymphoedema should be diagnosed using subjective and objective means via a standardised assessment template. Refer to [appendix II](#) for assessment templates or hse.ie/lymphoedema for HSE staff, or via www.lnni.org for HSC.

Evidence Grade: D

Strength of recommendation: Strong

GQ22.2 There are multiple ways of measuring lymphoedema and the choice of measurement should be based on accessibility of equipment, appropriateness for the patient with lymphoedema, and site of lymphoedema.

Evidence Grade: C

Strength of recommendation: Strong

GQ22.3 All HCPs should be aware of lymphoedema signs and symptoms related risk factors. HCPs should ensure appropriate and timely onward referral in cases of suspected lymphoedema. Refer to [education section](#) for further details on lymphoedema education for HCPs.

Evidence Grade: D

Strength of recommendation: Strong

GQ23: What subjective outcome measures are recommended in the assessment of lymphoedema?

Evidence Summary

Two measures were recommended by the Breast Cancer EDGE Task Force members to assess the quality of life and function in patients with BCRL - The Functional Assessment of Cancer Therapy – Breast (FACT-B+4) and Disability of Arm, Shoulder and Hand (DASH). These questionnaires came highly recommended for use in patients with BCRL due to their psychometric properties and clinical utility.

Choice of suitable outcome measures depend on multiple different factors (O'Donnell et al., 2020). The CREST CPG recommended that clinical history and physical examination with characteristic findings should establish the diagnosis in most cases. Currently, the combination of outcomes required to capture meaningful change in a lymphoedematous limb is unclear (Sierla et al., 2018). Agreement was not evident for what outcome measures were necessary to ensure all relevant changes are captured. The necessary or core outcome set to demonstrate clinically relevant change in lymphoedema remains unclear. Consensus on a core outcome set with standardised assessment protocols and reporting, and investigation into empirical minimum important differences (MID) is needed.

See below for a comprehensive list of patient report outcome measures (PROMs). See [appendix II](#) for subjective self-assessment and outcome measures for genital lymphoedema.

Published and validated lymphoedema PROMs (Gabe-Walters and Thomas, 2021)

Outcome Measure	Body Part
Lymph-ICF UL (De Vrieze et al, 2019) and Lymph-ICF LL (Devoogdt et al, 2014)	Upper and lower limb
LLIS (Weiss and Daniel, 2018)	Lymphoedema in any extremity
LyQLI (Klernäs et al, 2015) and Swedish version (SLQOLI) (Klernäs et al, 2010)	Upper/lower limb and genital lymphoedema
LFSQQ (Thomas et al, 2014)	Filarial lymphoedema
PBI-L (Blome et al, 2014)	Lymphoedema in any extremity
LYMQOL (Keeley et al, 2010)	Upper and lower limb
ULL QoI (Williams et al, 2018)	Upper limb (BCRL)
ULL 27 (Launois et al, 2002)	Upper limb (BCRL)
WCLS (Mirolo et al, 1995)	Cancer-related PROMs

Recommendations

GQ23.1 There are validated outcome measures which should be used to assess subjective symptoms of lymphoedema and response to treatment and should include levels of pain, fatigue and function.

Evidence Grade: C

Strength of recommendation: Strong

GQ23.2 The subjective outcome measure used should be based on lymphoedema aetiology and patient-specific factors.

Evidence Grade: C

Strength of recommendation: Strong

GQ24: What objective outcome measures should be used in the assessment of lymphoedema?

Evidence Summary

The choice of suitable objective measures to use on patients with lymphoedema depends on different factors (O'Donnell et al., 2020) and should take place at initial assessment and at regular intervals afterwards to assess response to treatment and impact of other clinical changes.

Methods of objectively assessing lymphoedema include:

- Clinical history and physical exam
- Circumferential measurements
- Perometry
- Water displacement
- Bioimpedance spectroscopy (BIS)
- Tissue Dielectric and Dielectric Constant (TDC)
- Imaging studies (MRI, CT scan, and duplex ultrasound) may assist
- Photography can be a useful outcome measure with appropriate consent and data security
- Emerging technologies (e.g., 3D Cameras)

Currently, the combination of outcomes required to capture meaningful change in a lymphoedematous limb is unclear (Sierla et al., 2018). Agreement was not evident for which outcome measures were necessary to ensure all relevant changes are captured. The necessary or core outcome set to demonstrate clinically relevant change in lymphoedema remains unclear. Consensus on a core outcome set with standardised assessment protocols and reporting, and investigation into empirically based minimum important differences (MID), is needed.

Recommendations

GQ24.1 There are validated outcome measures which should be used to assess objective signs of lymphoedema. See [appendix II](#) for objective outcome measure templates.

Evidence Grade: C

Strength of recommendation: Strong

GQ24.2 The outcome measure used should be based on lymphoedema aetiology, patient-specific factors, and accessibility of equipment.

Evidence Grade: C

Strength of recommendation: Strong

GQ25: What tools can measure sub-clinical lymphoedema?

Evidence Summary

Numerous reviews and guidelines have reported that bioimpedance has the ability to detect subclinical lymphoedema (Asklöf et al., 2018, Shah et al., 2016a, He et al., 2020, Shah et al., 2016b, Wanchai et al., 2016, Paskett et al., 2012, Shah and Vicini, 2011, International Society Of Lymphology, 2016, McLaughlin et al., 2017c, Levenhagen et al., 2017). Other tools have also been noted to detect subclinical lymphoedema including perometry (Shah et al., 2016a, McLaughlin et al., 2017c), Tissue Dielectric Constant values (McLaughlin et al., 2017c, International Society Of Lymphology, 2016), DEXA (Shah et al., 2016a, Shah and Vicini, 2011), ultrasound (McLaughlin et al., 2017c), indocyanine green (ICG) lymphography (He et al., 2020, Yamamoto et al., 2011a) and tonometry (He et al., 2020).

Subclinical lymphoedema is not appropriately measured by older/traditional methods including circumferential measurements, water displacement and self-assessment surveys (Shah et al., 2016b, Shah and Vicini, 2011, Brandon Dixon and Weiler, 2015). There is a lack of published reviews focused solely on bioimpedance in lymphoedema (Asklöf et al., 2018, Seward et al., 2016, Shah et al., 2016b).

In general, the evidence deems bioimpedance as a reliable and useful method with increased sensitivity for lymphoedema diagnosis (Asklöf et al., 2018, Sierla et al., 2018, Hidding et al., 2016, Shah et al., 2016a, DiSipio et al., 2013a, Shah et al., 2016b, Lee et al., 2015), especially when compared to older/traditional methods (Shah et al., 2016a, Shah et al., 2016b, Shah and Vicini, 2011). However, varying rates of sensitivity and specificity have been recorded (Seward et al., 2016) and only one study allocated an intra-class correlation coefficient to bioimpedance (Hidding et al., 2016). Much of the evidence supports a role for bioimpedance in the early diagnosis of lymphoedema (Cohn et al., 2017a, Hidding et al., 2016, Shah et al., 2016a, Kayiran et al., 2017, Shah et al., 2016b, Brandon Dixon and Weiler, 2015) while highlighting increased false negative rates in the assessment of advanced lymphoedema or lymphoedema \geq stage 2 (Asklöf et al., 2018, Cohn et al., 2017a, Kayiran et al., 2017). Published guidelines are also in agreement (International Society Of Lymphology, 2016, McLaughlin et al., 2017c, Lee et al., 2015, New South Wales Agency for Clinical Innovation, 2018).

One review was in disagreement with the consensus of this evidence and regarded bioimpedance as an accurate assessment tool for established lymphoedema but stated that it had not been validated for early diagnosis, due to the possibility of false positive results (Seward et al., 2016). Hidding et al. noted good reliability of bioimpedance for both lower and upper extremities but found that the majority of studies examined bioimpedance use in the upper extremities and called for further investigation of bioimpedance in the lower extremities (Hidding et al., 2016).

Recent evidence has called into question the use of Bioimpedance Spectroscopy (BIS) as a method of diagnosing subclinical lymphoedema, with a 2020 prospective study (Bundred et al., 2020) reporting relative arm volume increase to be a superior method of diagnosing lymphoedema when compared with BIS. However, later studies found bioimpedance to be reliable in assessment of lower limb lymphoedema (Asklöf et al., 2018, Cohn et al., 2017a). One review found that bioimpedance was useful for unilateral limb lymphoedema (Cohn et al., 2017a), while another review stated that it was not suitable for bilateral limb assessment (Fu, 2014). In summary, bioimpedance is a reliable method for subclinical lymphoedema diagnosis. Combination of bioimpedance with other diagnostic methods should further increase the sensitivity for early detection (Fu, 2014).

One review found that numerous imaging tools have not been successful in detecting subclinical lymphoedema but suggested a potential future role for Near-infrared fluorescent lymphatic imaging (Brandon Dixon and Weiler, 2015, Wigg and Cooper, 2017).

Recommendation

GQ25.1 Assessment of early fluid changes / subclinical lymphoedema can be accomplished using bioimpedance spectroscopy (BIS) or Tissue Dielectric Constant analysis (TDC).

Evidence Grade: B

Strength of recommendation: Strong

GQ26: What is the accuracy of circumferential measurements in lymphoedema measurement?

Evidence Summary

There have been numerous systematic and literature reviews published to address the use of circumferential measurements in the diagnosis and assessment of lymphoedema. Circumferential measurements are reported as the most frequently used method for lymphoedema diagnosis by assessment of limb volume (Bernas, 2013, Cavezzi et al., 2010, DiSipio et al., 2013a, Johnson et al., 2014, Murdaca et al., 2012, Shah et al., 2016a, Sierla et al., 2018, Wanchai et al., 2016, Brandon Dixon and Weiler, 2015, International Society Of Lymphology, 2016). Guidelines have recommended tape measurement for lymphoedema diagnosis in the limbs provided stringent measurements are taken along multiple anatomical points using a non-stretch tape (Damstra et al., 2017, Lee et al., 2015, Levenhagen et al., 2017, McLaughlin et al., 2017c).

However, both the published guidelines and literature have also noted significant variability and limitations associated with this method. Different mathematical formulas used for circumference to volume conversion, various intervals used for measurement, position of the patient during assessment and requirement for sufficient training have been highlighted as sources of variability (Cavezzi et al., 2010, Cohn et al., 2017a, Cormier et al., 2010, Deng et al., 2015, Hidding et al., 2016, Sayegh et al., 2017, Shah et al., 2016a, Shah and Vicini, 2011, Sierla et al., 2018, Wanchai et al., 2016). Nonetheless, good to excellent reliability has been reported for circumferential measurements (Cohn et al., 2017a, DiSipio et al., 2013a, Hidding et al., 2016, Levenhagen et al., 2017, Perdomo et al., 2014b, Sayegh et al., 2017). One systematic review highlighted varying sensitivity and specificity levels depending on the criteria applied (Hidding et al., 2016). Different cut-offs of change in volume (%) have been recommended by various groups.

Generally, a 10% difference qualifies as a diagnosis of lymphoedema (Hidding et al., 2016, Borman, 2018, Shah and Vicini, 2011) but cut-offs between 5% to 20% have been used (Shah and Vicini, 2011). A difference of > 2 cm in circumference and/or > 200 mL volume difference are common criteria used to diagnose lymphoedema (Borman, 2018, Kayiran et al., 2017, Wanchai et al., 2016, Murdaca et al., 2012, Shah and Vicini, 2011). However, a guideline published in 2017 did not recommend that this measurement of > 2 cm be used for diagnosis due to inaccuracy and a limb volume difference of < 200 mL does not rule out lymphoedema (Levenhagen et al., 2017, Johnson et al., 2014). Furthermore, lymphoedema may be present with circumferential differences < 2 cm (Hidding et al., 2016). Therefore, circumferential measurements have low sensitivity in the assessment/diagnosis of subclinical lymphoedema (DiSipio et al., 2013a, Levenhagen et al., 2017, Shah et al., 2016a). However, contradictory results were published in a different systematic review, the Breast Cancer EDGE Task Force recommends circumferential measurements (along with other tools) for early detection of BCRL (Perdomo et al., 2014b)

With regards to the conversion of circumferential measurements to limb volumes, the literature suggests using a truncated cone (frustum) formula (Borman, 2018, Cohn et al., 2017a, Johnson et al., 2014, Paskett et al., 2012, Perdomo et al., 2014b, International Society Of Lymphology, 2016). These formulas are preferred as they are a closer representative of the shape of the limb than cylindrical formulas, which have shown to overestimate the limb volume (Cohn et al., 2017a, Johnson et al., 2014, Paskett et al., 2012, Shaitelman et al., 2015). The efficacy of circumferential measurements for lymphoedema assessment may vary depending on the site of the lymphoedema. Lymphoedema of the limbs has been successfully measured via circumferential tape methods (Borman, 2018, Cohn et al., 2017a, Hidding et al., 2016, Levenhagen et al., 2017). A published guideline recommends circumferential measurements for upper limb assessment (Levenhagen et al., 2017); excellent reliability and validity has been reported for such (Hidding et al., 2016). Tape measurements have been used for the assessment of head and neck lymphoedema but issues regarding reference points and lack of valid data have been noted (Deng et al., 2015, Deng et al., 2011, Flores et al., 2015). Hand and foot lymphoedema assessment cannot be done easily with tape measurement (Borman, 2018). There is another method of measuring hand oedema with tape, 'figure of eight', which is not commonly used but may be considered. (Borthwick et al 2013).

While some of the literature suggests replacement of traditional diagnostic methods (i.e. tape measurement) with newer techniques (Flores et al., 2015, McLaughlin et al., 2017c, Sayegh et al., 2017), others suggest that multiple methods of assessment be used; the choice of which depends on feasibility, cost and stage of lymphoedema (DiSipio et al., 2013a, Hidding et al., 2016). However, it is important to note that tools for volume assessment are not interchangeable (Hidding et al., 2016, Levenhagen et al., 2017, Perdomo et al., 2014b).

In summary, circumferential measurement is a cost effective, accessible and useful method for lymphoedema assessment but is time consuming, requires a high level of experience for application and may be subject to variability and reduced reliability (Cavezzi et al., 2010, Cormier et al., 2010, DiSipio et al., 2013a, McLaughlin et al., 2017c, Sayegh et al., 2017, Brandon Dixon and Weiler, 2015)

Recommendations

GQ26.1 Circumferential measurements should be taken as part of the objective assessment of lymphoedema and limb volumes should be calculated.

Evidence Grade: B

Strength of recommendation: Strong

GQ26.2 Limb volume recording is recommended and can be calculated using an electronic tool, an example is the LNNI electronic tool which is available from the LNNI website (www.lnni.org).

Evidence Grade: D

Strength of recommendation: Strong

GQ27: How should lymphoedema be assessed in patients with head and neck cancer?

Evidence Summary

There are no standard diagnostic criteria for head and neck lymphoedema (Cohen et al., 2016a) and there is a lack of a standardised universal measurement tool, with only a few methods available to characterise head and neck lymphoedema (Tyker et al., 2019, Flores et al., 2015, Borman, 2018) and a large variety of tools being utilised (Anand et al., 2018, Tyker et al., 2019).

There is variability amongst the published literature regarding how head and neck lymphoedema should be diagnosed. There are significant obstacles to volumetric measurements in head and neck lymphoedema (Damstra et al., 2017, Hidding et al., 2016). In general, it appears that diagnosis should be made by using a combination of patient reported outcomes, physical examination and assessment tools (Levenhagen et al., 2017, Deng et al., 2015, Flores et al., 2015, Borman, 2018, Deng et al., 2019, Smith and Lewin, 2010). HN-ELAF (Levenhagen et al., 2017, Borman, 2018) and NCI CTCAE (Cohen et al., 2016a) are grading tools that have been recommended while Smith et al. devised a protocol for evaluation of head and neck lymphoedema using tape measurements but also advised that evaluation should include patient reported outcomes and functional assessment (Smith and Lewin, 2010).

The most common method used is tape measurement (Smith and Lewin, 2010, Levenhagen et al., 2017, Flores et al., 2015). A protocol for the MDACC HNL evaluation consists of the required measurement and is available from Smith et al. (Smith and Lewin, 2010). Other tools may have a potential role in the measurement of head and neck lymphoedema and include BIS, Tissue Dielectric Constant (TDC) values, tonometry, imaging modalities and photography (Borman, 2018, Cohen et al., 2016a, Damstra et al., 2017, Deng et al., 2015, Flores et al., 2015, Hidding et al., 2016, Smith and Lewin, 2010).

External and internal head and neck lymphoedema require different methods for diagnosis. Often external lymphoedema is diagnosed by physical examination but standardised tools to quantify the extent of swelling are lacking (Deng et al., 2015, Deng et al., 2019, Flores et al., 2015). The diagnosis of internal head and neck lymphoedema requires more invasive modalities including endoscopy (Deng et al., 2015, Deng et al., 2019, Flores et al., 2015). One study suggests that both internal and external anatomy should be examined due to the unique risk of internal lymphoedema with head and neck cancer (Shaitelman et al., 2015).

It has been suggested that head and neck lymphoedema may be underdiagnosed due to the inaccuracy of the methods used for diagnosis (Deng et al., 2015, Deng et al., 2019) and also due to the prevalence of subclinical or internal lymphoedema (Cohen et al., 2016a). Overall, the optimum method/criteria for diagnosing head and neck lymphoedema remains unclear.

Recommendations

GQ27.1 External head and neck lymphoedema measurement should include subjective assessment, to include screening questions about speech/swallow, and objective assessment of external oedema, tissue dielectric constant and photography. Please see [appendix II.V](#) for a Head and Neck Lymphoedema Assessment Template.

Evidence Grade: D

Strength of recommendation: Strong

GQ27.2 If subjective symptoms highlight swallow and/or speech impairment, internal measurements may be needed in conjunction with speech and language assessment in patients with head and neck cancer.

Evidence Grade: D

Strength of recommendation: Strong



Education Need:

HCPs working in head and neck services should be educated in the identification of at-risk patients and of early lymphoedema development.

GQ28: How can internal lymphoedema be diagnosed in patients with head and neck cancer?

Evidence Summary

Published studies on the topic of internal lymphoedema are generally lacking. No specific recommendations have been made by published guidelines with regard to the detection and/or measurement of internal lymphoedema (IL). It has been reported that both physical and functional examinations are the basis of IL measurement (Anand et al., 2018).

The Patterson Scale was developed in 2007 as a grading system for IL (Patterson et al., 2007). Endoscopy is required for the examination which involves the assessment of 11 named structures (Base of tongue, Posterior pharyngeal wall, Epiglottis, Pharyngoepiglottic folds, Aryepiglottic folds, Interarytenoid space, Cricopharyngeal prominence, Arytenoids, False vocal folds, True vocal folds, Anterior commissure) and 2 named spaces (Valleculae & Pyriform sinus), for which the oedema is graded as either none, mild, moderate or severe (Patterson et al., 2007). The Patterson Scale has been cited in several reviews (Anand et al., 2018, Deng et al., 2015, Deng et al., 2011, Deng et al., 2019) and was deemed the only available clinician-reported grading system by a recent review (Deng et al., 2019). The scale has very good intra-rater reliability and moderate inter-rater reliability (Patterson et al., 2007) but also has limitations including inability to measure certain anatomical sites (Anand et al., 2018, Deng et al., 2015, Patterson et al., 2007) and the associated risks with invasive endoscopy (Flores et al., 2015).

Other grading systems for IL include the RTOG/EORTC system and the LENT-SOMA systems which lack validity and reliability and cannot detect oedema of the pharynx nor the oral cavity (Anand et al., 2018, Deng et al., 2015).

Recommendations

GQ28.1 Clinicians should suspect internal oedema in patients with head and neck cancer who present with symptoms of swallow impairment.

Evidence Grade: D

Strength of recommendation: Strong

GQ28.2 Clinicians should consider referral to speech and language therapy in patients with head and neck cancer with suspected swallowing impairment due to internal oedema. Clinicians should also consider referral to dietetics if there is evidence of weight loss.

Evidence Grade: D

Strength of recommendation: Strong

GQ28.3 Oncology speech and language therapists should consider the presence of internal and external oedema as contributing factors to dysphagia after cancer treatment.

Evidence Grade: D

Strength of recommendation: Strong

GQ28.4 Patients with head and neck cancer should be referred back to the MDT for consideration of further assessment if lymphoedema is not resolving with conservative measures.

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Signs and Symptoms of swallow impairment:

- coughing/choking while eating or drinking
- regurgitating food
- sensation of food being stuck in throat
- persistent drooling
- being unable to chew food properly
- pain/discomfort when swallowing

GQ29: How should genital lymphoedema be diagnosed?

Evidence Summary

Genital lymphoedema is defined as increased volume of the genitals due to impairment of the lymphatic system. Genital lymphoedema may be internal or external. It can affect the scrotum, foreskin, penis, vulva, vagina, clitoris or labia minora and majora. It can also affect adjacent areas including the pubis, adductor region of the thighs and the groin (Noble-Jones et al., 2019). A genital lymphoedema scoring (GLS) system has been designed to assess the severity of symptoms associated with genital lymphoedema (Yamamoto et al., 2016). This assessment tool appears to be well associated with the pathophysiological GL severity staging system. Groin nodal lymphography has been validated as a means of diagnosing genital lymphoedema (Gómez et al., 2012).

Lymphoedema Network Wales has devised self-assessment tools for genital lymphoedema in both female and male patients (See appendix [II.IV.II](#)).

Recommendation

GQ29.1 Patients with suspected and self-reported genital lymphoedema should be requested to use the self-assessment tool for genital lymphoedema and this should be used in conjunction with clinician examination and/or photographic evidence. Please see appendix [II.IV.III](#).

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Signs and Symptoms of swallow impairment:

- Clinicians should routinely ask about genital lymphoedema symptoms in patients with relevant risk factors

GQ30: What is the impact of early detection of lymphoedema on patient outcomes?

Evidence Summary

The majority of research to date supports early diagnosis of lymphoedema due to earlier interventions in disease course, lower incidence of complications and overall superior prognosis (International Society Of Lymphology, 2016, Asklöf et al., 2018, Shah et al., 2016a, Cormier et al., 2010, Grada and Phillips, 2017, Bernas, 2013, Damstra et al., 2017, McLaughlin et al., 2017c, Levenhagen et al., 2017, New South Wales Agency for Clinical Innovation, 2018). Furthermore, studies have shown that treatment is most effective when implemented early in the course of disease, which highlights the importance of highly sensitive diagnostic methods and the self-reporting of symptoms (Fu, 2014, Paskett et al., New South Wales Agency for Clinical Innovation, 2018)).

The importance of BCRL surveillance was reported by two reviews (DiSipio et al., 2013a, Shah et al., 2016a). Shah et al. supported the implementation of early monitoring programmes while DiSipio et al. stated that there was little evidence to support the idea. DiSipio et al. also highlighted that lymphoedema may subside with or without treatment which presents a risk of overtreatment with early detection. However, they concluded that lymphoedema is more likely to be underdiagnosed than over diagnosed. Research would suggest that some groups are at higher risk and a targeted approach would seem pragmatic. Only one review stated that the effect of earlier diagnosis of lymphoedema was unclear, as the same recommendations are made for all patients at risk (Hidding et al., 2016). A recent study (n = 488) found that limb volume and circumference increased gradually 36 months post-treatment for breast cancer, whereas subjective reports of symptoms of lymphoedema were much lower, underpinning the importance of early prospective surveillance in these patients (Armer et al., 2019a).

Recommendation

GQ30.1 Early intervention and risk reduction strategies can reduce the impact of lymphoedema and reduce progression so patients should be diagnosed and referred to services as soon as possible.

Evidence Grade: C

Strength of recommendation: Strong

GQ31: What is the role of biological markers in the diagnosis of lymphoedema?

Evidence Summary

A review of biological markers in the clinical management of BCRL addressed this question (Invernizzi et al., 2020). The study of the genetics underpinning BCRL has resulted in a variety of insights which pave the way for the future of precision medicine in BCRL. There have been a number of gene variations for cytokines identified in BCRL patients. These variants include those of pro-inflammatory cytokines (e.g. IL-1, IL-2, NFKB-2) and anti-inflammatory cytokines (e.g. IL-4, IL-10). A selection of single nucleotide polymorphisms (SNPs) in NFKB-2, IL-10, and IL-4 have been shown to be significantly related to the development of unilateral arm swelling (Invernizzi et al., 2020). Other studies have shown the ratio of LTB4/HA may be a useful index to predict development of lymphoedema in BCRL patients (Hadizadeh et al., 2018).

There has been a recent increase in research on the molecular mechanisms controlling lymphangiogenesis and its associated therapeutic potential in the management of lymphoedema. In patients with BCRL several germline alterations in the genes implicated in lymphangiogenesis have been identified, suggesting a possible role for individual predisposition to the development of lymphoedema following breast cancer treatment. The genes identified include lymphocyte cytosolic protein 2 (LCP2), spleen associated tyrosine kinase (SYK), endothelial cell adhesion proteins interleukins, and K-channel genes (Invernizzi et al., 2020). IL-10 is thought to influence active transcription factor binding sites involved in lymphangiogenesis. IL-4, a pleiotropic cytokine produced by CD4+ T-cells with an important role in B-cell immune response modulation, has been shown to display variation in BCRL patients. Dysfunction in this pathway has been proposed as a key factor in the development of lymphoedematous tissues, such as fibrosis, adipose deposition, and lymphatic dysfunction. Cyclooxygenase 2 (COX 2) and prostaglandin 2 (PGE2), the latter of which acts on the EP4 receptor, have also been identified as overexpressed in breast cancer and potentially have a role in lymphangiogenesis. These findings may provide a future for BCRL risk stratification and pre-surgery risk assessment. Future prospective clinical studies should investigate whether NFKB2, IL-10, IL-4, or EP-4 can be employed as circulating biomarkers for lymphoedema development.

In addition, administration of lymphatic growth factors or related molecules may provide potential treatments to target lymphatic vessels in patients with lymphoedema (Saito et al., 2013). Further studies are required to design targeted therapies directed to improve lymphatic regeneration and function while concurrently attempting to modulate inflammatory pathways.

Recommendation

GQ31.1 While significant progress is being made clarifying the correlation between clinical and biological aspects of primary and secondary lymphoedema, further research is required.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

Clarifying the correlation between clinical and biological aspects of primary and secondary lymphoedema

GQ32: How should lymphoedema be staged?

Evidence Summary

The clinical staging of lymphoedema has been defined by the ISL (International Society Of Lymphology, 2016). This classification system consists of 4 specific stages of lymphoedema, where Stage 0 represents subclinical lymphoedema whereas Stages I to III denote overt oedema.

- Stage 0: Latent/subclinical lymphoedema with impaired lymph transport, subtle changes, swelling not yet evident
- Stage I: Accumulation of protein-rich fluid which subsides with limb elevation and may cause pitting
- Stage II: Limb elevation alone rarely reduces swelling, pitting may not be apparent due to the development of subcutaneous fat and fibrosis
- Stage III: Lymphostatic elephantiasis with pitting absent due to progressive development of fat and fibrosis, trophic skin changes seen and warty overgrowths develop

ISL note that the 4 stages are not static; more than one stage may be prevalent in a lymphoedematous limb, which may suggest issues in different lymphatic areas. Stage 2 can be broken down to early and late. This staging system is widely used for the classification of lymphoedema but the ISL also highlight that these stages only relate to the physical characteristics of lymphoedema and suggest that future advances in staging will rely on the incorporation of imaging modalities along with the combination of phenotypic, genotypic, immunological & physical characteristics (International Society Of Lymphology, 2016).

A separate imaging-based staging system was established by Yamamoto et al. in 2011 (Yamamoto et al., 2011b). Dermal backflow staging can be determined using indocyanine green lymphography. This severity staging system is qualitatively divided into 4 different patterns (linear, splash, stardust and diffuse) depending on the ICG lymphography findings as lymphoedema progresses. Furthermore, they noted that these staging patterns correlated to clinical staging.

Recommendation

GQ32.1 Lymphoedema should be staged using the ISL lymphoedema staging classification system.

Evidence Grade: D

Strength of recommendation: Strong

GQ33: When should new patients be referred to a lymphoedema service?

Evidence Summary

The guidelines recommend the following patients with lymphoedema be referred to specialist services without delay: breast cancer patients ((NICE), 2018, McLaughlin et al., 2017c), head and neck cancer patients (Innovation, 2018), oncology patients (Denlinger et al., 2018), patients for whom the diagnosis is unclear or with unknown swelling (Damstra et al., 2017, Lymphology, 2016), paediatric patients (Damstra et al., 2017, NSW Agency for Clinical Innovation, 2018) and patients presenting with cellulitis (New South Wales Agency for Clinical Innovation, 2018). Differences in the levels of specialist services remain unclear and access to such services vary depending on the health system. For example, the Australian Guideline recommends a different level of specialist service depending on the type of lymphoedema (New South Wales Agency for Clinical Innovation, 2018).

Referral to specialist services may be required for certain treatment regimens. Referral may be required if a treatment programme has been unsuccessful or if intensive treatment is needed (Damstra et al., 2017). Two guidelines recommend that patients be referred to a specialist service for treatment, if available, for compression garments, manual lymphatic drainage and resistance training (Cohen et al., 2016a, Denlinger et al., 2018). A review published in 2018 noted that complete decongestive therapy and compression garments should be provided and prescribed by lymphoedema specialists (Borman, 2018).

Recommendations

GQ33.1 Patients should remain with their primary health care provider unless they develop evidence of lymphoedema requiring onwards referral to a lymphoedema service. Evidence of lymphoedema requiring onward referral includes:

- a volume change $\geq 10\%$
- a BIS or TDC increase outside the normal reference range
- Complex presentations including head and neck, genital, and midline lymphoedema

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Signs and symptoms of Lymphoedema:

- Persistent swelling
- Sensation of heaviness/tightness
- Restricted range of motion
- Aching or discomfort
- Recurring infections
- Hardening and thickening of the skin (fibrosis)

Please see appendix I.I for further advice on the adult lymphoedema care pathway.

GQ33.2 Patients with complex palliative care needs should be referred to the specialist palliative team with shared care with the lymphoedema services where possible. See [palliative care section](#) for comprehensive guidance.

Evidence Grade: D

Strength of recommendation: Strong

GQ34: What percentage volume change requires a specialist lymphoedema service?

Evidence Summary

Owing to the heterogeneity of methods used to diagnose lymphoedema (Kayiran et al., 2017) there is currently no international consensus regarding the volume change requiring onward referral to a specialist. Simple non-complex lymphoedema should be managed by HCPs with appropriate education (not lymphoedema specialists).

This does not apply to oncology patients who should follow the oncology surveillance and screening pathway.

This guideline development group reached consensus to endorse the Dutch Lymphoedema Guideline which recommends a cut-off of 10% volume change or complex presentation requiring specialist lymphoedema management.

See the adult and oncology pathways ([appendix I.I](#) and [I.IV](#))

Recommendations

GQ34.1 Oncology patients with a subclinical change noted by BIS or an identified relative volume increase of $< 5\%$ identified via screening, should be managed and monitored as part of a screening/surveillance pathway. Refer to surveillance section GQ37.

Evidence Grade: D

Strength of recommendation: Strong

GQ34.2 Any patient with an identified volume change of 5% to 10% (without midline or other complexity) should receive treatment from a non-specialist with education in managing non-complex lymphoedema, including compression, skin care and exercise with lymphoedema services support as needed.

Evidence Grade: D

Strength of recommendation: Strong

GQ34.3 Patients with an identified volume change of $> 10\%$ should be referred to a lymphoedema specialist.

Evidence Grade: D

Strength of recommendation: Strong

GQ34.4 Complex presentations, irrespective of volume change, should be referred to a lymphoedema specialist.

Evidence Grade: D

Strength of recommendation: Strong

GQ35: How should new patients be prioritised once referred to a lymphoedema service?

Evidence Summary

As there is a lack of evidence examining prioritisation of care for lymphoedema patients, a round table discussion led to a consensus statement based on expert opinion. The opinion of the GDG is that patients should be prioritised in the following manner:

Priority 1

- Risk of imminent hospital admission if not seen e.g. cellulitis, lymphoedema with skin breakdown/ lymphorrhoea
- Acute oncology-related lymphoedema
- Palliative lymphoedema affecting quality of life

Priority 2

- Acute deterioration of oedema/symptoms
- Patients with suspected primary lymphoedema

Priority 3

- Stable patients who have been re-referred to services

Refer to [Paediatric section](#): patients will require more regular review for bespoke needs and thus should be prioritised.

Recommendations

GQ35.1 Patients with lymphoedema who may eventually be at risk of hospitalisation or cases of lymphoedema that progressively deteriorate should be considered first priority for assessment.

Evidence Grade: D

Strength of recommendation: Strong

GQ35.2 Patients with lymphoedema and evidence of active deterioration of oedema/symptoms should be seen as a second priority for assessment.

Evidence Grade: D

Strength of recommendation: Strong

GQ35.3 HCPs should ensure urgent assessment by lymphoedema clinicians for oncology (non-screening/surveillance) patients, paediatric patients or patients presenting with recurrent cellulitis.

Evidence Grade: D

Strength of recommendation: Strong

GQ35.4 Patients with lymphoedema who present to services systemically unwell should seek urgent medical attention.

Evidence Grade: D

Strength of recommendation: Strong

GQ36: How should patients be prioritised for a planned review within a lymphoedema service?

Evidence Summary

As there is a lack of evidence examining prioritisation of care for lymphoedema patients, a round table discussion led to a consensus statement based on expert. The opinion of the GDG is that patients should be prioritised in the following manner:

Priority 1

- Risk of imminent hospital admission if not seen e.g. cellulitis, lymphoedema with skin breakdown/ lymphorrhoea
- Acute oncology-related lymphoedema
- Early-onset oncology-related lymphoedema

Priority 2

- Acute deterioration of oedema/symptoms
- Previously stable patients needing compression modification
- New patients who are not improving with initial advice/compression

Priority 3

- Stable review patients

Refer to [Paediatric section](#): patients will require more regular review for bespoke needs and thus should be prioritised.

Recommendations

GQ36.1 Patients with lymphoedema who may eventually be at risk of hospitalisation or cases of lymphoedema that progressively deteriorate should be considered first priority for planned review.

Evidence Grade: D

Strength of recommendation: Strong

GQ36.2 Patients with lymphoedema and evidence of active deterioration of oedema/symptoms should be seen as a second priority for planned review.

Evidence Grade: D

Strength of recommendation: Strong

GQ36.3 Stable review patients with lymphoedema should be seen as a third priority for review.

Evidence Grade: D

Strength of recommendation: Strong

GQ36.4 Patients with lymphoedema who present systemically unwell should seek urgent medical attention.

Evidence Grade: D

Strength of recommendation: Strong

1.3 Treatment

GQ37: Is there any evidence to support specific models of care in improving patient outcomes?

Evidence Summary

Chronic Conditions
 A comprehensive review of international evidence (Flanagan et al., 2017) assessing the effectiveness of integrated care interventions in improving quality of life in patients with chronic conditions addressed this question. This review assessed 41 articles and categorised interventions into:

- Case management
- Chronic care model
- Discharge management
- Multidisciplinary teams
- Complex interventions
- Primary vs. secondary care follow-up
- Self-management

This review demonstrated the varied effectiveness of integrated care interventions. In general, interventions were more effective in condition-specific quality of life, in comparison to overall quality of life. Overall, ‘chronic care model’ and ‘case management’ interventions led to positive quality of life improvements and these tended to increase in effectiveness with increasing numbers of elements. ‘Self-management’ and ‘multidisciplinary teams’ were more mixed in terms of their improvement in quality of life in patients with chronic conditions.

Chronic Care Model
 Chronic care models are based on active participation by patients in the management of their health condition, self-efficacy and empowerment of the patient combined with a more hands-off approach by health care workers, who function in the background as a network of professionals sharing data and evidence-based management strategies. Using this model, the patient is at the centre of the care pathway and their care is arranged according to a network-of-care pyramid. At the top of the care pyramid is the highly-specialised expert care, provided to a subset of patients and at the bottom of the pyramid is the local- or home-based care offered to all patients (Pines, 2015). The effectiveness of this model of care has been noted in other chronic conditions such as atrial fibrillation (Hendriks et al., 2015) and diabetes (Barletta et al., 2017).

The Dutch Guidelines on lymphoedema management are also based on the Chronic Care Model, using the International Classification of Functioning (ICF) framework approach to create a holistic and functional pathway of care. The Dutch Guidelines propose an interdisciplinary approach to lymphoedema care using a “continuum of care from prevention to initial treatment phase, maintenance phase, and follow-up. An integrated treatment programme can be conservative or surgical. A therapeutic programme depends on stage and origin of lymphoedema; International Classification of Functioning, Disability, and Health-based limitations; needs of the patient; ability to perform self-management; and ability to reduce patient-related risk factors, such as overweight and lack of exercise”. The authors assert that this approach centres on patient awareness, early diagnosis and risk stratifying practices and they contend that early recognition facilitates early treatment which in turn reduces patient morbidity and risk of lymphoedema complications.

A literature-scoping review identified that participants experienced the programmes as beneficial according to less symptom distress, greater awareness of their own health, improved self-management strategies, peer support and learning (Stenberg et al., 2016). Barlett (1995) showed that for every dollar spent on patient education, four are saved. Kirsner (2018) produced a meta-analysis of 5 small studies, and suggested that exercise offers an additional benefit in patients with leg ulcers (61% healed at 12 weeks in comparison to 41%).

A Cochrane systematic review (Lane et al., 2017) concluded that there was high-quality evidence showing that exercise programmes provided important benefit compared with placebo or usual care in improving both pain-free and maximum walking distance in people with leg pain from intermittent claudication who were considered to be fit for exercise intervention. Literature-scoping review examined telephone consultations for people with chronic conditions. 47 articles were reviews and found this model can improve health behaviour, self-efficacy, and health status. The review found that telephone-based coaching can enhance the management of chronic disease, especially for vulnerable groups (Dennis et al., 2013). For every £1.00 spent on lymphoedema treatments that limit swelling and prevent damage and infection, the NHS saves an estimated £100 in reduced hospital admissions (NCAT, 2013).

Examples of appraised Lymphoedema Chronic Condition Models

- The Healthy Legs Project, Southern Health and Social Care Trust
- The St Oswald’s Lymphoedema Service, Newcastle Upon Tyne
- The Head and Neck Cancer Lymphoedema Model at Guy’s and St Thomas’ Hospital, NHS Foundation Trust, London

A typical four week programme would include:

Week 1	Week 2	Week 3	Week 4
Causes of swelling Signs and symptoms Complications associated with swelling	Self-management Skin care Positioning Physical activity	Principles of healthy eating Food labelling Weight control Onward referral	Role of compression garments Donning/doffing aids General care advice
EXERCISES	EXERCISES	EXERCISES	EXERCISES

Prospective Surveillance Model for Oncology
 Several lymphoedema guidelines recommend a comprehensive prospective surveillance program for all patients at risk of lymphoedema. International Society of Lymphology (2016), the National Lymphoedema Network (2011), the American Physical Therapy Association (Brunelle et al., 2018), the Australasian Lymphology Association (Boyages et al., 2020) and the American Society of Breast Surgeons (McLaughlin et al., 2017c), all recommend a prospective surveillance model of care be employed for patients at risk of lymphoedema.

A single-centre study (Yang et al., 2016) assessed the outcomes of a surveillance programme for lymphoedema management compared to standard care. In total 707 patients who underwent axillary lymph node dissection were included in the study. Overall 5-year incidence of lymphoedema was 6.4% in the surveillance group and 15.1% in the standard care group. The authors claim surveillance improves the rate of lymphoedema prevention compared to clinical assessment alone.

Based on their study they recommend the first visit to the clinic should be within 1 month post-op and follow-up visits should be less than 3 months apart in the in the first year. It is recommended that for surveillance purposes, an initial pre-op measurement should be followed by regular measurements for 3–5 years.

A review of the literature on the role of early detection of prospective surveillance care pathway concluded that current evidence supports the development of surveillance programs aimed at the early detection and management of BCRL (Shah et al., 2016a). Currently available research findings do not standardize early interventions or provide long-term follow-up to allow one recommended treatment pathway over another.

Lymphoedema Surgery Pathways

The Australian Lymphoedema Education Research and Treatment (ALERT) programme originated as an advanced clinic for patients considering surgery for lymphoedema (Boyages et al., 2020). The Australasian Lymphology Association endorses the need for all patients treated for breast cancer to have access to:

- 1) an educational programme informing them about lymphoedema
- 2) a prospective monitoring programme for changes indicative of developing swelling, particularly for those at higher risk of developing breast cancer-related lymphoedema”

Recommendations

GQ37.1 Lymphoedema is a chronic condition and requires the Chronic Care Model. The role of the expert patient is central, and must be addressed through education and empowerment to ensure responsibility for their care, and long term success.

Evidence Grade: D

Strength of recommendation: Strong

GQ37.2 A lymphoedema service should be developed with funded links to multi-professional services required for diagnosis and management. Services should include allied health professionals, health and social care professionals, tissue viability, dermatology, vascular services, psychology, bariatric services, oncology, paediatrics and associated surgical disciplines.

Evidence Grade: D

Strength of recommendation: Strong

GQ37.3 Cancer Prehabilitation is being developed as part of new cancer pathways, and should be utilised to provide pre-cancer management awareness of lymphoedema regarding risk reduction, and surveillance.

Evidence Grade: D

Strength of recommendation: Strong

GQ37.4 The risk of developing lymphoedema should be discussed as part of the medical consent process for any treatment with an identified risk of lymphoedema development.

Evidence Grade: D

Strength of recommendation: Strong

GQ37.5 Other non-cancer risk-reduction strategies may include upskilling those in general practice (e.g. for the successful provision and monitoring of life-long compression) alongside relevant public health messages.

Evidence Grade: D

Strength of recommendation: Strong

GQ37.6 Risk awareness education for those working with the at-risk populations should be built into relevant undergraduate and post graduate academic programmes for the MDT.

Evidence Grade: D

Strength of recommendation: Strong



Education Need:

Risk awareness education for those working with populations at risk of lymphoedema should be built into relevant undergraduate and post graduate academic programmes for the MDT.

GQ38: Is there any evidence to identify the most appropriate clinical settings in which initial lymphoedema treatment and follow up should take place?

Evidence Summary

There was no evidence available to answer this question. The Chronic Care Model supports delivery of services in an outpatient setting by encouraging the service to fit within the normal lifestyle of the patient. This service would ideally be delivered in a facility with access to MDT and specific medical teams as needed.

Recommendations

GQ38.1 The Chronic Care Model supports delivery of services in an outpatient setting by encouraging the service to fit within the normal lifestyle of the patient. This service would ideally be delivered in a facility with access to MDT (e.g. physiotherapy, tissue viability) and specific medical teams (e.g. dermatology) as needed.

Evidence Grade: D

Strength of recommendation: Strong

GQ38.2 In rare cases where patients may benefit from an acute admission to hospital, a funded bed should be made available.

Evidence Grade: D

Strength of recommendation: Strong

GQ38.3 Commissioning should be provided for inpatient lymphoedema services which are not currently available. Both in-reach and/or outreach services should be developed.

Evidence Grade: D

Strength of recommendation: Strong

GQ38.4 Domiciliary visits may be considered for patients with specific access needs who are unable to attend healthcare appointments, this may include nursing homes depending on local policy. This service should be monitored and further commissioning sought if there is increased demand.

Evidence Grade: D

Strength of recommendation: Strong

GQ38.5 Specific risk assessment should be undertaken before domiciliary visits to ensure the health and safety of the HCP and patient/carer. This may require the use of additional support staff.

Evidence Grade: D

Strength of recommendation: Strong

GQ39: Which criteria would indicate the need to provide CDT/DLT?

Evidence Summary

Complex Decongestive Therapy (CDT) / Decongestive Lymphatic Therapy (DLT) is the internationally recommended current best practice for the treatment of lymphoedema. CDT/DLT consists of two stages, the goal of the first stage or “intensive treatment” stage is to decongest the oedematous limb via clinician-delivered manual lymphatic drainage (MLD) and multi-layer compression bandaging for a duration of at least two weeks. The second stage consists of maintenance carried out by the patient in the form of self-massage and skin care along with daily compression, usually with compression garments.

CDT/DLT in Early Onset BCRL

A 2018 systematic review addressed this question in part (Jefferis et al., 2018). This review assessed 7 studies (5 RCTs and 2 descriptive studies) which assessed the efficacy of CDT in early onset lymphoedema (duration of less than 12 months of BCRL symptoms). The author concluded that there is some evidence that CDT is effective in patients with early-onset BCRL but that they were unable to draw any conclusions regarding the most effective treatment when patients first present. Similarly a review of therapies aimed at BCRL concluded that CDT is recommended for stage 0 and stage I BCRL only (Smile et al., 2018)

The Dutch Guidelines (Damstra and Halk, 2017) recommend that once there is a volumetric increase between 5%-10% in one limb compared to non-affected side (and to pre-op values) an adjusted lymphoedema programme including compression should be introduced (including thorax compression and a bra holder). These patients should have more frequent check-ups and a full lymphoedema programme should be initiated if there has been no improvement. Patients with a volumetric increase in the affected limb > 10% compared to the non-affected side (and to pre-op levels) should be commenced on a full lymphoedema treatment programme. This programme should involve decongestive lymphatic therapy (DLT) and compression.

In cases where patients feel the need to attend private therapists in addition to their public therapists, the GDG feel a collaborative relationship must be maintained between therapists once consent is given by the patient attending both. Any identified risks must be discussed, documented and managed.

Recommendations

GQ39.1 CDT may be considered in patients with a relative volumetric difference of greater than 10% between the affected limb/segment and the unaffected limb/segment, taking into consideration hand dominance and its impact on volume differences.

Evidence Grade: D

Strength of recommendation: Strong

GQ39.2 CDT may be considered in patients with a relative volumetric difference of less than 10% in those with midline swelling, head and neck swelling, digital swelling, or swelling which is complex in shape. These patient groups should be managed by a specialist lymphoedema service.

Evidence Grade: D

Strength of recommendation: Strong

GQ39.3 An adjusted lymphoedema programme including compression, should be introduced in patients with a relative volumetric increase between 5%-9% in one limb compared to the non-affected side. This can be managed by generalist staff if non-complex in shape and not midline.

Evidence Grade: D

Strength of recommendation: Strong

GQ39.4 In patients with bilateral limb swelling, CDT may be considered based on clinical judgment and patient-reported symptoms.

Evidence Grade: D

Strength of recommendation: Strong

GQ39.5 Treatment provided should always be determined by the clinical presentation and clinical judgement. If patients have other symptoms related to lymphoedema such as pain, these should be taken into consideration and treated accordingly.

Evidence Grade: D

Strength of recommendation: Strong

GQ40: Which criteria should be used to determine whether supplemental CDT should be considered?

Evidence Summary

There was no evidence available to answer this question.

Based on expert consensus, a Supplemental Treatment Policy was created which states when supplemental CDT should be provided.

As part of the review process, the therapist must:

- Use the Lymphoedema Service review form with all review patients to be able to assess and discuss concordance with the patient, and in particular the correct use of garments
- Ensure that the current maintenance programme is sufficient and appropriate for the individual and, if required, ensure additional support is provided to enable this practice. If a change in self-management or level of support is required, a further period of supported self-management prior to supplemental CDT may be considered.

Essential criteria for supplemental CDT:

- The patient must be fully concordant with the recommended maintenance programme and, where possible, the patient and/or a carer should routinely practice self-MLLB and SLD

And one of the following criteria:

- The patient has had a recent Acute Inflammatory Episode (AIE), which has resulted in a significant increase in limb volume and/or deterioration of the skin condition and thickening of the tissues
- The patient has had an acute exacerbation of a skin condition/a wound and they can no longer wear their compression garment(s)

The period of supplemental treatment should be no longer than two weeks, or until previous measurements are achieved or plateau, and will include revision sessions of self-MLLB and SLD etc. Patients should bring all their recent and current compression garments with them so that the therapist can assess fit, wear and garment care practices.

For those who require regular supplemental treatment (at least one supplemental treatment period every year), the therapist should consider maintenance CDT instead (i.e. one session each month on an ongoing basis). For example, those with uncontrolled genital/breast swelling, and who are concordant with self-care, should be offered a trial of regular MLD sessions.

The therapist must discuss all individual cases with the Team Lead before offering supplemental or maintenance CDT sessions.

Recommendation

GQ40.1 Lymphoedema services should adopt the lymphoedema supplemental CDT policy (as above) in conjunction with an individualised self-management discussion with all patients.

Evidence Grade: D

Strength of recommendation: Strong

1.4 Skin Care

GQ41: Which skin care practices are recommended in the treatment of Lymphoedema?

Evidence Summary

A review of skin and wound care in lymphoedema addressed this question (Fife et al., 2017). The authors of this paper made the following recommendations regarding skin care in the management of lymphoedema:

Daily hygiene with careful washing

- Soaps are drying, moisturising soap substitutes are recommended as an alternative. Avoidance of skin damage or trauma
- Use of protection from sunburn and avoidance from cuts, insect bites, injections, and overly hot water.
- Use of appropriate shoes for patients with lower extremity lymphoedema and gloves for certain activities involving the affected upper extremities (e.g. gardening).

Daily application of perfume-free emollients

Good nail hygiene

- Keep nails short and trim them often.
- Scrub the underside of nails with soap and water to reduce bacterial and fungal entry points.

A second review of the literature (Jones et al., 2019) lead to the following recommendations by the authors:

- Daily washing and drying of skin with appropriate cleansing products (with particular attention to skin folds)
- Emollient application as a moisturiser
- Active monitoring of both the affected and unaffected regions for any signs of trauma or cellulitis
- Active prevention of skin trauma via use of sun lotion and protective equipment (e.g. gardening gloves) for ADLs

Compression garments should be removed on a daily basis and not used for more than 24 hours consecutively. Anecdotally if garments are left in situ for prolonged periods, the normal shedding of dead skin cells does not happen and the build-up of dead skin cells under compression is likely to add to the problem of hyperkeratosis. If removing garments daily is not possible then the treatment used for dry skin needs to reflect this in terms of ensuring that dead skin cells do not build up causing hyperkeratosis or dry-skin conditions. Where appropriate certain patients should be assessed and managed by podiatry services.

Recommendations

GQ41.1 It is recommended that all patients at risk of lymphoedema or patients with lymphoedema follow a standard skin care protocol. Please see appendix III.I for a standard skin care protocol.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.2 It is recommended that the protocol includes; a thorough examination of the affected area looking for signs of changes in the skin condition (e.g. increased dryness, infection, injury, changes in shape or distribution of oedema), washing with warm water and soap substitute and applying moisturiser/emollients.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.3 It is recommended to use a soap substitute as per patient preference. Soap substitutes are water and oil-based and therefore do not have the same irritant or drying effect as perfumed soaps.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.4 It is recommended to dry between the fingers and toes and skin folds after washing and if this is too difficult then use of a spray cleanser is recommended.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.5 It is recommended to wait 30 minutes after applying moisturiser before attempting to apply a compression garment.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.6 It is recommended that compression garments be removed daily. If this is not possible then extra care should be taken to remove dead skin when washing, before applying moisturiser.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.7 When garments are removed daily, they should be washed as per manufacturer instructions and replaced with a clean, dry garment.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.8 For the removal of unwanted hair it is recommended to use a method that minimises the risk of damaging the skin (i.e. chemical irritation, cuts). The choice of hair removal method should be based on informed patient preference.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.9 It is recommended that good nail hygiene is followed and referral to a podiatrist should be made if required.

Evidence Grade: D

Strength of recommendation: Strong

GQ41.10 It is recommended that patients use moisturiser from a pump or a tube instead of tub-based moisturiser due to the risk of product contamination. If tub-based moisturiser is essential, hygiene measures such as gloves or a spatula should be considered as well as hand washing before use, in keeping with infection prevention and control practices.

Evidence Grade: D

Strength of recommendation: Strong

GQ42: Which types of skin care product should be used for patients with lymphoedema?

Evidence Summary

A review of skin and wound care in lymphoedema addressed this question (Fife et al., 2017). The authors of this paper made the following recommendations regarding skin care in the management of lymphoedema:

Daily application of emollients without perfume

- Emollients are moisturisers which help the epidermis to retain water and diminish water loss (e.g. bath oils). Regular use of ceramide-containing emollients re-establishes the protective lipid layer of the skin, thus preventing water loss.
- These products are available as either lotions or creams.
- Creams are often the best option for dry skin.
- As emollients may damage the elastic component of compression garments, current recommendations include avoiding application immediately before putting on hosiery.

Dermatologic preparations for specific skin problems

- Topical steroids, antifungals, and antimicrobials have been successfully used off-label for the conditions associated with lymphoedema, such as tazarotene gel 0.1% for which there is a case report in the treatment of elephantiasis nostras verrucosa.
- Topical tacrolimus has been suggested off-label for use in severe stasis dermatitis as a possible alternative to topical steroids.

All substances placed on the skin have the potential to penetrate and therefore be cleared by the lymphatic system and so HCPs should consider the effect a compromised lymphatic system might have on this process if absorption is anticipated. It is therefore very important that any excess topical preparation is avoided and indeed removed during the standard skin care regime. Emollient and moisturiser are terms often used synonymously. An emollient is a substance that smooths and softens the skin usually via occlusion. Moisturisers can actively add moisture to skin usually as humectants but their main role is to prevent loss of water from the skin. Humectants are hygroscopic substances which absorb water or retain moisture (e.g. urea, glycerine and sorbitol).

Recommendations

GQ42.1 It is recommended that perfume-free moisturisers/emollients (lotions, ointments or creams) are used as directed by clinicians, in order to protect the skin and prevent dryness. The choice of moisturiser should be based on patient preference, ability and availability.

Evidence Grade: D

Strength of recommendation: Strong

GQ42.2 Creams are recommended for dry skin, however, products containing lanolin should be avoided due to known potential for sensitivity and dermatitis.

Evidence Grade: D

Strength of recommendation: Strong

GQ42.3 It is recommended that excessive moisturising be avoided and any residue from previous treatment should be fully removed during washing before reapplying moisturiser.

Evidence Grade: D

Strength of recommendation: Strong

GQ42.4 HCPs, patients and carers should be aware of the fact that some paraffin-based emollients present a fire hazard particularly if soaked into garments or bandaging, therefore choice of emollient may need to be risk-assessed based on individual risks such as smoking status and use of oxygen therapy.

Evidence Grade: D

Strength of recommendation: Strong

GQ42.5 Patients and carers should be advised to allow sufficient time for moisturiser absorption before applying garments.

Evidence Grade: D

Strength of recommendation: Strong

ALERT:

Some paraffin based emollients present a fire hazard particularly if soaked into garments or bandaging, therefore choice of emollient may need to be risk assessed based on individual risks such as smoking status and oxygen therapy.

GQ43: Which extra skin care measures are needed in the management of skin conditions associated with lymphoedema?

Evidence Summary

The following skin complications may occur in patients with lymphoedema:

- Plantar dermatitis
- Atopic eczema
- Varicose eczema (with or without ulceration)
- Ulceration
- Hyperkeratosis
- Papillomatosis
- Skin fold maceration
- Lymphorrhoea
- Folliculitis
- Fungal infections
- Onychomycosis
- Lymphangiectasia

Patients experience red, itchy skin which can be broken and therefore some degree of exudate can be present. Consideration should be given to potential substances which may be causing the dermatitis and avoidance of exposure to suspected agents. If not improving with these simple measures or if the condition is severe, referral to a GP or dermatologist may be appropriate.

Liners may be required for use underneath garments/gloves. These may also cause increased perspiration and hold moisture to the skin, causing a moist warm environment for microbes to grow. In this case, a silver-impregnated liner may help. Washing detergent residue on clothing can cause sensitivity which is often mistaken for allergic dermatitis. If this is suspected, a localised trial of elimination and reintroduction can identify the offending agent. Allergic dermatitis can result in an increase in oedema due to the inflammatory response.

Recommendations

GQ43.1 It is recommended that for all associated skin conditions that the standard skin care protocol is followed with the following additional recommendations.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.2 It is recommended for dermatitis that potential irritants are eliminated. If no improvement is seen, then referral to the GP is needed for consideration of topical steroids. Owing to their antimicrobial effect, the use of silver liners may be considered in the treatment of dermatitis.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.3 In patients with plantar dermatitis, medicated bandaging should be used.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.4 Patients with lymphoedema who present with evidence of atopic eczema should be referred to primary care or to dermatology.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.5 It is recommended that varicose eczema with ulceration is treated with a multi-disciplinary approach combining lymphoedema management and the treatment of venous leg ulceration.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.6 It is recommended for varicose eczema without ulceration that the standard skin care protocol is followed and if no improvement is seen, then referral to the GP is needed for consideration of topical steroids.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.7 When washing or bathing, it is recommended for hyperkeratosis that moisturisers are added to the water. Exfoliator pads may also be used when washing.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.8 The use of an antimicrobial preparation (e.g. chlorhexidine) may be considered when washing limbs with evidence of papillomatosis and the choice of agent should be based on local antimicrobial guidelines. Patients should be advised, as with all topical preparations, to begin with a patch test of the prescribed agent on a small area of skin to assess for allergic or contact irritant reaction.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.9 Keratolytic preparations such as salicylic acid or creams-containing urea can be used depending on patient tolerance.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.10 It is recommended that skin folds are fully examined every day to allow for early detection and treatment of lymphoedema-associated skin changes.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.11 It is recommended for Lymphorrhoea that a barrier cream is used to protect the skin around the affected area. See the [Lymphorrhoea pathway](#) for further details on management.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.12 It is recommended for folliculitis that the area be swabbed for microbiology assessment and anti-bacterial wash be used. Topical antibiotics can be considered as second-line therapy for refractory folliculitis according to local microbiology guidelines.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.13 It is recommended for fungal infections that scrapings be taken to attempt fungus identification. Often fungal culture is not successful, however the presence of spores in scrapings should be considered diagnostic and be treated with an appropriate antifungal preparation. Antifungal preparations should be removed fully before reapplication. The area should be kept dry and aerated footwear is recommended.

Evidence Grade: D

Strength of recommendation: Strong

GQ43.14 It is recommended for onychomycosis that nail clippings be sent for analysis and that appropriate antifungal preparation be used to treat the area. If no improvement is seen then consider referral to a podiatrist or GP.

Evidence Grade: D

Strength of recommendation: Strong

GQ44: Which types of skin care products should be used in patients with lymphoedema who have allergies to topical agents?

Evidence Summary

There was no evidence available to answer this question however a review of skin care in lymphoedema management (Fife et al. 2017) recommended that: "In cases where clinical trial literature is scant, conflicting, or unclear, a consensus approach is recommended based on expert opinion regarding clinical vignettes. A group of experts should be asked to describe how to manage a particular patient in a primary care setting and address a set of questions about key areas of practice". Therefore the expert opinion of this guideline development group is that patients with lymphoedema with a history of allergy to topical agents should be reviewed by their GP/dermatologist to advise on what is best for that patient in cases where multiple products have been trialled.

Recommendation

GQ44.1 Patients with lymphoedema with a history of allergy to topical agents should be reviewed by their GP/dermatologist to advise on what is best for that patient in cases where multiple products have been trialled.

Evidence Grade: D

Strength of recommendation: Strong

1.5 Compression Therapy

GQ45: Is an assessment of Ankle Brachial Pressure Index (ABPI) necessary prior to providing compression?

Evidence Summary

An international consensus statement addressed this question (Rabe et al., 2020). It is recommended that clinicians check arterial circulation status before compression therapy is initiated. In cases where the foot and/or ankle pulse is weak or not palpable, the Ankle Brachial Pressure Index (ABPI) should be measured and calculated prior to applying compression therapy.

The BLS have previously published a guidance document on vascular assessment, however it is currently under review, with a view to updating current guidance.

Recommendations

GQ45.1 Routine ABPI measurements for patients who present with lymphoedema are not required in the absence of significant cardiovascular risk factors and clinical signs or symptoms of PAD (Peripheral Arterial Disease), provided the vascular status has been thoroughly assessed. If there are concerns in terms of reduced arterial flow, a referral for further vascular assessment and possible intervention should be pursued.

Evidence Grade: D

Strength of recommendation: Strong

GQ45.2 All lymphoedema clinicians should be competent to assess vascular status.

Evidence Grade: D

Strength of recommendation: Strong

GQ46: Are there any contraindications for medical compression?

Evidence Summary

An international consensus statement on the risks and contraindications of medical compression therapy answered this question (Rabe et al., 2020). Medical compression here refers to compression garments and bandaging. Expert consensus is that medical compression therapy is rarely associated with severe adverse events if it is used appropriately and contraindications are considered.

Cardiac Insufficiency

Cardiac insufficiency in itself is not a contraindication to compression therapy. Decompensated cardiac insufficiency is internationally regarded as a contraindication to medical compression and to MLD. In patients with New York Heart Association (NYHA) heart failure grade I or II, compression is possible. While not recommended, in NYHA stages III and IV use of compression may be possible in a limited manner, given there is a strict indication for compression and haemodynamic response is closely monitored.

Peripheral Arterial Disease (PAD)

A recent consensus paper by (Rabe et al., 2020) recommended contraindications for compression treatment are

severe peripheral arterial disease (PAD) with:

- Ankle brachial pressure index (ABPI) < 0.6,
- Ankle pressure < 60 mmHg,
- Toe pressure < 30 mmHg,
- Transcutaneous oxygen pressure < 20 mmHg
- Suspected compression of an existing epifascial arterial bypass
- Severe cardiac insufficiency (New York Heart Association [NYHA] class IV)

With compression bandaging applied, pressure and the material elasticity should be considered. This contraindication does not apply to patients with non-critical leg ischaemia treated with inelastic material applied with low resting pressure. In every patient with impaired perfusion (ABI < 0.9), the clinical effect of the compression stocking on blood supply should be closely monitored. Non-healing skin breaks may develop even with the use of low pressure stockings.

The guideline development group endorse the previous [HSE wound management recommendations](#) on compression therapy.

Recommendation

GQ46.1 The following contraindications for medical compression are recommended:

- Severe peripheral arterial disease (PAD) with an ABPI < 0.6, ankle pressure < 60 mmHg, toe pressure < 30 mmHg, or transcutaneous oxygen pressure < 20 mmHg
- Severe cardiac insufficiency (New York Heart Association [NYHA] class IV)
- Suspected compression of an existing epifascial arterial bypass
- Confirmed allergy to compression material
- Severe diabetic neuropathy with sensory loss or microangiopathy with the risk of skin necrosis (this may not apply to inelastic compression exerting low levels of sustained compression pressure, or modified compression)

Evidence Grade: D

Strength of recommendation: Strong

GQ47: What is the efficacy of compression therapy in the treatment of lymphoedema?

Evidence Summary

Compression is currently recommended as a key component of a combined treatment approach by several international guidelines and expert groups (Gloviczki, 2016, 2016, Damstra and Halk, 2017, McLaughlin et al., 2017a), however only a low level of evidence exists to support its use (Health, 2014). This is due to the lack of trial evidence examining compression therapy in lymphoedema.

The authors of a systematic review of the evidence supporting the use of compression in venous and lymphatic disease concluded that compression is the most important component of DLT for treatment and maintenance (Rabe et al., 2018). Conversely, an earlier systematic review and meta-analysis (Rogan et al., 2016) of available studies concluded that compression garments do not reduce lymphoedema volumes in the acute phase but they prevent additional swelling. Based on expert opinion and taking into account the quality of available evidence and improvements in recent compression technology, this guideline development group supports the use of compression as an integral component of lymphoedema management at all stages.

Recommendations

GQ47.1 There is evidence for the use of compression in all stages of lymphoedema. Compression should be used as part of a combined treatment approach and should not be used in isolation.

Evidence Grade: A

Strength of recommendation: Strong

GQ47.2 When lymphoedema is stable, compression garments or devices are recommended as part of the maintenance treatment.

Evidence Grade: A

Strength of recommendation: Strong

GQ48: What is the optimal level of compression prescription in each stage of lymphoedema?

Evidence Summary

A narrative review of the use of compression in lymphoedema addressed this question (Mosti and Cavezzi, 2019). Upon analysis of the literature the authors of this review call for lymphoedema experts to revise high compression regimens previously proposed. They conclude that pressure in the range of 20–30 mmHg is sufficient in the treatment of arm lymphoedema and 40–50 mmHg is sufficient for the treatment of lower limb lymphoedema.

The International Society of Lymphology recommend the highest level of compression tolerated within the range of 20-60 mmHg (ILS, 2016). The Dutch guidelines (Damstra and Halk, 2017) recommend compression class III (34-46 mmHg) or IV (> 49 mmHg) garments for lower limb lymphoedema and class II (23 - 32 mmHg) and III (34-46 mmHg) for upper limb lymphoedema. The American Venous Forum Guidelines (Gloviczki, 2016) also issue guidance for the use of compression in lymphoedema. These guidelines state that 30-40 mmHg and rarely 50-60 mmHg of pressure are recommended for advanced lower extremity fibrotic lymphoedema or for comorbidities such as morbid obesity. They state that a pressure of 20-30 mmHg is usually sufficient for upper limb lymphoedema. Authors of the STRIDE algorithm for compression garment selection (Bjork and Ehmann, 2019) recommend compression from 30-40 mmHg up to 50-60 mmHg for patients with lymphoedema. They recommend that effective prescription of compression is based on the presentation of the patient and not the diagnosis.

See appendix [III.II](#) for further guidance on suitable compression for various conditions.

Recommendation

GQ48.1 Effective compression prescription requires matching the compression selection to the patient presentation and patient choice, not to the diagnosis alone.

Evidence Grade: C

Strength of recommendation: Strong

GQ49: How should compression garments be selected?

Evidence Summary

A 2019 literature review addressed this question (Reich-Schupke and Stucker, 2019). To date, there appear to be no studies comparing circular/round-knit compression garments and flat-knit compression garments. The type of compression garment chosen should be based on the clinical presentation. Typically patients with significant differences in limb circumferences, severe oedema of the toes/forefoot or those with deep skin folds tend to require flat-knit garments. There are, however, some patients for whom circular/round-knit with a high degree of stiffness would be more appropriate. The authors suggest that typical indications for flat-knit garments include significant differences in leg circumference as well as deep skin folds and toe/forefoot.

The Dutch Guidelines (Damstra and Halk, 2017) recommend that flat knit compression stockings with high stiffness be prescribed in patients with lymphoedema where possible.

Custom Made versus Off-the-Shelf

The ILS recommend that a prescription of custom-made garments with specific measurements be obtained if needed but do not indicate which patients are in need of these (ILS, 2016). The American Venous Forum guidelines state that most patients can be fitted with off-the-shelf garments but that those with severe, misshapen lymphoedema may require custom made garments (Gloviczki, 2016). The highest compression level required to maintain oedema and which is tolerated by the patient is likely to be the most beneficial. If not maintaining volume reduction, then compression pressures should be increased as tolerated. Prior to maintenance therapy, it is essential to adequately decongest the area using compression bandages and/or adaptive compression systems.

See the [S.T.R.I.D.E guidance document](#) on compression garment selection for comprehensive guidance, including guidance on layering compression garments. See appendix [III.II](#) for indication for use of compression garments.

Recommendations

GQ49.1 Every patient requires a thorough assessment and clinicians should always use their clinical judgement to provide the best garment (s) suited to each individual patient.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.2 When selecting appropriate compression for a patient with lymphoedema, consider the patient's vascular status, ability to tolerate compression and ability to manage the garment.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.3 The pressure applied by the garment should counteract capillary filtration pressure which is higher in the leg while standing than while supine. Therefore, the garment needs to exert a higher pressure while standing.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.4 Typically patients with significant differences in limb circumference, severe oedema of the toes/forefoot or those with deep skin folds tend to require flat-knit garments.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.5 Patients who cannot tolerate off-the-shelf garments or those with misshapen limbs should be considered for custom made garments, in accordance with patient choice.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.6 Lower compression may be used for patients with palliative needs with comfort being the main treatment goal.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.7 Lower limb compression garments exert graduated pressure with the highest pressure gradient at the ankle. The highest compression level required to maintain oedema and which is tolerated by the patient is likely to be the most beneficial.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.8 Upper limb compression garments exert graduated pressure with the highest pressure gradient at the wrist. The highest compression level required to maintain oedema and which is tolerated by the patient is likely to be the most beneficial.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.9 In some circumstances, layering of garments might be necessary to control swelling.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.10 Layering garments may be considered to enable greater tolerance of higher levels of pressure and easier donning.

Evidence Grade: D

Strength of recommendation: Strong

GQ49.11 The total pressure applied by two garments, layered one over the other, may not be equal to the sum of their individual pressures in the upper limb. Furthermore, caution should be used to ensure maintenance of the pressure gradient from lower pressure proximally to higher pressure distally (e.g. at the wrist).

Evidence Grade: D

Strength of recommendation: Strong

GQ49.12 When using a combination of flat and circular knit garments, the inner layer should be flat-knit and the outer layer should be circular-knit.

Evidence Grade: D

Strength of recommendation: Strong

GQ50: What is the efficacy of compression wrapping devices (wraps) in the management of lymphoedema?

Evidence Summary

Compression wrap systems are becoming more common place in the management of lymphatic and venous conditions. Different wrap systems deliver pressures of 30-60 mmHg depending on the brand used. A review of the literature on adjustable compression wrap devices assisted in answering this question (Williams, 2016). This review concludes that the majority of evidence for the use of compression wraps is in the form of descriptive insights and anecdotal evidence. The research evidence is very limited for the use of adjustable compression wrap devices in people with lymphoedema. Most evidence is in the form of descriptive papers, case studies, or small research studies. There is clinical evidence that adjustable compression wrap devices provide improved quality of life (QoL) and independence for patients with lymphoedema.

A recent small study (n = 36) compared the efficacy of conventional multilayer short-stretch bandaging to a Velcro-adjustable compression wrap (Borman et al., 2021). Outcomes studied included: volume reduction, ultrasound measurements, function, and QoL during the active CDT period in patients with lower limb lymphoedema. All study participants received skin care education, MLD, and supervised lymphoedema exercises. They were then randomised to Group One (multilayer short-stretch bandaging-Rosidal-K®) or Group Two (adjustable-compression-velcro-wrap-Circaid Reduction-kit®) for 3 weeks, totalling 15 sessions. This adjustable compression velcro-wrap performed as a part of CDT greatly reduced limb volume similar to multilayer bandaging, and also led to improvements in QoL. It can be a comfortable alternative to the conventional multilayer bandages in the active treatment phase of the CDT.

An RCT by (Pujol-Blaya et al. (2019) compared the effectiveness of a precast adjustable compression system to that of MLLB in patients with BCRL. The trial included 42 patients who were randomised to a precast adjustable compression system or to MLLB. Both groups were found to have significant reductions in limb volume and symptoms after intervention and the between group difference was non-significant.

Expert opinion supports the use of compression wrap systems, prescribed by clinicians trained in the application of compression wraps, in the management of lymphoedema.

Recommendation

GQ50.1 Clinicians should consider the use of compression wrap systems in the treatment and long term management of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ50.2 All clinicians prescribing and fitting compression wraps should undergo adequate training to ensure competency and patient safety.

Evidence Grade: D

Strength of recommendation: Strong

GQ50.3 Clinicians should ensure that two liners are provided to each patient when they are being fitted for compression wraps.

Evidence Grade: D

Strength of recommendation: Strong



Education Need:

All clinicians prescribing and fitting compression wraps should undergo adequate training to ensure competency and patient safety.

GQ51: What is the efficacy of prophylactic compression garments in the prevention of lymphoedema?

Evidence Summary

There is a paucity of research available to answer this question. A systematic review of the use of medical compression in lymphatic disease reported that there is currently insufficient evidence to comment on the efficacy of prophylactic compression garment use in the prevention of lymphoedema after surgery (Rabe et al., 2018). The American Venous Forum state that while they recommend patients with lymphoedema wear compression while undertaking air travel, they cannot (based on the National Lymphoedema Network's advice) recommend prophylactic use of compression (Gloviczki, 2016).

Recommendations

GQ51.1 Compression garments should only be prescribed prophylactically if there is a diagnosis of sub-clinical lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ51.2 If a patient is diagnosed with sub-clinical lymphoedema then the short term wearing of compression garments is recommended during a set monitoring period. See question GQ52 for guidance on compression duration in subclinical lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ51.3 Further research is required to assess the efficacy of prophylactic compression garments in the prevention of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

Assessing the efficacy of prophylactic compression garments in the prevention of lymphoedema.

GQ52: For what minimum duration should patients with subclinical lymphoedema wear compression garments?

Evidence Summary

There is limited evidence available to answer this question. Expert opinion considers that volume and BIS should have returned to baseline as a marker of minimum duration. Small studies have suggested a minimum of 4 weeks for upper limb lymphoedema before reassessment (Stout Gergich et al., 2008) and 6 months for lower limb lymphoedema.

If the measurement has returned to baseline and this is maintained over a 3 - 6 month period, then the patient can be weaned off the garment by gradually reducing the number of days per week it is worn. During this weaning period, the patient must carefully monitor their limb and inform the therapist if they observe any deterioration. See appendices [I.IV.II](#) and [I.IV.III](#) for pathways.

Recommendations

GQ52.1 Patients with subclinical upper limb lymphoedema using compression should be reviewed by their lymphoedema therapist after a 4 week period. In cases where patients do not return to baseline measurements, compression should be continued with monthly reviews.

Evidence Grade: D

Strength of recommendation: Strong

GQ52.2 Patients with subclinical lower limb lymphoedema using compression should be reviewed after a 3 month period. In cases where patients do not return to baseline measurements, compression should be continued with 3-monthly reviews.

Evidence Grade: D

Strength of recommendation: Strong

GQ52.3 If lymphoedema progresses and limb volume increases by more than 10%, refer to specialist lymphoedema service for ongoing management.

Evidence Grade: D

Strength of recommendation: Strong

GQ52.4 If the measurement has returned to baseline and is maintained over a 3 - 6 month period, then the patient can be weaned off the garment by gradually reducing the number of days per week it is worn. During this weaning period, the patient must carefully monitor their limb and inform the therapist if they observe any deterioration.

Evidence Grade: D

Strength of recommendation: Strong

See appendices [I.IV.II](#) and [I.IV.III](#) for breast and gynaecological pathways

GQ53: When should compression garments be worn by patients with lymphoedema?

Evidence Summary

Guidelines published by the American Venous Forum (Gloviczki, 2016) state the following in relation to when compression should be worn:

- During intensive reduction periods: multi-layered bandaging is placed after MLD and should be worn constantly, except while washing.
- During maintenance phase: the patient is fitted with an elastic or inelastic compression garment to use during waking hours (some patients will need nocturnal compression as well).
- While carrying out exercise.

The Queensland Guidelines (2014) recommend that following intensive therapy, garments should be worn 23 out of 24 hours until the limb volume has stabilised.

- Patients with very mild/subjective symptoms – daily garment wear may be considered. Intermittent garment wear may be required for activities which exacerbate symptoms—for example, for heavy physical activities, exercise.
- People with severe lymphoedema—indefinite night time bandaging/garment wear may be required in combination with daytime garment wear.
- People who have undergone treatment for stage I (early) lymphoedema, particularly of the upper limb, may not require lifelong use of compression garments.
- Daytime garment wear may be recommended for people waiting long periods of time for intensive therapy and for those who do not require intensive treatment.
- Compression garments should maintain the volume reduction achieved in the initial management phase.
- People who have completed intensive therapy may require day and night time compression until stable limb volumes have been achieved.
- Garments worn at night should be lower in compression level than daytime garments. Once stability has been achieved, night time compression may no longer be necessary. However, to establish this, a trial of weaning night time compression should be undertaken.
- Limb volumes may take up to 6–12 months to stabilise.

Recommendations

GQ53.1 Compression is recommended to control lymphoedema and patients typically require life-long compression.

Evidence Grade: D

Strength of recommendation: Strong

GQ53.2 During intensive reduction treatment: compression (short stretch or inelastic bandage systems) should be applied after MLD and should be worn constantly, except while washing. New technology allows compression garments or wraps to also be considered in specific cases.

Evidence Grade: D

Strength of recommendation: Strong

GQ53.3 During the maintenance phase the patient is measured and fitted with an elastic or inelastic compression garment to use during waking hours.

Evidence Grade: D

Strength of recommendation: Strong

GQ53.4 Some patients may require nocturnal compression; this should be based on clinician judgment, particularly regarding duration of use. There is a variety of nocturnal techniques/products that may be considered.

Evidence Grade: D

Strength of recommendation: Strong

GQ53.5 Patients who undergo liposuction for the treatment of lymphoedema will require 24-hour compression, unless otherwise directed by the treating surgeon.

Evidence Grade: D

Strength of recommendation: Strong

GQ54: How often should compression garments be re-fitted or replaced?

Evidence Summary

It is currently recommended by a number of bodies and manufacturers to replace compression garments every 3 - 6 months (Gloviczki, 2016, Damstra and Halk, 2017, Bjork and Ehmann, 2019, Queensland Health, 2014). There does not appear to be any original trial evidence to support this recommendation. Garments may need to be replaced more frequently in more active patients or for those living with obesity.

See PQ14 for recommendations on garment replacement in paediatric patients.

Recommendations

GQ54.1 Compression garments should be replaced according to manufacturer guidelines, which would typically be every 6 months or sooner.

Evidence Grade: D

Strength of recommendation: Strong

GQ54.2 For children with lymphoedema, garments should be reviewed more often according to their growth pattern and additional garments should be supplied as required.

Evidence Grade: D

Strength of recommendation: Strong

GQ54.3 Patients should be supplied with a minimum of two garment sets per body part at each six month review.

Evidence Grade: D

Strength of recommendation: Strong

GQ54.4 Damaged garments should be replaced to ensure adequate compression is applied.

Evidence Grade: D

Strength of recommendation: Strong

GQ54.5 In cases of significant weight or oedema fluctuation, such that garments no longer fit correctly or are uncomfortable, garments should be remeasured and new sets supplied.

Evidence Grade: D

Strength of recommendation: Strong

GQ55: Should compression garments be removed if patients develop cellulitis?

Evidence Summary

The Australasian Lymphology Association consensus document (Association, 2015) on the management of cellulitis in lymphoedema recommends the removal of compression garments and avoidance of MLD during an acute attack of cellulitis. They recommend re-commencing compression once tolerated by the patient. If the swelling persists the patient should be reassessed by a HCP to ensure compression is correctly fitted.

The Queensland Health Guidelines (2014) on compression therapy in lymphoedema recommend that compression be applied after antibiotics have been initiated, as tolerated by the patient.

See the BLS cellulitis guidance document (www.thebls.com).

Recommendation

GQ55.1 Compression garments should be removed and MLD should be avoided during acute cellulitis presentation. Usual compression should be recommenced as soon as the patient achieves baseline tolerance of activity and compression.

Evidence Grade: D

Strength of recommendation: Strong

GQ55.2 Once antibiotics for cellulitis are initiated, compression may be applied if tolerated. Alternative modified compression (e.g. looser compression garments or bandaging) may be in considered in the initial stages.

Evidence Grade: D

Strength of recommendation: Strong

GQ56 Does compression therapy reduce recurrent cellulitis in patients with lymphoedema?

Evidence Summary

A recent RCT examined the impact of compression therapy on cellulitis in adults with chronic oedema (Webb et al., 2020) and found it resulted in a lower incidence of recurrent cellulitis than conservative treatment.

In this trial 84 patients with chronic oedema were randomised to one of two groups: one group received leg compression therapy plus education on cellulitis prevention (n = 41) and the control group (n = 43) received education alone. Follow-up took place every 6 months for up to 3 years. Participants in the control group who had an episode of cellulitis crossed over to the compression group. The trial was stopped for efficacy at the time of planned interim analysis. Of the compression group just 6 patients (15%) had an episode of cellulitis compared to 17 (40%) in the control group (hazard ratio, 0.23; 95% CI 0.09 - 0.59; P=0.002; RR, 0.37; 95% CI 0.16 - 0.84; P=0.02). No adverse events occurred during the trial and there was no significant difference in QoL measures between groups.

A large epidemiological study by (Burian et al., 2021) supports the use of compression to reduce recurrent cellulitis in patients with lymphoedema. While this study did not directly examine the impact of compression on cellulitis, it looked at the effect of controlled swelling (with compression as a composite of control, along with eczema care etc.) on incidence of cellulitis. Control of swelling in patients with lymphoedema was associated with a reduced risk of developing cellulitis (OR 0.59, 95% CI 0.51–0.67).

Refer to question GQ55 for guidance on the use of compression in patients with acute cellulitis.

Recommendations

GQ56.1 The significant role of compression in preventing cellulitis in patients with lymphoedema should be included in all educational material and opportunities.

Evidence Grade: D

Strength of recommendation: Strong

GQ56.2 Compression therapy is an integral component of clinical and subclinical lymphoedema management.

Evidence Grade: B

Strength of recommendation: Strong

GQ57: Should patients with lymphoedema wear compression garments while exercising?

Evidence Summary

A large systematic review examining exercise in cancer related lymphoedema answered this question (Singh et al., 2016). The authors concluded that there is insufficient evidence to support or refute the use of compression garments while exercising in patients with secondary lymphoedema. Expert consensus of the guideline group is that garments should be worn as tolerated by patients while exercising.

Recommendations

GQ57.1 Compression garments should be worn by patients with lymphoedema during exercise if they are tolerated.

Evidence Grade: D

Strength of recommendation: Strong

GQ57.2 Patients with lymphoedema should monitor their own limb during and after exercise regardless of whether they are wearing a compression garment or not.

Evidence Grade: D

Strength of recommendation: Strong

GQ58: Is MLD and/or compression bandaging safe in patients with open wounds?

Evidence Summary

While there do not appear to be any trials examining the safety of MLD or bandaging in patients with open wounds, one review (Towers, 2010) states that MLD and bandaging should be adapted in cases where tumours infiltrate the skin or subcutaneous tissue. Similarly, open wounds do not constitute an absolute contraindication to MLD or compression bandaging.

Recommendations

GQ58.1 Open wounds do not constitute an absolute contraindication to MLD or compression bandaging in patients with lymphoedema. Modified MLD should be considered in the treatment of oedema in patients with open wounds. Clinicians should evaluate the affected area regularly and observe for any local changes.

Evidence Grade: D

Strength of recommendation: Strong

GQ58.2 Clinicians should liaise with tissue viability services and/or nursing staff in the treatment of patients with lymphoedema and open wounds.

Evidence Grade: D

Strength of recommendation: Strong

GQ59: What is the efficacy of intermittent pneumatic compression pumps in the management of lymphoedema?

Evidence Summary

A systematic review and meta-analysis (Rogan et al., 2016) of available evidence in lymphoedema management reported that intermittent pneumatic compression (IPC) appears to reduce lymphoedema volume in the acute phase. A second systematic review (Phillips and Gordon, 2019) of the evidence regarding IPC dosage in lymphoedema found low-moderate quality evidence that 45-60 minutes of IPC at 30-60 mmHg may result in significant improvements. The American Venous Forum Guidelines recommend the use of compression pumps to reduce lymphoedema in some patients (Gloviczki, 2016) and the Queensland Guidelines recommend IPC can be effective as part of a combined treatment programme for BCRL in the short term and up to 2 months post treatment.

A 2020 RCT (Tastaban et al., 2020) examined the role of IPC in the treatment of BCRL (n = 76). The trial compared the addition of IPC to CDT, to CDT alone. Percentage limb volume reduction decreased in both groups and the difference was non-significant. Intermittent pneumatic compression seems to add no benefit when combined with complex decongestive treatment of lymphoedema, however it may assist in symptom management (e.g. sensation of limb heaviness).

A randomised waitlist trial of 43 patients examined the effectiveness of advanced pneumatic compression in patients with head and neck cancer-related lymphoedema (Ridner et al., 2021). The trial showed that advanced pneumatic compression was safe, with no serious adverse events reported. Patients who received advanced pneumatic compression reported significant improvement in their ability to control lymphoedema (p = 0.003), visible external swelling (p < 0.001), as well as reported pain.

Recommendations

GQ59.1 Intermittent pneumatic compression (IPC) can be used in conjunction with other treatments for lymphoedema, for limited durations and depending on patient compliance.

Evidence Grade: A

Strength of recommendation: Strong

GQ59.2 Due to the lack of evidence to support benefits of intermittent pneumatic compression, clinicians should use their clinical judgement to decide if it would be beneficial to individual patients.

Evidence Grade: D

Strength of recommendation: Strong

GQ59.3 Patients with lymphoedema should be taught simple manual lymphatic drainage in order to perform intermittent pneumatic compression, to ensure adequate movement of fluid and avoid development of genital lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ60: Does any evidence exist regarding the effectiveness of bandaging?

Evidence Summary

Compression bandaging is a common treatment for people with lymphoedema. Short-stretch bandages are typically used in the multi-layer lymphoedema bandaging (MLLB).

Indications for MLLB include (Queensland Health, 2014):

- Moderate to severe lymphoedema (20%–40% excess volume)
- Distorted limb shape
- Lymphorrhoea or broken skin
- Subcutaneous tissue thickening

A randomised clinical trial by (Torres-Lacomba et al., 2020) compared different bandage and taping regimens in 150 patients with BCRL. Patients were randomly assigned to one of five groups to compare the effectiveness of multilayer, simplified multilayer, cohesive or adhesive bandaging or Kinesio tape. There were significant differences between the bandage groups in absolute value of excess limb volume (P < 0.001). The most effective bandaging type was simplified multilayer (59.5%, IQR = 28.7) and cohesive bandaging (46.3%, IQR = 39). Kinesio tape (4.9%, IQR = 17.7) and adhesive bandages (21.7%, IQR = 17.9) had the least effect on limb volume. However, all groups exhibited significant reduction in symptoms.

An RCT demonstrated the efficacy of MLLB compared to elastic hosiery alone (Badger et al., 2000). The reduction in limb volume in the MLLB arm was approximately double that of the hosiery-alone cohort and this reduction was sustained at 6 months. A second small RCT (n = 42) found that MLLB and standard care had a significantly greater reduction in limb volume compared to patients treated with Kinesio tape (Tsai et al., 2009). A third RCT reported that mean volume reduction was significantly higher in an MLLB-treated cohort compared to a standard bandaging cohort (Didem et al., 2005). A small (n = 29) controlled comparative study (DAMSTRA et al., 2008) that found inelastic MLLB leads to an immediate reduction in limb volume both in lymphoedematous and normal lower limbs.

The Queensland Health Guidelines recommend that bandaging as a component of combined therapy is efficacious, but owing to a lack of evidence conclusions regarding its efficacy alone cannot be drawn. While the benefit of bandaging in upper limb lymphoedema management is established, there is a paucity of trials in the literature to investigate its effect in lower limb lymphoedema. The authors recommend that “As part of combined treatment programmes, MLLB is more effective at reducing upper limb lymphoedema volume than: compression garments, Kinesio tape, ‘standard’ bandaging, elevation and exercise.”

Recommendations

GQ60.1 If indicated, using MLLB bandaging may be more beneficial than using compression garments alone. The use of bandaging as part of lymphoedema treatment should always be individually assessed and based on clinical reasoning.

Evidence Grade: A

Strength of recommendation: Strong

GQ61: What is the effectiveness of bandaging-alone in the treatment in lymphoedema?

Evidence Summary

Multi-layer lymphoedema bandaging (MLLB) was shown to be similarly effective to CDT, in terms of limb volume reduction in a small prospective study (n = 103) of patients with lymphoedema (Zasadzka et al., 2018). Very few studies have examined the individual components of CDT, so the efficacy of any given individual element is not currently supported. A small study (n = 38) of patients with BCRL compared low stretch MLLB alone to MLLB combined with MLD (Johansson et al., 1999). The addition of MLD to MLLB resulted in a significantly greater reduction in arm volume than using MLLB alone. The American Venous Forum Guidelines (Gloviczki, 2016) recommend combined treatment programs, citing lack of evidence for the volume-reducing contribution of compression or MLD alone.

Recommendation

GQ61.1 The use of MLLB alone is not recommended for the treatment of Lymphoedema. MLLB should be used as an integral part of a combined treatment approach including skin care, exercise, and MLD if indicated.

Evidence Grade: C

Strength of recommendation: Strong

GQ62: Is there evidence to support effective treatment of lymphorrhoea?

Evidence Summary

While there is a distinct lack of trial evidence available to answer this question, case studies as well as expert opinion support the use of multilayer compression bandaging in patients with lymphorrhoea, and it is generally considered safe and effective for these patients (Towers, 2010). In some patients with severe lymphorrhoea, lower limb bandaging may be helpful even if it does cause some proximal swelling (a commonly reported side effect of treatment). It is therefore important to involve the patient in medical decision-making. Lymphorrhoea usually responds well to continuous compression bandaging. Frequent changes in bandaging may be required, often more than once a day (Regnard et al., 1997). Non-adherent dressing materials such as paraffin-impregnated gauze may be beneficial at the leaking area (Renshaw, 2007).

Pressures applied may need to be reduced or more gradually increased compared to standard care. It may be advisable to use fewer bandage layers and lighter materials for bandaging (e.g. tubigrip). Community lymphoedema therapists in some regions may not have access to all types of dressing materials and may need to involve hospital or community tissue viability nurses.

See Lymphorrhoea pathway in Appendix I.V

Recommendations

GQ62.1 Multilayer compression bandaging is effective in the treatment of lymphorrhoea. [See Lymphoedema Network Wales Lymphorrhoea Pathway](#) for further advice on lymphorrhoea management. (Appendix I.V)

Evidence Grade: D

Strength of recommendation: Strong

GQ62.2 Multilayer compression bandaging should be commenced as soon as possible in the treatment of lymphorrhoea in patients to prevent skin maceration.

Evidence Grade: D

Strength of recommendation: Strong

GQ62.3 Clinicians should consider liaising with nursing staff when treating patients with lymphorrhoea.

Evidence Grade: D

Strength of recommendation: Strong

GQ62.4 Clinicians should consider liaising with tissue viability nurses when treating patients with lymphorrhoea who have compromised skin integrity or those with open wounds.

Evidence Grade: D

Strength of recommendation: Strong

1.6 Manual Lymphatic Drainage

GQ63: Does any evidence exist regarding the effectiveness of MLD?

Evidence Summary

Evidence regarding the efficacy of MLD is mixed. A 2015 Cochrane Review examining the efficacy of MLD in the treatment of BCRL (Ezzo et al., 2015) concluded that MLD is safe and may be of additional benefit to compression bandaging in terms of reducing lymphoedema. The review cautioned that firm conclusions regarding its efficacy could not be drawn. When results from trials were pooled, swelling reduced by 30% to 37% in those treated with an intensive course of compression bandaging. The addition of MLD to this regimen reduced swelling by a mean additional 7.11%. The authors therefore concluded that MLD is safe and may offer additional benefit to compression bandaging for swelling reduction. They also concluded that compared to individuals with moderate-to-severe BCRL, those with mild-to-moderate BCRL may be the ones who benefit from adding MLD to an intensive course of treatment with compression bandaging.

The ISL recommends the use of MLD in specific populations (i.e. early BCRL, new oedema and/or early stage lymphoedema without adipose or fibrotic tissue deposition) but there is a need for more robust evidence. For symptoms such as pain and heaviness, 60%-80% of participants reported feeling better regardless of which treatment they received. An older systematic review and meta-analysis of RCTs (Huang et al., 2013) found that evidence does not support the use of MLD in the prevention or treatment of lymphoedema, however, clinical and statistical inconsistencies between the various studies confounded the evaluation. The effect of MLD on health-related QoL is unclear, owing to the lack of evidence in this field of study (Müller et al., 2018).

The effectiveness of MLD reduces as BMI increases, see the [obesity section](#) of this guideline for guidance on MLD for people living with obesity.

Prevention

A Cochrane Review published in 2015 examining preventative measures against BCRL included evidence from four trials. The authors concluded that at that time there was insufficient evidence to draw firm conclusions about the efficacy of MLD in preventing BCRL. A systematic review (Müller et al., 2018) of RCTs examining the efficacy of MLD in improving health-related quality of life (HRQoL) in patients with chronic oedema found that currently, the evidence is unclear as to the benefits of MLD on HRQoL in this cohort.

Recommendations

GQ63.1 MLD is recommended for the treatment of lymphoedema in the intensive phase, in conjunction with compression (CDT) but only when therapeutically indicated and for patients with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ63.2 The routine use of MLD without compression is not recommended. MLD alone may be useful in certain specific conditions (e.g. head and neck cancer, midline lymphoedema, palliative care).

Evidence Grade: D

Strength of recommendation: Strong

GQ63.3 MLD, as part of CDT, may be required as a supplemental management option for specific indications e.g. after severe cellulitis.

Evidence Grade: D

Strength of recommendation: Strong

GQ64: Is there any evidence to support frequency and length of MLD treatment?

Evidence Summary

The American Venous Forum (Gloviczki, 2016) recommends a minimum of 5 days per week in the lymphoedema reduction stage of treatment. MLD should be provided with compression therapy and, due to new bandaging systems, the frequency of MLD (as part of CDT) has been reduced.

Owing to expert consensus, however, within this guideline development group and the advent of newer technologies, it is recommended that patients should be provided with compression therapy between 2 - 5 sessions per week until symptoms have plateaued and agreed goals have been reached. The duration of MLD required is variable and depends on the complexity of the swelling, as well as the number of areas requiring treatment.

Recommendations

GQ64.1 MLD, if indicated, should be provided with compression therapy between 2-5 sessions per week and until symptoms have plateaued and agreed goals have been reached.

Evidence Grade: D

Strength of recommendation: Strong

GQ65: Are there any contraindications to manual lymphatic drainage?

Evidence Summary

Previously authors have postulated that MLD may cause spread of neoplastic disease or pre-existing cellulitis or exacerbation of heart failure secondary to increased venous return (Lawenda et al., 2009). Others have suggested that it may lead to tissue damage or to pulmonary embolism secondary to DVT propagation. According to a review of the evidence supporting therapies in the treatment of lymphoedema, these theories have been disproven by several studies (Smile et al., 2018) with no authors reporting complications associated with MLD. A study examining the haemodynamic response to MLD found no contraindication in heart failure (Leduc et al., 2011). To date, no studies have found evidence that MLD can cause neoplastic spread (Pinell et al., 2008).

However, the lymphoedema schools of Vodder, Casley Smith, Klose and Leduc list the following conditions as contraindications to MLD:

- Acute cellulitis
- Active lymphangitis
- Active erysipelas
- Acute inflammation
- Untreated/unstable heart failure
- Acute deep vein thrombosis (DVT)
- Superior vena cava (SVC) Obstruction

The above schools also recommend that caution be exercised when using MLD in the following patients:

- Risk of DVT
- Suspicion of undiagnosed malignancy or recurrence
- Renal disease
- Liver cirrhosis (especially with concurrent ascites)
- Cardiac arrhythmias, hyperthyroidism, hypersensitivity of carotid sinus, > 60 years of age (risk of arteriosclerosis), abdominal aortic aneurism (AAA)
- Pregnancy, menstrual period, recent abdominal surgery, radiation fibrosis, colitis, cystitis, IBS, Crohn's disease, diverticulosis, unexplained pain
- Osteoporosis, radiation fibrosis, bone metastasis
- Bronchial asthma
- Thyrotoxicosis
- Lymph nodes previously affected by TB
- Hypotension
- Unstable hypertension
- Cancer treatment (i.e. radiotherapy or chemotherapy)
- Toothache

Recommendations

GQ65.1 MLD should be avoided in acute cellulitis, lymphangitis, erysipelas, unstable heart failure, acute inflammation, SVCO, and untreated deep venous thrombosis (DVTs).

Evidence Grade: D

Strength of recommendation: Strong

GQ65.2 There are precautions that should be considered after full assessment and risk screening which include malignancy, pregnancy, hypotension, unstable hypertension and renal disease.

Evidence Grade: D

Strength of recommendation: Strong

GQ66: Does any evidence exist regarding the effectiveness of self-lymphatic drainage (SLD)?

Evidence Summary

While an older systematic review (Ridner et al., 2012) suggested evidence was lacking to recommend any self-management method for practice, more recent evidence appears to support the effectiveness of self-lymphatic drainage (SLD) in lymphoedema management (Temur and Kapucu, 2019, Arinaga et al., 2019, Arinaga et al., 2016).

A systematic review (Douglass et al., 2016) assessing various methods of self-management in lymphoedema concluded that SLD is effective in reducing limb volume in patients with cancer-related lymphoedema. Four studies which used SLD with compression garments and one study which used SLD without compression, found significant decreases in limb volume reported (2.59%-60% reduction) over 3 and 6 months in a 122 patient cohort. The same review found that studies including SLD lead to improved QoL at all-time points and for up to 6 months of follow up. Empowerment of people with lymphoedema to care for themselves with access to supportive professional assistance has the capacity to optimise self-management practices and improve outcomes from limited health resources.

See EQ2 for advice on available educational supports.

Recommendations

GQ66.1 Self Lymphatic Drainage should be taught to all people who have the capability and competence to perform the techniques.

Evidence Grade: D

Strength of recommendation: Strong

GQ66.2 Self Lymphatic Drainage should be taught to families and carers where the person with lymphoedema is unable to perform the techniques themselves.

Evidence Grade: D

Strength of recommendation: Strong

GQ67: How should patients be instructed to carry out self-lymphatic drainage?

Evidence Summary

There does not appear to be evidence advocating specific skills required for self-lymphatic drainage (SLD). Some authors highlight the importance of patient knowledge for optimal adherence to a self-management regimen as part of lymphoedema management (Alcorso et al., 2016). Expert opinion recommends that SLD should be taught by a lymphoedema therapist and that patients be provided with written or online instruction be supported by reviewing individual technique.

Recommendations

GQ67.1 Patients undertaking self-lymphatic drainage (SLD) should be taught by a lymphoedema therapist and be provided with written or online instruction be supported by reviewing individual technique. The LNNI website (www.lnni.org) has an open directory of suitable written material and a link to a mobile app, all of which support SLD.

Evidence Grade: D

Strength of recommendation: Strong

GQ67.2 If a patient is unable to undertake self-lymphatic drainage, family members or carers should be provided with online instruction once patient consent has been provided.

Evidence Grade: D

Strength of recommendation: Strong

G67.3 Patients should be directed to online education videos to assist in educating patients and/or family members or carers to undertake self-lymphatic drainage. The videos are available from the [LNNI website](http://www.lnni.org).

Evidence Grade: D

Strength of recommendation: Strong

Good Practice Point

The following guidance on self-lymphatic massage is provided by a Canadian University Health Network:



- Use a light pressure and keep your hands soft and relaxed.
- The pressure of your hands on your skin should be just enough to gently stretch the skin as far as it naturally goes, and then releasing. If you can feel your muscles underneath your fingers, then you are pressing too hard.
- Use the flats of your hands instead of your fingertips. This allows more contact with the skin to stimulate the lymph vessels.
- Massage towards areas of your body that have not been treated for cancer
- Try to do the massage when you are comfortably warm because your muscles will be more flexible.
- Make sure you are comfortable while doing the massage. You can try a seated, standing or lying down position.
- Try to do self-massage everyday
- If you need to do the massage on both sides of your body, start on one side of your body and go through each step. Once you have completed the steps on one side, repeat them on the other side of your body

1.7 Pharmacological Treatment

GQ68: Is there any evidence to support the pharmacological treatment of lymphoedema?

Evidence Summary

A systematic review in 2019 of pharmacotherapy agents in the management of lymphoedema addressed this question (Forte et al., 2019a). In total, 7 studies assessing the efficacy of pharmacotherapy in lymphoedema were identified. The agents identified by studies include cyclophosphamide (injection), sodium selenite / selenium (PO), ketoprofen or Pegsunercept (soluble TNF- α receptor R1) (Subcutaneous injection), tacrolimus (topical). All of these agents appear to exert their effects by attenuating the inflammatory response, a component of lymphoedema. All agents appear to show promising results but larger scale randomised control trials are required to validate these preliminary findings. The agents appeared to be well tolerated by patients and no adverse effects were reported in the trials.

Selenium

Three studies (Kasseroller and Schrauzer, 2000, Micke et al., 2003, Zimmermann et al., 2005) assessed the effectiveness of selenium in the treatment of lymphoedema associated with breast and head and neck cancer, all of which reported positive results with no adverse effects reported in study cohorts.

Ketoprofen

Ketoprofen is a nonsteroidal anti-inflammatory drug (NSAID). One pilot study including 21 patients with lymphoedema assessed the efficacy of ketoprofen in humans with lymphoedema in an open-label trial (Rockson et al., 2018). Subjects with primary and secondary lymphoedema were given 75mg ketoprofen PO three times daily for 4 months. Skin thickness and histopathology were significantly reduced compared to baseline. Based on these findings a placebo-controlled study was carried out (n = 34). This trial showed a reduction in skin thickness, decreased plasma G-CSF and improved histopathological markers. No adverse events were reported.

Topical Tacrolimus

The use of topical tacrolimus was investigated in one experimental animal study (Gardenier et al., 2017). The results of this have yet to be examined in human trials.

Soluble TNF- α Receptor R1

Pegsunercept, a soluble TNF- α receptor 1 inhibitor, was trialled in an animal study and it did not result in a positive impact on lymphoedema compared to controls (Nakamura et al., 2009).

Cyclophosphamide

A case series (n = 4) described the use of cyclophosphamide injections in patients with secondary lymphoedema due to metastatic breast cancer (Kitchen and Garrett, 1971). Two of the four patients improved in terms of symptom relief and lymphoedema.

Evidence Summary (cont.)

Benzopyrones

The use of 5, 6 Benzo-[Alpha]-Pyrone (coumarin) has previously been investigated for its potentially beneficial effects in lymphoedema management. This is an organic chemical compound found in many plants. Its mechanism of action is thought to be mediated via increased proteolysis of high protein oedema fluid by macrophages (Casley-Smith et al., 1993). A Cochrane Review on the use of benzopyrones to reduce lymphoedema concluded that it was not possible to draw firm conclusions on the effectiveness of benzopyrones in reducing limb lymphoedema or associated symptoms. The International Society of Lymphology (2016) stated that the role of benzopyrones is, as of yet, undetermined and cautioned that coumarin has been linked to hepatotoxicity in some patients (ILS, 2016).

Steroids

Corticosteroids may be helpful in the management of lymphoedema associated with advanced cancer management. The most commonly used steroid in palliative care for these purposes is dexamethasone.

Targeted Therapies

There has been a recent increase in studies examining the efficacy of targeted therapies, both systemic and topical, to treat lymphoedema. A systematic review (Forte et al., 2019b) examining six such experimental trials on animals found overall positive outcomes. The interventions studied included two broad categories of therapy:

- 1) induction of lymphangiogenesis with vascular endothelial growth factor-C hydrogel or fibroblast growth factor
- 2) inflammatory modulators e.g. tacrolimus, topical collagen or troxerutin-phosphatidylcholine

While these therapies have yet to be assessed in human, results from these preliminary experimental studies appear promising.

Recommendations

GQ68.1 Due to lack of evidence routine pharmacological treatment for lymphoedema is not recommended

Evidence Grade: A

Strength of recommendation: Strong

GQ68.2 In rare, specific conditions associated with lymphoedema (e.g. lymphangiodyplasia), pharmacotherapeutic options may be prescribed under specialist supervision.

Evidence Grade: D

Strength of recommendation: Strong

GQ69: Are diuretics effective in the management of lymphoedema?

Evidence Summary

The majority of evidence for diuretic therapy in the treatment of lymphoedema was carried out on small cohorts in the 1950s and 1960s in patients with primary lymphoedema (Cattell et al., 1965). There do not appear to be any recent trials examining the effectiveness of diuretics in the management of either primary or secondary lymphoedema. In their 2016 consensus document, the International Society Of Lymphology (2016) recommend that diuretics have limited use in the initial stage of CDT in select patients with specific comorbidities. They recommend long-term administration of diuretics should be avoided as they are of limited value in peripheral lymphoedema and they may cause harmful side effects including electrolyte disturbance.

Recommendation

GQ69.1 The use of diuretics is not recommended for lymphoedema treatment but may be used where applicable for the treatment of co-morbidities.

Evidence Grade: D

Strength of recommendation: Strong

GQ70: Are calcium channel blockers contraindicated in patients with lymphoedema?

Evidence Summary

A nested case-control study of adult female patients with breast cancer was reported including 717 cases and 1,681 matched controls (Stolarz et al., 2019). After controlling for baseline characteristics, calcium channel blocker (CCB) use (28.3% vs. 23.3%; $P = 0.0087$), was higher in patients who also developed lymphoedema. In adjusted analysis, CCB exposure was significantly associated with increased risk of lymphoedema (OR = 1.320; 95% confidence interval, 1.003-1.737). However while these results may imply a correlation between CCB use and lymphoedema, they do not prove causation.

A 1998 epidemiological study of patients with lymphoedema also found CCB use to be higher in patients with lymphoedema than those without (Lee et al., 2015, Stolarz et al., 2019, Michelini et al., 1998). The IUAP Guidelines recommend calcium-channel blocking agents should be avoided as they impair lymphatic pumping (Lee et al., 2015).

Recommendation

GQ70.1 While there is currently insufficient evidence to contraindicate the use of calcium channel blockers in patients with or at risk of lymphoedema, it is advised that patients should be aware that there are alternative types of drugs available and should discuss this with their doctor.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

The impact of calcium channel blockers on lymphatic function should be further examined.

GQ71: What is the role of non-medical prescribers in lymphoedema?

Evidence Summary

A Cochrane Review of the literature on non-medical prescribing addressed this question (Weeks et al., 2016). This review included 46 studies (n = 37,337) on non-medical prescribing. Of the included studies, 26 studies included nurses and 20 included pharmacists. This review concluded that non-medical prescribers were as effective as medical prescribers. These nurses and pharmacists include subjects practising with varying levels of autonomy in terms of prescribing and in a variety of settings. This review suggests that non-medical prescribing delivers similar outcomes for Hr-QoL, patient adherence and satisfaction. Non-medical prescribing compares favourably in relation to outcomes for systolic blood pressure, HbA1c and LDL levels.

Inconsistency and reporting variability across studies meant that the review could not determine adverse outcomes and resource use in relation to non-medical prescribing.

Recommendations

GQ71.1 Lymphoedema teams should consider the role of non-medical prescribing to improve the effective and efficient prescribing of garments and other lymphoedema-related products.

Evidence Grade: A

Strength of recommendation: Strong

1.8 Cellulitis

GQ72: What differential diagnosis should be considered for suspected cellulitis?

Evidence Summary

Cellulitis is over diagnosed and treated in patients with lower limb redness (Walsh et al., 2016). A diagnosis of red leg syndrome should be considered in patients with bilateral redness, warmth, swelling and tenderness without systemic symptoms. Cellulitis is a bacterial infection, nearly always unilateral, painful, often with raised serum CRP. Red legs can be attributed to gravitational eczema, dermatitis or other chronic conditions which do not respond to antibiotics. Management of red legs involves good skin care, antipruritic cream if itchy or antihistamines or topical steroids.

See [Appendix I.VII](#) for the BLS Red Legs Pathway for comprehensive guidance.

Recommendations

GQ72.1 Clinicians should use the BLS Red Leg differential diagnosis pathway to ensure that a correct diagnosis is made for suspected cellulitis.

Evidence Grade: D

Strength of recommendation: Strong

GQ72.2 If cellulitis is suspected following the diagnosis pathway, clinicians should refer the patient to their GP for assessment.

Evidence Grade: D

Strength of recommendation: Strong

GQ72.3 If the condition is not suspected to be cellulitis, follow the guideline which may result in a referral to dermatology.

Evidence Grade: D

Strength of recommendation: Strong

GQ73: Are antibiotics effective in the prevention of recurrent cellulitis in patients with lymphoedema?

Evidence Summary

There do not appear to be any clinical trials investigating the effectiveness of antimicrobial prophylaxis in patients with lymphoedema specifically. A systematic review and meta-analysis of 5 RCTs (n = 535) including healthy patients found that antibiotic prophylaxis significantly reduced the rate of recurrent cellulitis (RR: 0.46; 95% CI: 0.26 - 0.79).

The BLS issue guidance on the [prevention and treatment of cellulitis](#) in people living with lymphoedema and this guidance is reviewed on an annual basis (Society, 2016). This guideline development group endorses these current recommendations, which are due to be updated in 2022.

The National Institute for Health and Care Excellence (NICE) also offer comprehensive guidance on the prevention and management of cellulitis. The guideline development group also endorse these recommendations available [here](#).

Recommendation

GQ73.1 Antibiotic prophylaxis may be beneficial in preventing recurrent cellulitis in certain patients with lymphoedema. See the [BLS guidance](#) on this topic for a list of comprehensive and up-to-date recommendations. (www.thebls.com)

Evidence Grade: D

Strength of recommendation: Strong

GQ74: Which antibiotic regimen is recommended in the treatment of cellulitis in patients with lymphoedema?

Evidence Summary

The efficacy of antibiotic therapy in the treatment of skin infections is well established. Patients with lymphoedema are more prone to developing skin infections such as cellulitis and erysipelas and the use of antibiotics in such cases is recommended.

The BLS issue guidance on the [prevention and treatment of cellulitis](#) in people living with lymphoedema and this guidance is reviewed on an annual basis (British Lymphology Society, 2016). This guideline development group endorses these current recommendations, which are due to be updated in 2022 (www.thebls.com).

The National Institute for Health and Care Excellence (NICE) also offer comprehensive guidance on the prevention and management of cellulitis. The guideline development group also endorse these recommendations available [here](#).

Recommendation

GQ74.1 There are several antibiotic regimens available to treat cellulitis in patients with lymphoedema. Clinicians should be aware of the [BLS guidance](#) on this topic, however they should consult their local antimicrobial guidelines in the first instance.

Evidence Grade: D

Strength of recommendation: Strong

1.9 Physical Activity

GQ75: What is the role of physical activity in the treatment of lymphoedema?

Evidence Summary

Current evidence suggests that all types of exercise can improve subjective and objective parameters in patients with BRCL. A large systematic review (Baumann et al., 2018) including 11 RCTs of 458 women with BRCL examined the efficacy of exercise on outcomes. Exercises studied included aerobic exercise, resistance exercise, yoga, swimming, aqua lymph training and gravity-resistance exercise. Four of these trials found significant reductions in arm volume and seven of the studies reported significant subjective improvements. No studies reported adverse effects of exercise.

Recommendations

GQ75.1. The benefits of exercise should be strongly highlighted, and an agreement reached with the patient as soon as possible to include regular exercise as an integral part of their lymphoedema treatment programme.

Evidence Grade: D

Strength of recommendation: Strong

GQ75.2 Consideration should be given to provide services which support self-management and include exercise. This could be in the form of an external contract or managed within services.

Evidence Grade: D

Strength of recommendation: Strong

GQ75.3. All exercise should be started at a low level and progressed slowly.

Evidence Grade: D

Strength of recommendation: Strong

GQ76: What is the effectiveness of water-based exercises in Lymphoedema management?

Evidence Summary

A 2018 systematic review and meta-analysis addressed this question (Yeung and Semciw, 2018). Four RCTs, of moderate-quality evidence assessed the effectiveness of water-based aqua lymphatic therapy (ALT) exercises and found no significant short term effect of these compared to land-based regimens in terms of lymphoedema outcomes, as measured by limb volume (SMD: 0.14; 95% confidence interval [CI]: -0.37 to 0.64, I²=0%, p=0.59). There was low-quality evidence of no significant difference in improving upper limb function (SMD: -0.27, 95% CI: -0.78 to 0.23, I²=0%, p=0.29).

A small pilot study (n = 7) of lower limb lymphoedema secondary to gynaecological cancer found aquatic training enabled patients with lower limb lymphoedema to engage in vigorous exercise which correlated with an increase in functional capacity and QoL (Dionne et al., 2018).

A controlled trial comparing land exercise to aquatic exercise effects on body composition in patients recovering from breast cancer, found that land-based exercise produced a greater decrease in body composition but water-based exercise improved breast symptoms (Fernández-Lao et al., 2013). One RCT examined the use of ALT and found that it resulted in a clinically significant immediate effects on limb volume but no long-term effect was apparent (Tidhar and Katz-Leurer, 2010).

Recommendations

GQ76.1 Water-based activity programmes can be beneficial with no adverse effects reported, therefore can be recommended to patients with lymphoedema.

Evidence Grade: A

Strength of recommendation: Strong

GQ76.2. All exercise must be progressed slowly according to tolerance in patients with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ77: Is resistance exercise safe in patients with lymphoedema?

Evidence Summary

2015 Cochrane Systematic Review (Stuiver et al., 2015) suggests that progressive resistance exercise therapy does not increase the risk of developing lymphoedema, provided that symptoms are closely monitored and adequately treated if they occur. These findings were echoed in a recent large systematic review of 23 papers examining the impact of resistance exercise on BCRL. The authors of this review concluded that resistance exercise appears safe and does not increase the risk of developing lymphoedema in at-risk patients (Hasenoehrl et al., 2020). This echoes findings of their earlier review on the same topic (Keilani et al., 2016) and those of a further systematic review (Paramanandam and Roberts, 2014). A recent systematic review of systematic reviews examining the role of rehabilitation interventions after treatment for breast cancer concluded that resistance training has positive effects on limb volume reduction and muscle strengthening (Olsson Möller et al., 2019).

These findings were mirrored in recommendations by the expert panel convened by the American Society of Breast Surgeons (McLaughlin et al., 2017a), who made the following recommendation: "... Resistance and aerobic exercise is safe. Patients with BCRL should work with a trained lymphoedema professional to learn to exercise safely". The 2018 NICE guidelines on Early and locally advanced Breast Cancer Management also recommend that there is no clear evidence that exercise causes, prevents or worsens lymphoedema (NICE, 2014).

Recommendation

GQ77.1 Progressive resistance exercise is safe and should be utilised in the management of patients with lymphoedema.

Evidence Grade: A

Strength of recommendation: Strong

GQ78: What precautions should be taken by patients with lymphoedema when exercising?

Evidence Summary

A large systematic review examining exercise in cancer related lymphoedema, including 25 studies answered this question (Singh et al., 2016). The authors concluded that there is insufficient evidence to support or refute the use of compression garments while exercising in patients with secondary lymphoedema. There was no effect of exercise on lymphoedema or on lymphoedema-related symptoms. In subgroup analysis there was no difference in effect comparing aerobic, resistance and mixed-methods exercise, nor was there any difference in exercise duration (interventions greater than 12 weeks versus under 12 weeks). Exercise appears safe in patients with secondary lymphoedema and is not associated with worsening of lymphoedema or related symptoms. A systematic review of multiple modalities of exercise including 11 RCTs reported no adverse effects of exercise on BCRL (Baumann et al., 2018). A further meta-analysis of multiple modalities including aerobic, stretching, resistance training, yoga and Pilates concluded that all of these exercises appear safe in patients with or at risk of BCRL (Panchik et al., 2019).

Recommendations

GQ78.1 Exercise is safe for patients with lymphoedema and patients should be advised to monitor for any changes to symptoms. Any persistent changes (e.g. numbness, or increased swelling) should be reported to their lymphoedema clinician.

Evidence Grade: D

Strength of recommendation: Strong

GQ78.2. All exercise must be progressed slowly in patients with lymphoedema and HCPs should consider patient comorbidities.

Evidence Grade: D

Strength of recommendation: Strong

GQ79: What exercise prescription is recommended for patients with lymphoedema?

Evidence Summary

There is no specific evidence for exercise prescription specifically in patients with lymphoedema. However, it is recommended that patients with lymphoedema can follow the most recent American College of Sports Medicine (ACSM 2018) guidelines on exercise. ACSM guidelines on exercise are as follows:

- Cardiorespiratory Exercise: adults should get at least 150 minutes of moderate-intensity exercise per week.
- Resistance Exercise: adults should train each major muscle group on two or three days each week using a variety of exercises and equipment.

The BLS have produced a document highlighting the role of exercise and physical activity in patients with lymphoedema (Society, 2020). This guidance recommends that clinicians consider the following exercises in patients with lymphoedema:

- Deep breathing exercises or activities involving this e.g. singing
- Swimming or aqua aerobics
- Yoga and Pilates
- Tai chi and qigong
- Nordic walking and any other type of brisk walking
- Set programmes of muscle pumping and strengthening exercises
- Gym work
- Any additional preferred activity for general fitness e.g. dancing.

Recommendations

GQ79.1 It is recommended that patients with lymphoedema follow the most recent ACSM guidelines on exercise.

Evidence Grade: D

Strength of recommendation: Strong

GQ79.2 Consider the use of pelvic floor exercises for both men and women if there is genital swelling.

Evidence Grade: D

Strength of recommendation: Strong

GQ79.3 In patients with head, neck or facial swelling consider the use of range-of-movement exercises and use of a beaded collar and facial stretches.

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

The following modifications to exercise are recommended in patients with lymphoedema:

- Maintain hydration and avoid extreme heat
- Ensure adequate rest intervals between sets
- Compression sleeves or bandages should be worn if acceptable to patient and does not impede movement

GQ80: Is there evidence that abdominal or deep breathing increases lymphatic flow?

Evidence Summary

There does not appear to be any evidence available to answer this question. A published debate by lymphoedema experts discusses the influence of breathing on lymphatic drainage (Piller, 2006). Most experts interviewed support the use of deep breathing exercises and believe anecdotally that they increase lymphatic flow despite a lack of evidence demonstrating this. Older animal studies have shown that exercise and deep breathing do increase lymphatic flow (Browse et al., 1974, Browse et al., 1971).

Deep breathing exercises are recognised to have a calming effect and overall benefit to general well-being. In relation to SLD, deep breathing aids in the preparation for SLD by helping to slow down movements. Deep breathing is usually encouraged with simultaneous arm movements or stretches which involves movement of the skin that is essential in SLD. Deep breathing is also associated with syphoning fluid centrally, however it could be argued that there is more evidence for incorporating deep breathing techniques into the self-management due to the psychological and movement benefits.

Recommendation

GQ80.1 Deep breathing exercises, with emphasis on diaphragmatic breathing, should be incorporated into treatment plans to increase lymphatic flow in patients with oedema.

Evidence Grade: D

Strength of recommendation: Strong

1.10 Nutrition and Lymphoedema

GQ81: What is the impact of weight gain versus maintaining a healthy weight on lymphoedema management outcomes?

Evidence Summary

There was limited evidence available to answer this question. One small case series (Greene et al., 2015b) proposed that patients with a BMI > 50 kg/m² may have permanently damaged lymphatics due to their body mass.

A threshold may exist for lymphoedema development with a tipping point between 50 kg/m² and 60 kg/m² identified (Greene et al., 2015) at which point lower extremity lymphatic function appears to become dysfunctional. Greene et al. performed lymphoscintigraphy in 15 people with severe obesity with no prior history of lymphoedema. The average BMI of those with lymphoedema was 70.1 kg/m² (range, 59.7 to 88.1), significantly greater than the average BMI of 42.0 kg/m² (range, 30.7 to 53.3) in those without lymphoedema (p < 0.001). All patients with a BMI above 59 kg/m² had lymphoedema, whereas each patient with a BMI less than 54 kg/m² had normal lymphatic function.

See the [obesity section](#) of this guideline for more detail on the impact of BMI on people with lymphoedema.

Recommendations

GQ81.1 All lymphoedema patients should be encouraged to follow a healthy diet and be referred to dietetics if appropriate.

Evidence Grade: D

Strength of recommendation: Strong

GQ81.2 HCPs must ensure that patients with a high BMI are encouraged and supported to maintain or lose weight, as patients with a BMI > 50 kg/m² may have permanently damaged lymphatics due to their body mass. This may include partnership-working with bariatric teams.

Evidence Grade: C

Strength of recommendation: Strong

GQ82: Is there any evidence to support medical nutritional intervention in patients with lymphoedema?

Evidence Summary

Nutrition is important for everyone; regardless of body size, weight or health status. (Brownell et al., 2010) Nutritional interventions for obesity-induced lymphoedema should be nutritionally adequate, culturally acceptable and affordable for long-term adherence. HCPs should adapt nutrition interventions and/or adjuvant therapy to meet their patients' individual values, preferences and treatment goals. However, to date, it appears that there is 'no one-size-fits-all' nutritional intervention for obesity-induced lymphoedema (Koliaki et al., 2018). Nutritional interventions should be based on a collaborative care approach with a registered dietitian who has experience in obesity management and medical nutritional therapy. Dietitians can support people living with obesity who also have: other chronic diseases, malnutrition, food insecurity or disordered patterns of eating (Williams et al., 2019).

Individualised medical nutritional therapy for obesity-related lymphoedema should promote a healthy relationship with food, consider the social context of eating and promote eating behaviours that are sustainable and realistic for the individual (Puhl and Heuer, 2010, Brownell et al., 2010, Ramos Salas et al., 2019). Systematic reviews and meta-analyses of randomised controlled trials have shown that individualised nutrition consultations by a registered dietitian decrease weight by an additional -1.03kg and BMI by -0.43 kg/m² in participants with a BMI \geq 25 kg/m² compared with usual care or written documentation (Williams et al., 2019). Calorie restriction can achieve short-term reductions in weight (i.e. < 12 months) but has not shown to be sustainable long-term (i.e. > 12 months). Caloric restriction may in some individuals lead to pathophysiological drivers to promote weight gain via exaggerated hunger, appetite and decreased satiety. In addition, caloric restrictions may impair skeletal health and muscle strength, contributing to the role of individualizing nutrition interventions that are safe, effective and meet the values and preferences of the patient with obesity-related lymphoedema.

However, to date, there appears to be no single best nutrition intervention to sustain weight loss long-term, and literature continues to support the value of long-term adherence, regardless of the intervention. It is worth noting that obesity-related lymphoedema may cause irreversible lymphatic dysfunction which may not resolve with weight loss. Nevertheless, systematic reviews and meta analyses of RCTs assessing weight loss interventions for the treatment and prevention of BCRL have found that dietary advice to reduce energy intake can reduce BCRL (Schmitz, 2010).

Consequently, nutritional interventions for obesity-related lymphoedema should emphasise individualised eating patterns, food quality and a healthy relationship with food. Such interventions may consider mindfulness-based eating practices that may help lower food-cravings, reduce reward-driven eating, improve body satisfaction and improve awareness of hunger and satiety (Todd, 2019). Future research should assess nutrition-related outcomes, health-related behavioural changes in addition to weight and body composition outcomes instead of weight loss outcomes alone across all weight spectrums (Wharton et al., 2020). Based on the lack of current evidence it is not possible to recommend "a single best nutritional intervention plan" for people living with obesity-related lymphoedema (Koliaki et al., 2018, Williams et al., 2019, Johnston et al., 2014).

Recommendations

GQ82.1 There is currently not enough evidence to recommend "a single, best nutritional intervention plan" for people living with obesity-related lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ82.2 Patients with lymphoedema should be advised to follow national guidelines on nutrition.

Evidence Grade: D

Strength of recommendation: Strong

GQ83: Is there a safe level of alcohol intake for patients with lymphoedema?

Evidence Summary

There is no literature discussing the effects of alcohol on lymphoedema. Several practitioners advise against alcohol intake owing to the known diuretic effect of alcohol however studies examining the effect of alcohol intake on lymphoedema are lacking in the literature.

There is inconclusive evidence regarding the effects of alcohol on recurrence rates of breast cancer. Studies have shown a link between alcohol consumption and an increase in breast cancer recurrence. Other studies have not established such a link, with some highlighting the cardioprotective effect of alcohol when consumed in lower volumes (Rock et al., 2012).

The ESPEN guidelines (Arends et al., 2017) recommend: "In cancer survivors we recommend to maintain a healthy weight (BMI 18.5–25 kg/m²) and to maintain a healthy lifestyle, which includes being physically active and a diet based on vegetables, fruits and whole grains and low in saturated fat, red meat and alcohol."

Recommendations

GQ83.1 Patients with lymphoedema should be advised to follow national guidelines on consumption of alcohol.

Evidence Grade: D

Strength of recommendation: Strong

GQ84: What is the impact of dietary supplements in the treatment of lymphoedema?

Evidence Summary

According to a large 2017 review of integrative therapies delivered during and after breast cancer treatment, there is no strong evidence to support the use of dietary supplements in this patient population (Greenlee et al., 2017). A Cochrane Review examining the efficacy of selenium in reducing side effects of cancer treatments including chemotherapy, radiotherapy and surgery (Dennert and Horneber, 2006) concluded that there is insufficient evidence to make recommendations in favour or against selenium supplementation in patients living with cancer. Conversely, other authors have reported that selenium can reduce upper limb breast cancer related lymphoedema after surgery and radiotherapy (Samuels et al., 2014, Micke et al., 2003, Kasseroller, 1997). Other trials have found that vitamin E supplementation does not lead to a significant difference in upper limb lymphoedema post mastectomy or post radiotherapy (Gothard et al., 2004).

A number trials have looked at novel supplement therapies. One such supplement is Robuvit®, a natural extract from French oak wood. The authors of a trial examining its efficacy in reducing limb volume post mastectomy and post radiotherapy found that supplementation of a CDT programme with Robuvit® can further reduce limb volume (Belcaro et al., 2018). A review (Wanchai et al., 2013) of complementary and alternative medicines including horse-chestnut complex (Wheat et al., 2009), coumarin (Casley-Smith et al., 1993, Loprinzi et al., 1999), vitamin E (Gothard et al., 2004), Ginkor Fort (Cluzan et al., 2004) concluded that there is insufficient evidence to support the use of these therapies in patients living with cancer and lymphoedema.

A study examining patients' own experience with complementary therapies including vitamin supplementation, found perceived effectiveness ratings to be similar between mainstream treatment (mean ± SD: 5.3 ± 1.5) and complementary treatments (Mean ± SD: 5.2 ± 1.6) (Finnane et al., 2011).

Recommendations

GQ84.1 Due to the inconsistent evidence we do not recommend the use of dietary supplements in the treatment of lymphoedema. Further research is required in this area.

Evidence Grade: D

Strength of recommendation: Strong

1.11 Psychological Intervention

GQ85: How can practitioners identify the need for psychological intervention in lymphoedema?

Evidence Summary

There was no evidence available to answer this question however expert opinion is that clinicians working within lymphoedema services should manage local and non-complex psychological support for their patients.

Current PROMs do not have specific trigger scores to support onward referral to psychology services. There is a requirement to support patients to access the correct level of psychological intervention. This may be achieved by the development of PROM threshold scores.

Recommendations

GQ85.1 Results of patient reported outcome measures may be used to guide clinicians to appropriate pathways options. See appendix [I.VI](#) for an algorithm to support access to psychological care.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

The potential utility of patient related outcome measures to predict need for, and level of, psychological support should be studied.

GQ86: How should clinicians offer psychological support to patients living with lymphoedema?

Evidence Summary

It is widely recognised that mental health issues are common in patients who suffer with many chronic conditions. Body-image related distress has been reported as a frequent symptom in patients who have undergone treatment for breast cancer (Sherman et al., 2018).

It is the expert opinion of the International Lymphoedema Framework (2010) that all patients with lymphoedema should receive a psychological screening assessment to identify those who require additional psychological support and those who require specialist psychological referral. They recommend the following be included in the psychological screening:

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

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

Screening for depression

NICE recommends that screening for depression should include the use of at least two questions concerning mood and interest e.g.

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

See psychological Care Pathway (Appendix [I.VI](#))

Recommendations

GQ86.1 All patients with lymphoedema should receive a Quality of Life screening assessment, which includes screening for depression, to identify those who require additional psychological support and those who require specialist psychological referral.

Evidence Grade: D

Strength of recommendation: Strong

GQ86.2 Lymphoedema services should have funded access to psychological services.

Evidence Grade: D

Strength of recommendation: Strong

GQ86.3 As part of every clinician’s duty of care, consideration should be given to refer to non-specialist locally provided, supportive care, if required.

Evidence Grade: D

Strength of recommendation: Strong

GQ86.4 As part of every clinician’s duty of care, referral should be made to the patient’s GP or consultant if there is evidence of self-reported psychological distress associated with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ87: What is the impact of support groups for patients with or at risk of developing lymphoedema?

Evidence Summary

A small qualitative study of Japanese Breast Cancer survivors (Tsuchiya et al., 2012) revealed that many patients with lymphoedema felt uncomfortable disclosing their symptoms to others. The authors recommend support programmes to allow breast cancers survivors to discuss their lymphoedema. It is recognised as important that patients with Lymphoedema may want to talk to other people with lymphoedema.

While there is minimal specific evidence for the efficacy of support groups in lymphoedema, the guideline development group recommend that support groups are beneficial in this patient cohort based on evidence of benefit in other medical conditions.

Recommendations

GQ87.1 Patients should be made aware of and have access to support groups on various platforms (e.g. Lymphoedema Support Network, Lymphoedema Ireland).

Evidence Grade: D

Strength of recommendation: Strong

1.12 Adjunctive Therapies

GQ88: Is there any evidence that fluoroscopy is beneficial in the treatment of lymphoedema?

Evidence Summary

Fluoroscopy guided manual lymphatic drainage (FG-MLD) also known as the “Fill and Flush” method, involves the visualising the lymphatic network to identify the location most in need of lymphatic drainage. Currently, only preliminary evidence of its effectiveness is available. In two small studies (Belgrado et al., 2016, Tan et al., 2011), the physiological effect of one session of FG-MLD was shown.

A double-blinded multicentre RCT, “Effort-BCRL trial” examining the effectiveness of this technique is currently underway (De Vrieze et al., 2018). Until results of this trial are available, conclusions regarding the efficacy of FG-MLD cannot be drawn.

Recommendations

GQ88.1 At present there is insufficient evidence to support the use of fluoroscopy in the standard management of lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

The effectiveness of fluoroscopy in the treatment of lymphoedema should be further examined.

GQ89: Is there any evidence to support the use of Kinesio tape in treatment of lymphoedema?

Evidence Summary

Several systematic reviews have addressed this question (Kasawara et al., 2018, Gatt et al., 2017). One review included seven studies (Kasawara et al., 2018), each showing a positive effect in reducing lymphoedema after treatment. The authors however caution that there are no trials comparing Kinesio tape to controls or other treatments. Kinesio taping appears effective in post-mastectomy BCRL however it does not appear to be superior to other treatments due to the increased risk of dermatological complications.

A second review (Gatt et al., 2017) examining the effectiveness and safety of Kinesio tape in the management of BCRL, when compared to compression bandaging or hosiery included six randomised controlled trials. A meta-analysis of this trial data revealed no significant difference between the treatment groups in terms of limb measurement and an increased risk of skin complications was found in the Kinesio tape arms of several studies, affecting 10%-21% of subjects. Patients treated with Kinesio tape reported better lymphoedema-related symptoms compared to compression therapy. The authors conclude that Kinesio tape should only be used with caution if bandaging is not possible. Several RCTs have assessed the efficacy of Kinesio tape in the treatment of BCRL with differing results (Pekyavas et al., 2014, Tsai et al., 2009, Pop et al., 2014, Malicka et al., 2014, Pajero Otero et al., 2019, Melgaard, 2016, Tantawy et al., 2019, Martins Jde et al., 2016, Ozsoy-Unubol et al., 2019, Taradaj et al., 2014).

A 2020 randomised clinical trial compared the effectiveness of four types of bandages and Kinesio tape for treating BRCL (Torres-Lacomba et al., 2020). The trial found Kinesio tape to be the least effective in terms of absolute reduction in limb volume, however it was perceived as the most comfortable by women, and multilayer as the most uncomfortable ($P < 0.001$).

Recommendations

GQ89.1 The use of Kinesio tape in the treatment of lymphoedema is recommended only as part of a combined approach along with the conventional CDT approach.

Evidence Grade: A

Strength of recommendation: Strong

GQ89.2 The use of Kinesio tape in the treatment of lymphoedema is based on patient and clinician preference.

Evidence Grade: D

Strength of recommendation: Strong

GQ89.3. A skin test should be carried out prior to application of Kinesio tape. When Kinesio tape is used, the skin should be closely monitored for changes or reactions to the tape.

Evidence Grade: D

Strength of recommendation: Strong

GQ90: What training is required to use Kinesio tape in the treatment of lymphoedema?

Evidence Summary

There appears to be a lack of research studies examining training required to treat patients with lymphoedema. Two review articles on Kinesio taping (Bosman, 2014, Finnerty, 2010) highly recommend that practitioners undergo certified training in Kinesio taping before treating patients with lymphoedema.

Of note, most training courses for Kinesio taping in lymphoedema require that the attendee be a certified lymphoedema therapist as a prerequisite.

Recommendation

GQ90.1 Lymphoedema clinicians using Kinesio tape should undertake relevant CPD including formal and informal training, to ensure competency in Kinesio taping.

Evidence Grade: D

Strength of recommendation: Strong

GQ91: What is the efficacy of acupuncture in the treatment of lymphoedema?

Evidence Summary

There is evidence that acupuncture may be beneficial in the treatment of lymphoedema in patients with BCRL (Zhang et al., 2019). This meta-analysis and systematic review of 6 RCTs found that acupuncture is effective at reducing BCRL. The trials included in this review (Smith et al., 2014, Yao et al., 2016, Bao et al., 2018) were deemed of good reporting quality overall. A more recent systematic review and meta-analysis concluded however that while acupuncture tends to improve lymphoedema symptoms, it does not appear to significantly change arm circumference in BCRL (Chien et al., 2019).

A review of complementary medicines and their role in lymphoedema treatment included trials examining acupuncture, concluded that there is insufficient evidence to recommend any complementary therapies at this time (Wanchai et al., 2013). This review included a number of older studies which reported mixed results in terms of acupuncture efficacy (Kanakura et al., 2002, de Valois et al., 2012, Alem and Gurgel, 2008, Cassileth et al., 2011).

Recommendations

GQ91.1 Acupuncture may be considered as an adjunctive treatment to assist in symptom control as part of the overall treatment plan for lymphoedema. It is not currently recommended as a treatment to reduce limb volume.

Evidence Grade: D

Strength of recommendation: Strong

GQ91.2 Acupuncture needling should not be applied to the affected area in patients with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

GQ92: What is the effectiveness of Low level laser therapy (LLLT) in the treatment of lymphoedema?

Evidence Summary

Low-level laser therapy (LLLT) also known as photobiomodulation therapy refers to the use of thermally safe energy level photons to alter biological activity. LLLT is used in some countries in the treatment of BCRL. It has been proposed that LLLT stimulates an immune response, in particular that of macrophages, which facilitates breakdown of scar tissue and hence improved lymphatic flow. LLLT is also proposed to have positive effects in the lymphangiogenesis pathway.

Findings of a 2015 meta-analysis (Smoot et al., 2015) support the use of LLLT in BCRL treatment. The pooled results of these studies showed that LLLT led to reductions in arm volume and pain. These results were validated by a more recent systematic review on the topic, which recommends LLLT be considered an effective treatment in BCRL but cautions that there is a need for well-designed high-quality trials to research the area (Baxter et al., 2017). LLLT does not however appear superior to other treatments according to another systematic review examining the efficacy of LLLT, which failed to find studies comparing LLLT to complex physical therapy (E Lima et al., 2014). Some authors postulate that LLLT can increase cancer recurrence or metastasis rates but there is currently no evidence available to address this theory (Borman, 2018, E Lima et al., 2014).

LLLT is recommended as a "C-graded" therapeutic modality in the treatment of Lymphoedema by the Society of Integrative Oncology (Greenlee et al., 2017), a recommendation endorsed by the American Society of Clinical Oncology (Lyman et al., 2018). This recommendation is based off the assessment of 2 clinical trials in LLLT (Ahmed Omar et al., 2011, Ridner et al., 2013), both of which report mixed results.

Recommendation

GQ92.1 Evidence supports safe use of laser therapy however due to the time consuming nature of laser therapy and the costs involved, conventional treatments (e.g. CDT) for lymphoedema should be considered first.

Evidence Grade: C

Strength of recommendation: Strong

GQ93: What is the efficacy of deep oscillation therapy in the management of lymphoedema?

Evidence Summary

An early RCT (Jahr et al., 2008) randomised 21 patients to a treatment group (n = 11) who received 12 sessions of MLD plus Deep Oscillation, or to the control group (n = 10) who received only MLD.

In this trial deep oscillation resulted in significant pain and swelling reduction in the treatment group, suggesting the addition of deep oscillation to MLD was beneficial in patients with secondary breast lymphoedema, compared to MLD alone.

Tio (2016) did a very small study which concluded that deep oscillation reduces lower-limb oedema and is at least as efficacious as MLD in achieving volume reduction. Prospective studies with larger numbers of participants are required to further evaluate this treatment option.

Recommendation

GQ93.1 There is currently insufficient evidence to recommend the use of deep oscillation therapy as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

A high-quality, sufficiently powered prospective study examining the efficacy of deep oscillation therapy in the treatment of lymphoedema should be carried out.

GQ94: What is the efficacy of negative pressure therapy in the management of lymphoedema?

Evidence Summary

Negative pressure therapy is also known as “Physiotouch” or “LymphaTouch”.

In small case studies in clients with lymphoedema, when compared with manual lymphatic drainage, “LymphaTouch” was shown to improve treatment outcomes, decrease treatment time and improve patient satisfaction (Vuorinen et al, 2013; Osborne, 2015; Whitaker, 2015). Gott (2018) concluded that negative pressure therapy offers a new and innovative method for treating lymphoedema and can potentially improve complex lymphoedema therapy outcomes.

Further randomised controlled trials are required in this area before conclusions can be drawn regarding effectiveness.

Recommendations

GQ94.1 There is currently insufficient evidence to recommend the use of negative pressure therapy as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong

GQ95: What is the role of platelet-rich plasma in the management of lymphoedema?

Evidence Summary

Platelet-rich plasma (PRP) is an autologous, concentrated preparation of platelets, which is thought to have lymphangiogenic and tissue-repairing effects. Although PRP has been safely used in many different fields, there are few studies examining the use of PRP in lymphoedema treatment in humans.

One RCT (Akgul et al., 2020) assessed the clinical outcomes of PRP in patients (n = 45) with lower extremity lymphoedema. Patients were randomly allocated to one of three groups: treatment with PRP and CDT, low-level laser therapy with CDT, and CDT alone.

While significant differences in LYMQOL, LEC, NRS, and TDC values were found in all three groups, there were no statistically significant differences between the three groups.

Recommendations

GQ95.1 There is currently insufficient evidence to recommend the use of PRP as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

A study examining the efficacy of PRP in the treatment of lymphoedema.

GQ96: What is the efficacy of aromatherapy in the treatment of lymphoedema?

Evidence Summary

Small single-centre studies have reported that while patients find aromatherapy and essential oil based therapy beneficial in terms of symptom-burden, these interventions do not appear to lead to clinically meaningful reduction in limb volume (Barclay et al., 2006, Arinaga, 2012). Qualitative studies have reported positive perceived benefits of improved lymphoedema symptoms after aromatherapy massage in patients living with cancer (Ho et al., 2017).

There is no significant evidence to recommend aromatherapy in the treatment of lymphoedema, however there is some evidence of positive experience with no adverse effects. The evidence supports improvements in QoL rather than direct improvement in limb volume.

Recommendations

GQ96.1 Aromatherapy may be considered as an adjunctive treatment to assist in symptom control as part of the overall treatment plan for lymphoedema. It is not currently recommended as a treatment to reduce limb volume.

Evidence Grade: D

Strength of recommendation: Strong

GQ97: What is the efficacy of reflexology in the treatment of lymphoedema?

Evidence Summary

A qualitative study (Whatley et al., 2018) examining the perceived benefit of reflexology in patients with lymphoedema yielded positive results, with many patients reporting that they find reflexology beneficial, both psychologically and physically.

Recommendations

GQ97.1 Reflexology may be considered as an adjunctive treatment to assist in symptom control as part of an overall treatment plan for lymphoedema. It is not currently recommended as a treatment to reduce limb volume.

Evidence Grade: D

Strength of recommendation: Strong

GQ98: What is the efficacy of reiki in the treatment of lymphoedema?

Evidence Summary

There is no evidence available to answer this question. A large scale systematic review (Greenlee et al., 2017) of integrative therapies during and after breast cancer treatment failed to find evidence to support any complementary treatment in this population, aside from laser therapy.

Recommendation

GQ98.1 There is insufficient evidence to make a recommendation on reiki as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong

GQ99: What is the efficacy of cannabidiol (CBD) oil in the treatment of lymphoedema?

Evidence Summary

There is no evidence available to answer this question. A large scale systematic review (Greenlee et al., 2017) of integrative therapies during and after breast cancer treatment failed to find evidence to support any complementary treatment in this population, aside from laser therapy.

Recommendation

GQ99.1 There is insufficient evidence to make a recommendation on CBD as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong

GQ100: What is the impact of therapeutic massage on lymphoedema?

Evidence Summary

Therapeutic massage is classified as a complementary or alternative therapy in the treatment of lymphoedema. Manual lymphatic drainage is a more complex technique, applied by the therapist to assist with lymphatic drainage. This technique is not included in the training for routine therapeutic massage.

Several reviews on alternative therapies include therapeutic massage and conclude that currently there is insufficient evidence to recommend for or against it as a treatment for lymphoedema (Wanchai et al., 2013).

Recommendation

GQ100.1 There is insufficient evidence to make a recommendation on therapeutic massage as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong

GQ101: What is the effectiveness of dry brushing in the treatment of lymphoedema?

Evidence Summary

Dry brushing involves the use of a dry brush to softly stroke the skin and is self-administered by the patient. It is thought to increase lymphatic movement and anecdotally, patients report it provides symptomatic relief, however at this time there appears to be a lack of evidence to support these claims.

Recommendation

GQ101.1 There is insufficient evidence to make a recommendation on dry brushing as a standard treatment for lymphoedema until further research is published.

Evidence Grade: D

Strength of recommendation: Strong

GQ102: What benefits would a specific lymphoedema minimum data set provide services?

Evidence Summary

One of the early objectives of the NLP was to develop a minimum data set, to stimulate the gathering of consistent data about patients accessing lymphoedema services.

An MDS should help to describe the picture of who is receiving care. This is important at both national and local levels. Out of this we hope that a national strategy will be prioritised. At a local level, a service that is able to collate the data of its individual patients will be in a better position to justify future resourcing of lymphoedema services.

There was limited evidence available to answer this question however a pilot study carried out by the NLP concluded that it appears that it is feasible to collect information at the initial patient assessment that allows the minimum data set to be completed. The pilot service plans to incorporate the MDS questions into the ordinary clinical documentation within the electronic record so that the data can be reported electronically.

From discussion at a national level, the MDS has been modified and simplified to facilitate the gathering of comparable data with the view to strengthening the argument for improved resourcing of lymphoedema service provision. The National Lymphoedema Partnership and BLS, as one of its constituent members strongly encourage the use of the MDS to achieve this aim. Further work will take place to facilitate its roll out and the MDS tools will be made available on the BLS website in the near future.

Please see appendix [IV.II](#) for the LNNI (NLP modified, 2017) Lymphoedema MDS. The HSE Lymphoedema MDS will soon be available in excel format for download at www.hse.ie/lymphoedema.

Recommendations

GQ102.1 Each service should maintain a minimum lymphoedema data set available in appendix [IV.II](#). An Excel version is available on hse.ie/lymphoedema

Evidence Grade: D

Strength of recommendation: Strong

GQ102.2 The results of the minimum lymphoedema data set should be accessible at a local and national level.

Evidence Grade: D

Strength of recommendation: Strong

Good Practice Point

Benefits of having a minimum data set for lymphoedema:

- 1) *To assess the breakdown of the patients' attending services.*
- 2) *To gather data on how many patients present with different aetiologies/ severities / sites of oedema and the prevalence of cellulitis / wounds.*
- 3) *To demonstrate service workload and capacity to commissioners.*
- 4) *To have a baseline to demonstrate service efficiency and to benchmark services against others.*
- 5) *To enable campaigning for improved service provision for people with lymphoedema both locally and nationally.*



2. Chronic Oedema

The National Lymphoedema Partnership (NLP) agreed a definition for lymphoedema / chronic oedema (2015a) which reflects international thinking regarding the condition of lymphoedema and the symptom, which is chronic swelling. Chronic oedema is a term used to describe a group of conditions characterised by the presence of swelling within tissues of the body, caused by the accumulation of excess fluid within the interstitial space of the affected area.

The term 'chronic oedema' is traditionally used to describe oedema that has been persistent for at least 3 months to distinguish from acute causes of oedema such as post-operative oedema, acutely decompensated heart failure, infection, burns etc.

Although we are in agreement that the terms chronic oedema and lymphoedema are interchangeable, this subsection recognises the different perspectives across professions. Hence there is some duplication with the general lymphoedema section, but this was agreed to clarify management of simple, non-complex oedema.

CQ1: How should chronic oedema be diagnosed?

Evidence Summary

There was no trial evidence available to answer this question. Chronic oedema is defined as an oedema that is present for 3 months, therefore it is recommended that patients presenting with oedema present for more than 3 months be diagnosed with chronic oedema. Chronic oedema if left untreated can lead to the development of lymphoedema at a later stage, therefore all patients with chronic oedema require prompt assessment and management. Patients may require onward referral to: a nurse-led leg ulcer clinic, to a vascular consultant or to a specialist lymphoedema therapist or appropriately trained healthcare professional for assessment of aetiology and appropriate treatment of their condition. Some services may also have appropriately trained community nurses and practice nurses who may treat these patients in the community.

Investigations which may be carried out to assist in diagnosing the aetiology of lymphoedema:

- Full blood count, renal and liver (especially albumin) function tests may be useful
- Further imaging, such as lymphoscintigraphy or MRI, depending on clinical assessment

Where there is suspicion of venous oedema, the following investigations may be indicated:

- D-dimer ([see HSE guidance](#))
- Compression ultrasonography
- Contrast venography
- Venous duplex ultrasound
- Computed tomography (CT) of the abdomen and pelvis

Recommendations

CQ1.1 Clinicians should diagnose chronic oedema in patients presenting with oedema greater than 3 months, as it will progress to lymphoedema if left untreated.

Evidence Grade: D

Strength of recommendation: Strong

CQ1.2 Further consideration should be given to the aetiology of chronic oedema, and onward referral to other health professionals including specialist lymphoedema therapists, tissue viability nurses, appropriately trained community nurses / practice nurses, GPs or vascular consultants as required.

Evidence Grade: D

Strength of recommendation: Strong

CQ2: Which healthcare professionals should treat simple chronic oedema?

Evidence Summary

Expert opinion recommends that any HCP may treat patients with chronic oedema provided they are appropriately trained to manage this condition. HCPs treating chronic oedema should be proficient in doppler ultrasound assessment, clinical vascular assessment and compression therapy.

Recommendations

CQ2.1 Non-specialist, appropriately-trained healthcare professionals may treat simple chronic oedema. Complex presentations should be referred to a specialist lymphoedema service.

Evidence Grade: D

Strength of recommendation: Strong



Education Need:

There is a need for non-specialist healthcare professionals to be appropriately trained to manage simple chronic oedema.

CQ2.2 Healthcare professionals treating chronic oedema should be proficient in or have access to doppler assessment, clinical vascular assessment and compression therapy.

Evidence Grade: D

Strength of recommendation: Strong

CQ3: How should patients with simple chronic oedema be treated?

Evidence Summary

A recent single-centre, non-blinded RCT (Webb et al., 2020) of patients (n = 183) with chronic oedema of the leg and cellulitis, showed that compression therapy resulted in a lower incidence of recurrence of cellulitis than conservative (education only) treatment. In order for compression therapy to be commenced, arterial compromise must be excluded. Where possible, a doppler assessment, combined with a holistic clinical vascular assessment should be performed prior to commencing compression therapy.

One RCT (n = 36) carried out to assess optimal compression in chronic oedema (Partsch et al., 2011) concluded that for inelastic bandages, the upper limit of compression that should be used is 30 mmHg on the upper limb and 50-60 mmHg on the lower limb.

Reminder: The terms Chronic Oedema and Lymphoedema are interchangeable. Refer to the General Section for comprehensive management guidance on complex chronic oedema / lymphoedema.

Recommendations

CQ3.1 The care of patients should follow a clearly defined two-way care pathway, which should be an integrated specialist and non-specialist pathway, with patients given access to patient support groups.

Evidence Grade: D

Strength of recommendation: Strong

Please see [appendix I.I](#) for the Adult Lymphoedema Pathway and [appendix I.III](#) for the Children and Young People Lymphoedema pathway.

CQ3.2 The goal of simple chronic oedema management should be to alleviate swelling, improve function and minimise complications e.g. cellulitis, ulceration.

Evidence Grade: D

Strength of recommendation: Strong

CQ3.3 Skin care, physical activity and compression therapy should be the mainstays of treatment for simple chronic oedema.

Evidence Grade: D

Strength of recommendation: Strong

CQ3.4 Arterial compromise must be excluded before compression therapy is commenced. ABPI may not be required, depending on the vascular assessment outcome. See question GQ40 for further guidance on ABPI requirements in lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

3. Primary Lymphoedema

Primary lymphoedema is rare, affecting 1 in 100,000 people, however the true incidence is likely significantly underestimated. It may be present at birth or develop later in life, and often occurs as a result of a genetically determined malformation of the lymphatic system. Whilst this can most often be recognised in the periphery (i.e. swelling of the limbs), there may be internal lymphatic dysfunction, for example, chylous reflux causing pleural effusions or ascites (Gordon et al., 2020). Lymphoscintigraphy may be considered during diagnostic assessment.

Primary lymphoedema should be considered an umbrella term to describe several different diseases. Primary lymphoedema was traditionally classified according to age at presentation i.e. congenital (present at birth to 1 year), praecox (from 1 to 35 years), and tarda (occurring after the age of 35 years) (Mandell et al., 1993). However, this old classification system fails to consider the other health problems that may be associated with the type of primary lymphoedema. Recent progress in genetics has provided better understanding of lymphoedema and has contributed to a new classification algorithm (St. George's Classification of primary lymphatic anomalies, see figure 2) which describes sporadic, familial and syndromic forms. It incorporates the known gene mutations, including *VEGFR3/FLT4* (Milroy disease), *VEGFC* ('Primary lymphoedema of Gordon', another type of congenital lower limb primary lymphoedema similar to Milroy disease), *FOXC2* (lymphoedema distichiasis syndrome), *CCBE1* (Hennekam syndrome), and *GATA2* (Emberger syndrome). This classification system sub-groups primary lymphoedema into 5 main categories:

1. Primary lymphoedema associated with other genetic syndromes (e.g. Noonan or Turner syndrome)
2. Lymphoedema associated with systemic involvement (e.g. Hennekam syndrome)
3. Congenital limb lymphoedema (e.g. Milroy disease)
4. Late-onset limb lymphoedema (e.g. lymphoedema distichiasis syndrome or Meige disease).
5. Primary lymphoedema associated with overgrowth disorders and lymphatic malformations.

It is worth noting that certain conditions e.g. Meige disease can present later in adulthood or can be misdiagnosed due to other secondary causative factors e.g. obesity, immobility. Clinicians should also consider primary lymphoedema for patients without a positive family history but with symptoms associated with primary lymphoedema and where other causes have been excluded. Similarly, for those who present with recurrent cellulitis of unclear cause, it can be difficult to decide which came first, as a patient with subclinical primary lymphoedema will be at increased risk of cellulitis due to the associated immune deficiency of the affected limb (Damstra et al., 2008). Meige disease is the commonest form of primary lymphoedema but the underlying genetic cause is not yet known.

Some patients may have a level of lymphatic dysfunction not attributable to other recognisable causes. This may be related to a low level of primary dysfunction not associated with a recognised syndrome. Clinicians should use their clinical judgement and experience to decide which patients should be offered to undergo genetic testing. Genetic testing alone cannot confirm the diagnosis of primary lymphoedema in all patients. Genetic mutations are only detected in 20%-40% of patients with primary lymphoedema seen in specialist primary lymphoedema clinics, so the probability of detecting mutations in the general lymphoedema clinic are likely to be significantly less (Gordon et al., 2020).

The Genetic Assessment Service in Northern Ireland suggests that patients who may be at risk of developing lymphoedema (as a result of a condition which predisposes to it) should receive lymphoedema awareness education including risk reduction advice and information on local referral pathways. In Northern Ireland, NGS panel testing is completed with samples sent to England for analysis.

PLQ1: How should primary lymphoedema be diagnosed?

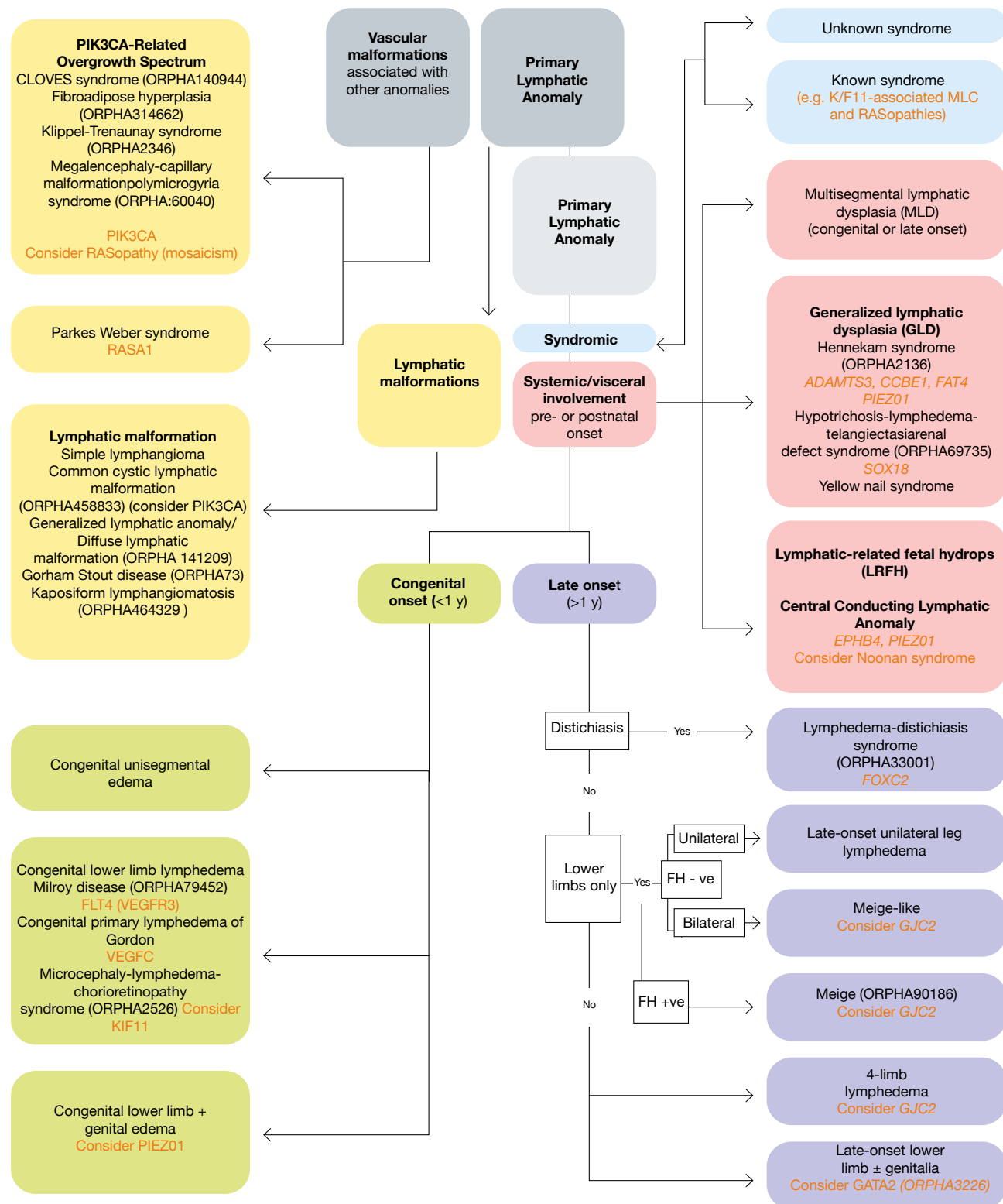


Figure 2. St. George's Classification of primary lymphatic anomalies (Gordon et al., 2021)

Evidence Summary

A review of the literature and an expert opinion document addressed this question. Children with suspected primary lymphoedema should be examined for syndromic characteristics. The St. George's classification algorithm should be used to assess presenting anomalies.

It is worth noting that certain conditions e.g. Meige syndrome present later in adulthood and may be misdiagnosed due to other secondary causative factors e.g. obesity or immobility. Therefore, adults presenting with oedema, and a non-oncological family history of oedema, should be screened using the primary lymphoedema pathway (figure 2).

According to the International Union of Phlebology (IUP) 2013 consensus document (Lee et al., 2013a) "the future of the diagnosis and classification of primary lymphoedema is likely to be determined by the pathophysiology or genetic basis of the underlying condition which would make the present classification (e.g. congenital, praecox or tarda) unnecessary."

VASCERN guidelines on Primary and Paediatric Lymphoedema (VASCERN, 2019) recommend that patients presenting with swelling at birth or persistent swelling for 3 months or more should be assessed and investigated for presence of lymphoedema. VASCERN suggest that a thorough patient history should include the following:

- Age of onset
- Distribution
- Cellulitis
- Systemic involvement
- Warts
- Skin problems
- Segmental overgrowth
- Family history
- Associated problems e.g. congenital cardiac disease
- Venous incompetence
- Previous surgery

A classification system for primary lymphoedema has been proposed which may assist in highlighting which patients require referral for genetic testing (Connell et al., 2010). Children with any of the following features should undergo genetic testing:

- Syndromic Features
- Systemic/visceral involvement (e.g. chylous, pericardial/pleural effusions, ascites, pulmonary/intestinal lymphangiectasia)
- Altered growth
- Cutaneous Features
- Vascular abnormalities
- Congenital onset lymphoedema
- Distichiasis

The International Lymphoedema Framework (2010) recommend that children with lymphoedema and dysmorphic features and/or learning difficulties should be referred for genetic testing.

See appendix II.V for an adapted Paediatric Assessment Form to assist in the assessment of children with lymphoedema.

Recommendations

PLQ1.1 All patients with suspected primary lymphoedema should be assessed using the guideline assessment form ([appendix II.V](#)).

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.2 All children presenting with swelling at birth or swelling for 3 months or more should be assessed and investigated for the presence of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.3 A complete history and clinical assessment may be sufficient to diagnose lymphoedema. Additional investigations may be sought based on individual clinical presentation.

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.4 If required, the choice of investigation depends on the clinical presentation and the resources available.

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.5 Patients with lymphoedema and any of the following features should undergo genetic testing to assess for the possibility of primary lymphoedema:

- Congenital onset lymphoedema
- Syndromic features
- Systemic/visceral involvement (e.g. chylous, pericardial/pleural effusions, ascites, pulmonary/intestinal lymphangiectasia)
- Altered growth
- Cutaneous features
- Vascular abnormalities
- Distichiasis
- Learning difficulties
- Family history of lymphoedema

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.6 If there is suspicion for a genetic aetiology, the St. George's algorithm (figure 2) should be used to direct clinical assessment.

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.7 All patients with primary lymphoedema should be offered genetic testing and counselling.

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.8 Genetic testing results should be shared with St. George's Lymphoedema Service to share information and data recording.

Evidence Grade: D

Strength of recommendation: Strong

PLQ1.9 Clinicians should encourage family and carer involvement where appropriate in the genetic assessment to support the development of new diagnostic modelling.

Evidence Grade: D

Strength of recommendation: Strong

PLQ2: Which genetic tests are indicated for the investigation of primary lymphoedema?

Evidence Summary

The Royal Brompton Hospital offers a single primary lymphoedema 22 known genes panel which can be accessed via all local genetics teams; this has replaced the requirement to request single individual tests thereby reducing cost. However, the genetic tests for Turner and Noonan syndrome are conducted separately.

A small case series (n = 3) concluded that neurological assessment including electroencephalography (EEG) should be carried out on children with generalised lymphoedema and facial involvement to identify generalised lymphoedema associated with neurologic signs (GLANS) syndrome (Berton et al., 2015).

VASCERN guidelines (VASCERN, 2019) recommend that where appropriate, patients should be referred to a genetic specialist for genetic testing.

St. George's Hospital previously published a Paediatric Investigation Pathway for Primary Lymphoedema in Childhood based on expert consensus (CLSIG, 2016) which provides a further breakdown of the genetic tests available:

If a child looks dysmorphic or has learning difficulties:

- Array CGH (detailed chromosome analysis)

Congenital lower limb lymphoedema:

- VEGFR3 for suspected Milroy disease
- KIF11 if microcephaly present
- Turner syndrome if female
- Noonan panel if dysmorphic

Congenital generalised lymphoedema*:

- Consider CCBE1 / FAT4 / PIEZO1 / Noonan gene panel

* hydrops fetalis / chylous effusions / ascites / intestinal lymphangiectasia / pericardial effusions / widespread lymphoedema

Childhood onset of bilateral lower limb lymphoedema (after the age of 1):

- FOXC2 (especially if distichiasis present)
- GATA2 (especially if genital involvement, low monocyte count)
- GJC2 (especially if hands are also swollen)
- Noonan panel (if dysmorphic and other associated features)
- Full blood count; refer to Haematology if any concerns

Multi-segmental lymphoedema with evidence of overgrowth:

- Consider taking a skin biopsy for PIK3CA gene testing

Isolated genital lymphoedema:

- Consider Noonan syndrome (gene panel test)
- Consider anogenital granulomatosis: take skin biopsies of scrotum and/or penis looking for granulomas within the dermis
- Refer to gastroenterology for consideration of endoscopy and biopsy looking for Crohn's disease

Recommendations

PLQ2.1 Genetic testing should be offered to patients in all cases of suspected primary lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PLQ3: Should all patients with primary lymphoedema be offered genetic counselling?

Evidence Summary

The Children's Lymphoedema Special Interest Group (CLSIG) Charter of Care for Children and Young People with Lymphoedema recommend that in cases where a genetic cause is suspected, the child or young person should have access to genetic counselling (CLSIG, 2016). The ILF state that children with inherited forms of lymphoedema should undergo genetic counselling (2010).

Patients with lymphoedema-distichiasis syndrome (LDS) should undergo genetic counselling as it is inherited in an autosomal dominant manner (Mansour et al., 1993).

Patients with Milroy disease (MD) should undergo genetic counselling as MD demonstrates an autosomal dominant pattern of inheritance (Brice et al., 1993). Ultrasonography carried out during pregnancy may detect oedema of the dorsum of the foot, pleural effusions and rarely more extensive oedematous states (e.g. hydrops fetalis). Prenatal testing may be carried out in at-risk families but is rarely requested.

Recommendations

PLQ3.1 Genetic counselling should be offered to any patient with a suspected genetic cause of their lymphoedema, prior to genetic testing.

Evidence Grade: D

Strength of recommendation: Strong

4. Surgery and Lymphoedema

Circumferential Suction Assisted Lipectomy (CSAL) (also known as liposuction) is a well-researched, effective and safe procedure for end-stage lymphoedema that has been unresponsive to conservative treatment (Forte et al., 2019). CSAL changes lymphoedema back into the original state; it produces a long-lasting, 100% volume reduction in limbs when proper compression garments are used post-operatively and for life. CSAL significantly reduces the number of episodes of cellulitis/erysipelas (infection), and dramatically improves quality of life and facilitates self-care. CSAL should be embedded in an integrated lymphoedema service protocol.

While the principle of microsurgery (reconstructing the lymphatic system) is logical, it does not address the reversal of hypertrophied adipose tissue. Therefore, micro surgery is proposed before signs of lymphoedema occur; however, this is controversial since the occurrence of lymphoedema is unpredictable. While microsurgical approaches are developing, further work needs to be undertaken to effectively define indications for such surgery (Hirche et al., 2019).

There are some risks associated with surgery which include but are not limited to:

- Infection
- Bleeding
- Abnormal scarring
- Lymphorrhoea
- Limited improvement of limb volume

A full assessment should be completed by a lymphoedema therapist before any referral to surgery is planned. The therapist is also essential to ensure post-surgical follow up care is provided.

Surgical Options for the treatment of lymphoedema

CSAL / Lymphoedema Liposuction

CSAL differs from cosmetic liposuction in that it enables the surgeon to remove larger volumes of fat than would be expected during cosmetic liposuction surgery. Liposuction is only suitable for a small percentage of lymphoedema patients. In patients with longstanding lymphoedema there are often fatty changes in the limb which become resistant to compression garments and manual lymphatic drainage. Liposuction can remove these fatty deposits permanently. Liposuction may be the most suitable option for patients with a large volume of fatty excess on their limbs who are not suitable for microsurgical interventions (lymphaticovenous anastomosis or lymph node transfer). Following this surgery, compression garments are required lifelong. For more information, see the [National Institute for Health and Care Excellence \(NICE\)](#) guidance on liposuction for chronic lymphoedema (2017 version due update publication in 2022).

Microsurgery

Micro surgical techniques have become increasingly popular in recent years, however they are considered more suitable for very early stage lymphoedema. Some centres have trialled the use of robotic instruments to assist in these microsurgical procedures with preliminary positive outcomes (Gourd, 2020). Lymphaticovenous anastomosis (LVA) is a minimally invasive procedure which diverts lymph into the dermal venous drainage system. According to recent evidence this combined with bandaging and compression garments leads to superior results with minimal lymphoedema at the donor site. Vascularized lymph node transfer (VLNT) is another microsurgical technique, which is often combined with autologous free flap breast reconstruction and it has been shown to improve lymphoedema rates and reduces cellulitis risk. Vascularized lymph vessel transfer (VLVT) consists of harvesting certain lymph vessels, sparing the donor site's lymph nodes. The combination of LVA and VLNT with other methods is thought to maximise their effectiveness (Gasteratos et al., 2021).

Lymphaticovenous Anastomosis (LVA)

Lymphaticovenous Anastomosis (LVA), Lymphovenous Anastomosis and lymphaticovenular anastomosis are interchangeable terms. This recent development in the treatment of lymphoedema aims to improve the underlying malfunction of the lymphatics. The surgery offers patients an opportunity to significantly reduce the amount of time they are required to wear their compression garments and in some cases may remove this requirement completely. LVA has been shown to significantly reduce the risk of cellulitis infections which can exacerbate the lymphoedema (Gennaro et al., 2017, Mihara et al., 2014). LVA is a form of microsurgery which joins lymphatic vessels in the region of 0.3 mm to similar-sized veins using sutures with finer calibre than that of a human hair. This allows the excess lymphatic fluid to drain directly into the venous system. LVA surgery is an option for patients with very early stages of lymphoedema, without any fibrosis and sclerosis of the remaining lymphatic vessels.

Lymph Node Transfer (LNT)

This procedure may be an option for patients who have had lymph nodes removed from their groin or axilla during cancer surgery. LNT surgery moves functioning lymph nodes to replace nodes that have been removed. Nodes are taken from the groin and placed in the axilla or vice versa. The blood vessels that supply the nodes are also transferred and joined to blood vessels in the region of poor drainage. The transferred lymph nodes have been shown to release cytokines (cell signalling chemicals) which encourage old lymphatic pathways to open up and new networks to develop. Over time, the lymphatic vessels from the transferred lymph nodes reconnect to the local vessels (Winters et al., 2021). The surgery also aims to release tight scar tissue in the axilla or groin and improve limb movement.

There are currently no lymphoedema surgical services in Northern Ireland or the Republic of Ireland. At present in the UK, only Scotland and Wales have routine NHS access to LNT surgery. Other hospitals may offer temporary access to surgery, linked to clinical trials. There is a growing number of private clinics offering lymphoedema-related surgeries; such clinics may not require patients to be assessed by a lymphoedema specialist and have undergone a trial of conservative management prior to surgery. This inequity has the potential to raise unrealistic access expectations across UK and Ireland populations, and disrupt agreed surgical pathways.

There are several implantable devices in research development e.g. the FACILISFLOW. A current trial is examining the use of a cyclic vacuum to affect lymphatic pumping pressure and velocity (Moore, 2021). A 2020 systematic review examined quality of life outcomes between lymphoedema treated with surgery versus without surgery (Fish et al., 2020). This review concluded that based on currently available data, such a comparison of outcomes cannot be made with authority.

New surgical developments to reduce the risk of developing lymphoedema

Axillary Reverse mapping

Axillary reverse mapping (ARM) identifies nodes that drain the upper extremity from those draining the breast allowing preservation of the limb lymphatics thereby reducing the risk of lymphoedema. A systematic review found lymphoedema in 0% to 6% of patients undergoing ARM plus SLNB and 5.9% to 24% of patients undergoing ARM plus ALND (Ahmed et al., 2016). Concerns surrounding ARM include reliable and consistent ARM identification rates, crossover lymph nodes / lymphatics (breast SLNB is also ARM node), and feasibility of axillae with heavy tumour burden. The Alliance A221702 trial is currently evaluating SLNB or ALND with and without ARM to formally evaluate the feasibility and utility of ARM (NCT03927027).

Immediate Lymphatic Reconstruction

Immediate Lymphatic Reconstruction (ILR) also known as Lymphatic Microsurgical Preventive Healing Approach (LYMPHA) (Cook, 2021). ILR seeks to identify arm lymphatics in the axillary field and then perform lymphatic to venous anastomoses (LVA) via microsurgical techniques when a competent venous valve is present. Boccardo et al. (2011) published the first series noting lymphoedema in 4% of patients receiving LYMPHA after ALND which increased to 10.5% if they included those patients with transient postoperative lymphoedema. More recently, Feldman et al. (2015) found lymphoedema in 8% of patients at 24 months, which increased to 12.5% when those with transient lymphoedema were included. Collectively, these studies suggest some benefit to immediate lymphovenous anastomosis during ALND; however, further investigation is needed, as the added operative time and need for specialised microsurgical training must be considered if LYMPHA is to be widely adopted for all patients undergoing ALND.



Research Idea:

The effectiveness of axillary reverse mapping (ARM) and the Lymphatic Microsurgical Preventive Healing Approach (LYMPHA) in the management of breast cancer requires further research. The efficacy of LYMPHA in the management of other cancers should also be explored.

SQ1: Is there evidence to support the use of liposuction (circumferential suction assisted lipectomy) in the management of lymphoedema?

Evidence Summary

The most recent systematic review on this topic was published in 2019 by Forte et al. It included 8 original research articles examining the use of lipoaspiration in chronic lower limb oedema. All patients who received lipoaspiration had a volume reduction greater than 50% in their affected lower limb. At four to five years of follow-up, the volume reduction persisted. Patients with secondary lymphoedema had greater reductions in volume when compared to primary lymphoedema. Improvement was found in functionality, quality of life, and rates of infection. Based on these findings the authors recommend lipoaspiration for patients with lower limb lymphoedema (stage II and III) followed by controlled compressive therapy to ensure that volume reduction persists (Forte et al., 2019c).

In 2017, NICE published guidelines (NICE, 2017) on the use of liposuction in chronic lymphoedema. NICE concluded that current evidence supports the use of liposuction in the treatment of chronic lymphoedema, on the basis that its safety and efficacy is well supported in the existing literature. It also states that patient selection must only be done by a multi-professional team as part of a lymphoedema service. Similarly, the International Lymphoedema Framework recommends the use of liposuction in patients with lymphoedema refractory to other interventions in their 2012 document "Surgical Intervention: A position document on surgery for lymphoedema" (Cormier, 2012) when embedded in an integrated lymphoedema service protocol including the follow up and lifelong use of compression garments. The document also noted the significant reduction in infections, dramatic improvements in quality of life, and ability to self-care. The American Society of Breast Surgeons (ASBrS) expert panel also recommends that liposuction and long-term compression is an effective combination approach for severe late-stage BCRL that is refractory to conservative management (McLaughlin et al., 2017b) and as part of a multimodality treatment plan.

The American Venous Forum (2009) recommend that all interventions for chronic lymphoedema should be preceded by at least 6 months of non-operative compression treatment. They also recommend that liposuction be used only in patients with late stage non-pitting lymphoedema, who fail conservative measures (Gloviczki, 2009). While much of the existing literature focuses on liposuction of the limbs in the treatment of post-cancer lymphoedema, a recent small scale RCT (n = 20) examined the use of submental liposuction following head and neck cancer treatment. The authors reported a statistically significant improvement in patients' self-perception of their appearance and subjective scoring of appearance (Alamoudi et al., 2018). As with conservative management, lifelong 24-hour use of compression garments is considered mandatory for maintaining the effect of liposuction (Schaverien et al., 2018).

A small 2019 (Chen, 2019) prospective trial showed a significant decrease in seroma/haematoma formation, contour irregularity and skin necrosis with the introduction of skin excision alongside liposuction.

Recommendations

SQ1.1 Liposuction should be considered as part of a multi-modality treatment plan for people with late stage / chronic lymphoedema where conservative therapy is no longer effective, and with long term monitoring by an established lymphoedema service.

Evidence Grade: A

Strength of recommendation: Strong

SQ1.2 A trial of conservative treatment for at least 6 months should be completed prior to consideration of liposuction/CSAL in patients with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

SQ1.3 In the provision of liposuction/CSAL it is recommended it is provided in a specialist centre and that standard arrangements and service protocols including clinical governance, consent and audit are developed and adhered to.

Evidence Grade: D

Strength of recommendation: Strong

SQ1.4 Lifelong compression therapy must be continued after liposuction/CSAL.

Evidence Grade: D

Strength of recommendation: Strong

SQ2: Is there evidence for the effective use of lymph node transfer (LNT) to manage lymphoedema?

Evidence Summary

A 2021 systematic review and meta-analysis of studies examining the efficacy of LNT addressed this question (Winters et al., 2021). There were 17 studies included in the systematic review and eight studies included in the meta-analysis. This review concluded that current evidence indicates that LNT can significantly improve volume difference (up to 40%) between arms in patients with unilateral lymphoedema. Based on current evidence it also seems likely that LNT has a positive impact on QoL, cellulitis incidence and may lead to reduced need for compression garment use.

A second meta-analysis examined the efficacy of vascularised LNT in reducing limb volume and cellulitis episodes in patients with cancer related lymphoedema (Ward et al., 2021). This metanalysis included the findings of 31 individual studies with a total of 581 patients with cancer related upper or lower limb lymphoedema. Vascularised Lymph Node Transfer (VLNT) led to a significant reduction in UL lymphoedema (above elbow: 42.7% [95% CI: 36.5-48.8]; below elbow = 34.1% [95% CI: 33.0-35.1]). VLNT also led to a significant reduction in lower limb lymphoedema (above knee: 46.8% [95% CI: 43.2-50.4]; below knee: 54.6% [95% CI: 39.0-70.2]).

This study also found that VLNT flaps from extra-abdominal donor sites were associated with greater volume reductions (49.5% [95% CI: 46.5-52.5] versus 39.6% [95% CI: 37.2-42.0], $p < 0.05$) than those from intra-abdominal donor sites. VLNT flaps from extra-abdominal donor sites were also associated with greater reductions in volume than synchronous autologous breast reconstruction/VLNT flaps (32.7% [95% CI: 11.1-54.4], $p < 0.05$). VLNT was also found to reduce the mean number of cellulitis episodes per year by 2.1 episodes (95% CI: -2.7 to -1.4) and it increased Lymphoedema-Specific Quality of Life (LYMQOL) "overall domain" score.

The American Society of Breast Surgeons expert panel recommends that LNT may be effective in the treatment of early secondary BCRL. They recommend patients be assessed by a MDT with an understanding of lymphoedema and receive aftercare with the understanding that surgical treatment is part of a multimodal treatment plan. They also recommend baseline and follow-up assessments be made which include assessments of lymphatic functionality (McLaughlin et al., 2017b). The International Society of Lymphology appears to not endorse LNT in their 2016 consensus document (Health Service Executive, 2016), stating that long-term follow-up data regarding efficacy and risk of harm were lacking at the time.

A 2015 review (Raju and Chang, 2015) of LNT procedures identified 10 studies that examined the effectiveness of LNT in patients with both upper and lower limb lymphoedema. The most common donor sites for lymph nodes were from the submental, supraclavicular, thoracic and inguinal regions. The most common site of transfer was to the nodal basins of the affected upper or lower limb. The reported change in limb volumes ranged from an increase of 13% to a decrease of 64%. The authors concluded that while the results were promising, more evidence was required before this practice could be definitively recommended (Raju and Chang, 2015). For patients with primary lymphoedema, the International Union of Phlebology (IUP) recommend LNT surgery is best utilised in patients with lymphadenodysplasia - clinical stage 2 and 3. However, they caution that LNT remains a controversial procedure, the effectiveness of which has not yet been fully established (Lee et al., 2013a).

Recommendations

SQ2.1 Lymph Node Transfer appears to be effective in the management of lymphoedema.

Evidence Grade: A

Strength of recommendation: Strong

SQ2.2 There is currently not enough high-quality evidence to support the use of Lymph Node Transfer to reduce the risk of developing lymphoedema. Further high-quality research is recommended.

Evidence Grade: D

Strength of recommendation: Strong

SQ3: Does lymphovenous anastomosis (LVA) surgery reduce the risk of lymphoedema?

Evidence Summary

A 2019 Cochrane review (Markkula et al., 2019) assessing the efficacy of surgical techniques in preventing breast cancer-related lymphoedema addressed this question. Two studies focused on the surgical technique of lymphaticovenular anastomosis (LVA). These studies found that patients who undergo LVA have a reduced risk of developing lymphoedema (255 fewer cases per 1,000 women) compared to those who do not. LVA resulted in a reduction in the incidence of lymphoedema compared to non-operative treatment (RR = 0.20, 95% CI 0.06 to 0.63, $P = 0.006$) in these RCTs. Between these studies, statistical variation was low, which increases the reliability of the evidence. However, the two RCTs were conducted in the same centre and did not evaluate any of the secondary outcomes, meaning the evidence is of low certainty. There is low-certainty evidence that LVA is effective in preventing the development of lymphoedema after treatment of breast cancer. Further high-quality RCTs are required to further assess the effectiveness of surgical interventions in the prevention of lymphoedema after breast cancer treatment.

A 2018 systematic review examining the efficacy of prophylactic LVA supported these findings. The review included 16 trials and found that patients treated with prophylactic LVA had a significant reduction in lymphoedema incidence (RR: 0.33; 95% CI: 0.19 - 0.56) compared to no prophylactic treatment ($P < 0.0001$). However, the authors concluded that while these results were promising, there is insufficient high-quality evidence to conclude on the efficacy of prophylactic LVA at this time (Jørgensen et al., 2018).

Recommendation

SQ3.1 At this time, while results are promising, there is not enough high-quality evidence to support the use of LVA to reduce the risk of lymphoedema.

Evidence Grade: A

Strength of recommendation: Strong

SQ4: Is lymphovenous anastomosis (LVA) surgery effective in the treatment of lymphoedema?

Evidence Summary

The International Society of Lymphology appears to endorse LVA in their 2016 consensus document (Health Service Executive, 2016), stating that the procedure is carried out in multiple international sites and long-term follow-up data regarding efficacy and patency are well documented (Health Service Executive, 2016). The American Society of Breast Surgeons (ASBrS) expert panel also recommends that LVA may be effective in the treatment of early secondary BCRL. They recommend patients be assessed by an MDT with an understanding of lymphoedema, and receive aftercare with the understanding that surgical treatment is part of a multimodal treatment plan. They also recommend baseline and follow-up assessments be made which include assessments of lymphatic functionality (McLaughlin et al., 2017b).

A large systematic review examined 17 studies (n = 2,251) including patients who underwent LVA for upper or lower limb lymphoedema or head and neck lymphoedema. This trial reported lymphoedema volume reductions ranging from 2% to 91.7%, with follow-up duration ranging from 8.9 to 120 months (Raju and Chang, 2015). Studies with larger populations (upper and lower limbs) demonstrated better research outcomes. Currently all patients should be fitted for lifelong compression garments post-operatively after LVA.

Recommendations

SQ4.1 LVA may be effective in the treatment of early secondary lymphoedema following node removal in the axilla, groin or head and neck region.

Evidence Grade: A

Strength of recommendation: Strong

SQ4.2 In the provision of LVA, standard arrangements and service protocol including clinical governance, consent and audit should be established.

Evidence Grade: D

Strength of recommendation: Strong

SQ4.3 A multi-disciplinary team should complete patient selection and follow up after LVA, as part of a lymphoedema service.

Evidence Grade: D

Strength of recommendation: Strong

SQ4.4 Consult individual surgeon protocols for guidance on whether there is a requirement for lifelong compression garments post-operatively after LVA.

Evidence Grade: D

Strength of recommendation: Strong

SQ4.5 We recommend further research comparing LVA and conservative treatment.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

Compare the effectiveness of LVA versus conservative treatment of lymphoedema.

SQ5: At what stage of lymphoedema is LVA recommended?

Evidence Summary

Evidence for the optimal time to deliver surgical interventions for lymphoedema treatment is lacking. According to the Dutch guidelines, procedures such as LVA are mainly indicated in early-stage lymphoedema without irreversible changes (stage I-II) (Damstra and Halk, 2017). They further state that reconstructive surgeries such as LNT and LVA are currently not routinely carried out in the treatment of lymphoedema and that non-operative treatment for early stage lymphoedema remains efficacious. The International Society of Lymphology issued similar advice. In their 2016 consensus document they recommend that procedures such as LVA should be performed early in the course of lymphoedema before lymphatic wall damage or impaired lymphatic contractility has occurred (Executive, 2016). The American Venous Forum also suggests that in cases of secondary lymphoedema, microscopic lymphatic reconstruction should be performed early in the course of the disease (Gloviczki, 2009).

Recommendation

SQ5.1 LVA is recommended for early stage lymphoedema, however non-operative treatment for early stage lymphoedema remains effective.

Evidence Grade: D

Strength of recommendation: Strong

SQ6: Is there evidence that sentinel lymph node biopsy (SLNB) compared to ALND reduces the risk of developing lymphoedema?

Evidence Summary

A Cochrane review (2017) addressed this question (Bromham et al., 2017). Three RCTs in patients (n = 1,965) undergoing treatment for breast cancer were included in this analysis. Lymphoedema, defined by increase in arm circumference, was less likely after SLNB compared to Axillary Lymph Node Dissection (ALND) (OR 0.04- 0.60). The authors concluded that low-quality evidence suggested that compared to SLNB or no axillary surgery, patients treated with ALND are at greater risk of developing lymphoedema. Based on this evidence, it is expected that for every 1,000 patients treated with ALND, 132 experience lymphoedema at one-year post-surgery, versus 22-115 of those receiving SLNB. Compared to SLNB, ALND is associated with more long-term negative outcomes such as reduced arm movement, pain, and paraesthesia.

A 2015 systematic review (Li et al., 2015) examined the outcomes of SLNB alone versus ALND in patients with early breast cancer and sentinel node metastasis and found that patients who underwent ALND had greater risk of lymphoedema (RR 0.28, 95% CI 0.20-0.41; p < 0.01) than those had SLNB alone. It may therefore be inferred that SLNB reduces this risk of developing lymphoedema in this cohort. Similarly, a 2013 systematic review (DiSipio et al., 2013b) of 18 studies examining risk factors associated with lymphoedema development in patients with breast cancer found that those who underwent ALND had a greater risk of developing lymphoedema at 2 years post-op than those who underwent SLNB (19.9% compared to 5.6% respectively).

Similar findings have been reported in other patient cohorts. A 2017 systematic review (Huang et al., 2017) examining risk factors for developing lower limb oedema in patients with

Evidence Summary Cont.

vulvar cancer found that those who underwent SNBL had a much lower rate of lower limb lymphoedema compared to pelvic lymph node dissection (5.9% versus 32.1% respectively). There was a lack of evidence examining SLNB in other cancers.

Additionally, results from a long term prospective screening trial found the rate of lymphoedema in a group treated with ALNB alone was 29% compared to 7.7% in the group treated with SNLB alone (Naoum et al., 2020).

Recommendations

SQ6.1 SLNB is recommended where possible in preference to regional lymph node clearance to reduce the risk of lymphoedema in patients undergoing certain surgeries for cancer treatment.

Evidence Grade: A

Strength of recommendation: Strong

SQ6.2 Further research is recommended to assess the efficacy of SLNB in reducing the risk of lymphoedema in all types of cancer surgery.

Evidence Grade: D

Strength of recommendation: Strong



Research Idea:

The efficacy of SLNB in reducing the risk of lymphoedema in all types of cancer surgery should be further evaluated.

SQ7: What is the role of compression therapy in the post-surgical phase?

Evidence Summary

There was limited evidence to answer this question. Each surgical procedure has a different role for post-op compression therapy. For example liposuction will require life-long compression post-surgery (Schaverien et al., 2018). The following are based on expert consensus available in surgical patient information leaflets:

- LVA aims to improve the underlying malfunction of the lymphatics. The surgery offers patients an opportunity to significantly reduce the amount of time they are required to wear their compression garments and in some cases may remove this requirement completely.
- With LNT, lymphoedema improves slowly over 3 years. During this time, both normal lymphoedema reviews and compression garments are maintained. After this period, it may be possible to reduce the time in garments or stop wearing them altogether, depending upon the final surgical outcome.

Recommendations

SQ7.1 Compression therapy post lymphoedema surgery should be initiated as per surgeon specific protocol, and based on type of surgery.

Evidence Grade: D

Strength of recommendation: Strong

SQ7.2 Lifelong compression therapy is essential after liposuction as a treatment for lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

5. Oncology-related lymphoedema

Lymphoedema is a recognised complication related to cancer and its treatments e.g. lymph node removal, and/or radiation therapy (Gillespie et al., 2018). It can also be caused by the tumour itself obstructing the lymphatic system. The prevalence is variable depending on the cancer location and stage of disease. The estimated prevalence, the number of new patients diagnosed per year and the estimated number of potential patients affected by lymphoedema per year is summarised in Tables 10 and 11.

Table 10. Estimates of oncology related lymphoedema (Republic of Ireland)

Malignancy	Prevalence %, as suggested by research, likely to develop lymphoedema	Average no. of new cases per year (National Cancer Registry Ireland 2018)	Research suggested estimated no. of new lymphoedema patients in Ireland (from 2018)
Breast	12%-25%	3,516	422-879 (median 651)
Gynaecological	33%	3,602	1188
Melanoma	20%	1,092	218
Prostate	10%	3,474	347
Bladder	10%-20%	471	47-94 (median 71)
Head and neck	62%-75.3%	1,400	910-1,054 (median 982)
Total		13,555	3,457

Table 11. Estimates of oncology related lymphoedema (Northern Ireland)

Malignancy	Prevalence %, as suggested by research, likely to develop lymphoedema	Average No. of new cases per year (Cancer Registry 2013-2017)	Research suggested estimated no. of new lymphoedema patients in Northern Ireland (from 2019))
Breast	12%-25%	1,398	167.76- 349.5 (median 258.63)
Gynaecological	33%	647	213.51
Melanoma	20%	451.58	90.32
Prostate	10%	1,133	113
Bladder	10%-20%	219	21.9 – 43.8 (median 32.85)
Head and neck	62%-75.3%	328	203.36 – 246.98 (median 225.17)
Total		4,176.58	933.48

The 2018 Welsh and LNNI caseload breakdown states that 40% of the lymphoedema caseload is secondary cancer related lymphoedema.

Sentinel lymph node biopsy (SLNB) has reduced the number of nodes being removed in breast surgery therefore reducing the risk of upper limb lymphoedema. This is recorded as being the first line procedure in 77% of cases (2017, BHSCT data); of this, 7% required an additional follow up axillary node clearance (ANC) post SLNB. Recently there have been reports of a new trend for breast oedema with ANC surgery. Radiotherapy treatment can also damage nodes. There is an increase in the use of neo-adjuvant chemotherapy, which may impact on future anticipated risk. Longer duration of neo-adjuvant chemotherapy (e.g. taxane-based therapy) in the treatment of breast cancer has been associated with increasing lymphoedema incidence (Armer et al., 2019). These findings underscore the importance of prospective surveillance and evaluation of both limb measurements and symptom assessment.

Prehabilitation

In 2019, the Department of Health (United Kingdom) and Macmillan Cancer Support, 2020 launched new prehabilitation guidelines (Support, 2020). People with cancer who have poor physical, nutritional and/or mental health are known to have fewer cancer treatment options available, be more vulnerable to the adverse effects of cancer treatments (including lymphoedema), and have worse long-term health prospects irrespective of cancer type and stage of disease (Naughton and Weaver, 2014). Prehabilitation enables people with cancer to prepare for treatment through assessment, needs-based prescribing for healthy behaviour, tumour specific education, public health initiatives, and follow-up.

Prehabilitation is relevant for many tumour groups providing risk reduction guidance, onward referral pathways, and first line management skills for some of the consequences of cancer, such as lymphoedema. A second aim is to introduce activities to address existing health disparities including reduced levels of activity and poor weight management control. Prehabilitation and rehabilitation are key components of the pathway of care from diagnosis to survivorship, and focus on enablement and empowerment.

The Northern Ireland Cancer Strategy draft prehabilitation recommendations include:

- All patients with a cancer diagnosis will have access to prehabilitation and, where required, continuation to rehabilitation.
- **Workforce** - Adequate numbers of prehabilitation/rehabilitation staff, and support team, across all tumour sites, and in all relevant care settings.
- **Resources** – Technology will be available to support prehabilitation including lymphoedema screening and surveillance), rehabilitation communication, assessment, and processes.

Alert Box!

Clinicians who assess patients with a history of a cancer diagnosis who present with a late onset, acute and severely swollen limb, should consider potential malignancy progression and refer back to oncology for urgent review.

Risk of lymphoedema after treatment of cancer is covered in detail in the general section on [risk reduction and awareness](#).

OQ1: Does prehabilitation reduce the risk of developing lymphoedema for patients undergoing surgical cancer treatment?

Evidence Summary

There is a paucity of evidence available to answer this question. The role of 'prehabilitation' in improving postoperative outcomes has been demonstrated in several cohorts of patients living with cancer including breast, colorectal and head and neck cancers (Shun, 2016). Expert opinion recommends that prehabilitation should be a component of the patient care pathway in patients living with cancer and may assist in reducing lymphoedema incidence.

Recommendations

OQ1.1 Prehabilitation should be a component of the care pathway in all patients diagnosed with cancer.

Evidence Grade: D

Strength of recommendation: Strong

OQ2: Does education reduce the risk of developing lymphoedema in at-risk patients living with cancer?

Evidence Summary

Patient education reduces BCRL risk and associated symptoms (Fu et al., 2008, Lu et al., 2015, Basen-Engquist et al., 2006) probably because of risk-reducing lifestyle changes such as exercise and weight loss. A prospective randomized trial demonstrated significantly lower rates of BCRL with education and active intervention compared with education only (Torres Lacomba et al., 2010). One further small prospective study (n = 180) found the degree or duration of lymphoedema was lower in patients that had been educated about lymphoedema compared to the patients who had not been, but the difference was non-significant (Borman et al., 2017).

The National Comprehensive Cancer Network (NCCN) Breast Cancer Panel recommendations stress the importance of including lymphoedema education as a central component of long-term follow-up care for patients living with cancer. They emphasise the crucial role of patient education in establishing risk reducing behaviours and promoting early self-detection of deterioration. When these activities are combined with prompt interventions, they can have significant positive impact on patient outcomes and quality of life. The goals of patient education are to raise awareness of the risk of developing lymphoedema, particularly in the 3-5 years post-op. Clinicians should also highlight to patients the early signs and symptoms that precede visible swelling: unilateral/ipsilateral aching, stiffness, heaviness, fullness, or tightness. They should inform patients that if clothing or jewellery become tight, this may be further evidence of evolving lymphoedema.

In their guideline, an expert panel from The American Society of Breast Surgeons recommends that "surgeons should admit and accept that lymphoedema risks exist and educate themselves and their patients about these risks at preoperative and follow-up visits. Education should continue into survivorship and be incorporated into survivorship care plans." (McLaughlin et al., 2017c).

Recommendations

OQ2.1 All patients who are at risk of developing lymphoedema post treatment, should be educated in risk reduction strategies and early signs and symptoms of lymphoedema development.

Evidence Grade: D

Strength of recommendation: Strong

OQ2.2 Education should be combined with active intervention to reduce risk of cancer-related lymphoedema development, for example during prehabilitation.

Evidence Grade: D

Strength of recommendation: Strong

OQ2.3 The wider MDT should accept that lymphoedema risks exist and educate themselves and their patients about the risks of developing lymphoedema at preoperative/pre-treatment and follow-up visits.

Evidence Grade: D

Strength of recommendation: Strong

OQ2.4 Education should continue into survivorship and be incorporated into survivorship care.

Evidence Grade: D

Strength of recommendation: Strong

OQ2.5 Education should include referral pathways to specialist lymphoedema services.

Evidence Grade: D

Strength of recommendation: Strong

OQ3: What is the impact of surveillance / early detection programmes on the development of lymphoedema in at-risk patients living with cancer?

Evidence Summary

There has been a great shift in lymphoedema surveillance over the last decade, with increasing emphasis now placed on identifying early-stage or subclinical lymphoedema (volume changes of 5% to 10%). The reason for this is because an early-stage diagnosis offers the best opportunity for early intervention and treatment (Bar Ad et al., 2010, Torres Lacomba et al., 2010, Lahtinen et al., 2015, Johansson and Branje, 2010). Additionally, data suggest that the early identification and surveillance strategies are more cost effective than waiting for the overt evidence of lymphoedema to present (Stout et al., 2012). Two ongoing trials are assessing the impact and importance of subclinical lymphoedema.

Prospective surveillance models (PSM) have been developed to assist in the early detection of lymphoedema leading to earlier and more efficacious treatment. PSMs typically include a pre-op assessment which should include baseline limb volume and functional mobility measurements and a regularly scheduled follow-up protocol e.g. 3 month intervals for the first year and less frequently thereafter (Yang et al., 2016). To identify meaningful change associated with sub-clinical onset of lymphoedema, typical assessments at follow-up appointments include:

- Psychosocial support
- Reassessment of limb volume
- Reassessment of functional mobility

Evidence Summary (cont.)

Sub-clinical lymphoedema is identifiable at low diagnostic thresholds (3% to 5% limb volume change from baseline in a swelling not due to weight change) by measuring both limbs. Initially sub-clinical lymphoedema can present in just one segment of a limb. Early identification may also offer the opportunity to provide treatment options only suitable for early stage lymphoedema e.g. lymphatic-venous shunts.

A systematic review (Shah et al., 2016a) identified two small (n = 185) randomised trials which demonstrated that early intervention was efficacious in reducing the rate of BCRL (> 50% reduction). Early interventions included physiotherapy and manual lymphatic drainage. The findings of these studies were confirmed with larger prospective and retrospective cohorts. There are currently two further trials (n = 1,280) being carried out in this area. The current body of evidence appears to support the development of surveillance programmes aimed at early detection and treatment of BCRL.

International guidance recommends that ideal detection tools for subclinical lymphoedema should be objective and reproducible, providing standardised metrics that could be used to justify treatment decisions. It is recommended that for surveillance purposes, an initial pre-op measurement should be followed by regular measurements for 3–5 years. Currently, available research findings do not standardise early interventions or provide long-term follow-up to allow us to establish treatment pathways. Longer term outcome trials are required to establish if favourable outcomes are associated with early lymphoedema detection at subclinical or early clinical stages.

Early Detection

A systematic review (Shah et al., 2016a) examined 13 original research articles assessing early detection and surveillance of BCRL. Several studies included in the review demonstrated that modern diagnostic modalities bioimpedance spectroscopy (BIS) and perometry have increased sensitivity, which may allow for the earlier detection of BCRL. Existing international guidance, however, does not at this time recommend any of these techniques over one another, citing a lack of robust evidence to support their validity. The American Society of Breast Surgeons advocate for the importance of early detection and surveillance however they state that all detection tools have advantages and disadvantages and as such they cannot recommend any specifically. The expert panel recommend lymphoedema assessment should include comprehensive subjective and objective elements (McLaughlin et al., 2017c, 2016).

Recommendations

OQ3.1 Screening and surveillance should be established for all identified at-risk oncology patient populations.

Evidence Grade: A

Strength of recommendation: Strong

OQ3.2 Patients undergoing treatment for cancer which put them at risk of developing lymphoedema, should have pre-treatment measurements (e.g. tape measurements, BIS) taken to establish their baseline.

Evidence Grade: D

Strength of recommendation: Strong

OQ3.3 Surveillance should include a pre-op assessment which should include functional mobility measurements. These patients should have regular surveillance follow-up. See the surveillance pathway in appendix [I.IV](#)

Evidence Grade: D

Strength of recommendation: Strong

OQ3.4 As there is currently no evidence to recommend one early detection tool over another, clinicians should use tools as per their preference and availability. Examples of tools which may assist in early detection include BIS and perometry. Assessment should include both subjective and objective components.

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Pre-treatment baseline measurements should be recorded alongside other vital signs in each patient's medical record.

OQ4: What is the recommended period of surveillance for cancer-related lymphoedema?

Evidence Summary

There is a paucity of trial evidence available to answer this question, however expert consensus documents advocate for surveillance at baseline (pre-op), surgical review (6-8 weeks post-op), 9 months, and thereafter at 1, 1.5, 2, 2.5 and 3 years. Relative volume change or bioimpedance spectroscopy (BIS) should be undertaken at these points. In cases where measurements have returned to baseline and are maintained over a 3-6 month period, the patient may be weaned off compression garment use by gradually reducing the number of days per week they are worn. Earlier expert consensus recommended cancer related lymphoedema surveillance for up to 2 years however recent consensus documents have recommended surveillance for 3 years (McLaughlin et al., 2020).

Recommendations

GQ4.1 Patients at high risk of cancer related lymphoedema should be on the screening and surveillance pathway and should be monitored at baseline (pre-op), surgical review (6-8 weeks post op), 9 months, and thereafter at 1, 1.5, 2, 2.5 and 3 years. Relative volume change, TDC or BIS should be undertaken at these points to assess for lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

OQ5: How should cancer related lymphoedema be diagnosed?

Evidence Summary

The diagnosis of lymphoedema is challenging, particularly in the early stages, with varying definitions and a wide range of diagnostic methods and tools available. There are no robust comparison trials validating one technique over another, hence no gold standard of lymphoedema measurement exists. None of the existing guidelines recommend a particular tool, device or method of measurement as there is a lack of evidence to support one tool or method of measurement over another (McLaughlin et al., 2017c). BCRL assessment tools include: circumferential measurement (CM), water displacement, bioelectrical impedance spectroscopy (BIS), perometry, tonometry, and patient self-report tools. Existing guidelines suggest that circumferential tape measurement is the minimum acceptable standard, provided non-stretch tape measures are used and measurements taken at several points on both arms. A 2 cm increase in measurement of limb circumference is likely the most common definition of lymphoedema used (Armer et al., 2013b). Others recommend calculating limb volumes using the formula of a truncated cone (Taylor et al., 2006). The NLN and ISL state that the following tools can assist in the early detection of subclinical lymphoedema: infrared perometry, tissue dielectric- and bioimpedance-spectroscopy (BIS). These bodies recommend that the above measures may also be superior methods of lymphoedema measurement, with higher specificity and sensitivity compared to circumferential tape assessment (McLaughlin et al., 2017c).

The American Society of Breast Surgeons expert panel state that every measurement method and tool has both advantages and disadvantages. A selected measurement method should be considered as part of a comprehensive measurement strategy, which includes a combination of objective and subjective assessments (McLaughlin et al., 2017b). This combination of assessment techniques has been shown to optimise lymphoedema diagnosis (Ostby et al., 2014, Armer et al., 2003). The International Society of Lymphology (ISL), the National Lymphoedema Network (NLN), the National Accreditation Programme for Breast Centres, and the National Comprehensive Cancer Network (NCCN) recommend pre-op assessment and ongoing surveillance of both arms at regular intervals (Armer et al., 2013a). None of these bodies recommend one particular technique over another, as there is yet to be an agreed upon gold standard screening method. The ISL recommend clinicians always consider other potential causes of unilateral limb lymphoedema. The differential diagnosis of which should include: lymphomas, solid organ tumours (primary or metastatic), soft tissue sarcomas and thrombotic events such as DVT. To exclude these aetiologies, a thorough medical evaluation should be carried out in each patient presenting with signs or symptoms of lymphoedema.

The International Society of Lymphology (ISL) recommend that most cases of lymphoedema can be diagnosed by history and clinical exam alone. They recommend that imaging only be used in cases where diagnosis is unclear or requires superior definition for therapeutic and/or prognostic purposes. The diagnosis of lymphoedema is challenging, particularly in the early stages with definitions varying and a wide range of objective methods and tools available (McLaughlin et al., 2017a).

Breast Cancer

The Breast Cancer EDGE Task Force (Perdomo et al., 2014a) recommends circumferential arm measurement, water displacement, and BIS as assessment tools for early detection of BCRL and to monitor response to treatment. Owing to its lack of clinical utility, perometry cannot be recommended at this time. Further research is also required to standardise the diagnostic criteria to detect early-onset BCRL.

Evidence Summary (cont.)

Head and Neck Cancer

There is no standardised set of diagnostic criteria for lymphoedema in patients with head and neck cancer, hence accurate and timely diagnosis is challenging (Cohen et al., 2016b). Methods that have been described in this population include:

1. Tape measure
2. Ultrasound
3. Endoscopic evaluation of mucosal oedema
4. Photographs

Clinicians unfamiliar with these methods of diagnosis should consider referral to a specialist.

Urogenital Cancer

The Urogenital Cancer EDGE Task Force (Cohn et al., 2017b) recommends circumferential limb measurement and water displacement measurement for use as reliable methods to assess lower limb volume as part of lymphoedema diagnosis, as well to monitor change of volume in patients treated for urogenital cancers. There is no “index” limb with which to compare in this cohort. There appear to be no studies validating methods of genital lymphoedema volume measurement.

Recommendations

QQ5.1 There is currently insufficient evidence to recommend a gold standard diagnostic device for the diagnosis of lymphoedema in patients living with cancer.

Evidence Grade: D

Strength of recommendation: Strong

QQ5.2 Patients who have undergone treatment for cancer may not require the same level of investigation to determine the aetiology of their swelling.

Evidence Grade: D

Strength of recommendation: Strong

QQ5.3 The choice of measurement tool depends on clinician preference and availability.

Circumferential tape measurement is the minimum acceptable standard, provided non-stretch tape measures are used and measurements are taken at several locations on both arms. Other acceptable methods include: water displacement, BIS, tonometry, patient self-report tools, infrared perometry and TDC.

Evidence Grade: D

Strength of recommendation: Strong

QQ6: With patients treated for cancer, is the use of imaging required for the diagnosis of lymphoedema?

Evidence Summary

The role of diagnostic imaging is mainly that of assisting in the assessment of unclear swelling or arm enlargement after breast cancer or other cancer surgery (McLaughlin et al., 2017b). There is a number of imaging modalities currently available to assist in the diagnosis of lymphoedema (Bernas et al., 2018). No single modality has been shown to be superior to another and none of the international lymphoedema bodies recommend one method of diagnosis over another. Most international guidance sources recommend that subjective assessment of patient symptoms as well as objective measurement of the patient’s limb is the optimal method of diagnosis (McLaughlin et al., 2017a).

Lymphoscintigraphy

Lymphoscintigraphy involves injection of a radioactive tracer dye to demonstrate dilation of the collecting lymphatics prior to swelling becoming clinically apparent. This method enables differentiation between lymphatic swelling from other causes of limb enlargement and as such is considered the gold standard for radiological assessment of lymphatic pathology (McLaughlin et al., 2017a). Lymphoscintigraphy can predict success or failure of CDT (Hwang et al., 2007). Lymphoscintigraphy should only be carried out when swelling aetiology is unclear or patients are unresponsive or poorly responsive to standard treatment.

Tissue Dielectric Constant

Measurement of tissue dielectric constant (TDC) is a technique that quantitatively measures local total tissue water content, both intracellular- and extracellular fluid in the skin and subcutaneous tissue. The technique has been validated experimentally on skin phantoms. Clinical studies on healthy subjects have demonstrated good intraobserver and interobserver validation. It has been demonstrated that TDC values are influenced by anatomical measurement site, measurement depth and subject sex, body mass index (BMI) and age. TDC measurements have been applied successfully in clinical studies evaluating oedema of varying aetiologies including oedema-changes in: skin irritation, skin irradiation, haemodialysis, post-op patients following cardiac surgery, breast-cancer-related arm lymphoedema and lymphoedema of the lower extremities.

Venous occlusion plethysmography

Venous occlusion plethysmography also shows that total arm blood flow is increased in BCRL (Stanton et al., 1998).

Indocyanine green fluorescence

Indocyanine green (ICG) fluorescence has been demonstrated as superior to standard lymphoscintigraphy in the diagnosis of early lymphoedema (Mihara et al., 2012). It identifies the location of lymphatics and dermal backflow and provides a dynamic functional assessment. Currently, ICG fluorescence lacks the quantification feasible for radionuclide lymphoscintigraphy (McLaughlin et al., 2017a).

Non-invasive imaging

Non-invasive imaging e.g. MRI, CT, and single-photon emission CT (SPECT) can define and detect early lymphoedema changes e.g. honeycomb distribution of fluid within epifascial planes, sub-fascial compartment fluid, and the absence of oedema within muscle tissue (Vaughan, 1990).

Recommendations

QQ6.1 Imaging is typically not required in the diagnosis of lymphoedema following cancer treatment.

Evidence Grade: D

Strength of recommendation: Strong

QQ6.2 Measurement modalities such as TDC may be used for patients with lymphoedema where there is difficulty with measurement e.g. head and neck, genital and breast lymphoedema.

Evidence Grade: B

Strength of recommendation: Strong

QQ7: Does lymph node dissection and/or radiation treatment increase the risk of lymphoedema development?

Evidence Summary

A large review of patients given pelvic radiation mono- or adjuvant-therapy found a varying impact of radiation on the development of lower limb lymphoedema (Lindqvist et al., 2017). Several studies reported that radiation treatment significantly increased the risk of lower limb lymphoedema (Ryan et al., 2003, Biglia et al., 2015, Todo et al., 2015, Tanaka et al., 2007, Tada et al., 2009, Todo et al., 2010, Yost et al., 2014, Rowlands et al., 2014, Bae et al., 2016, Kim et al., 2015). However, a small number of studies failed to find a statistically significant association (Achouri et al., 2013, Kitchener et al., 2009, Abu-Rustum et al., 2006).

In an older study (Tada et al., 2009) of patients with ovarian and uterine cancer who underwent pelvic lymph node dissection, post-operative radiotherapy (OR: 1.79; 95% CI: 1.20-2.67; $p = 0.006$) was statistically significantly associated with lymphoedema prevalence. Radiation has been consistently linked to increased risk of BCRL, especially additive regional nodal irradiation (Kim et al., 2016, Hayes et al., 2008, Kilbreath et al., 2016, Donker et al., 2014, Ozcinar et al., 2012, Ashikaga et al., 2010). The American Society of Breast Surgeons (ASBrS) expert panel recommend that clinicians should “question the routine use of post-mastectomy or regional node irradiation” (McLaughlin et al., 2017c). It is worth noting, however, that when compared to axillary LND, results from the AMAROS trial showed lymphoedema was noted significantly more often after axillary LND compared to axillary radiotherapy at 1, 3, and 5 years (Donker et al., 2014).

Recommendations

QQ7.1 LND may increase the risk of lymphoedema development and clinicians, and patients receiving LND should be made aware of this.

Evidence Grade: A

Strength of recommendation: Strong

QQ7.2 Radiation may increase the risk of lymphoedema development and patients receiving radiation treatment should be made aware of this.

Evidence Grade: A

Strength of recommendation: Strong

QQ7.3 Clinicians should be aware that LND may be associated with a higher risk of developing lymphoedema compared to radiation treatment and should be conscious of this when risk stratifying patients.

Evidence Grade: A

Strength of recommendation: Strong

QQ8: Does axillary web syndrome increase the risk of lymphoedema development?

Evidence Summary

A large ($n=1,181$), recent, prospective study of patients who developed axillary web syndrome (also known as lymphatic cording) post-treatment for breast cancer had 2.4 times the probability (odds ratio = 2.40; 95% confidence interval = 1.40-4.11; $P= 0.002$), of developing BCRL compared to patients without cording. Since patients with evidence of lymphatic cording are at higher risk of BCRL, evidence of such should be incorporated into BCRL risk stratification (Brunelle, 2020).

By contrast, older, long-term prospective trials (Wariss et al., 2017, Ferreira et al., 2018) examined the incidence of lymphoedema following axillary surgery and found no apparent increased risk of developing lymphoedema between those diagnosed with axillary web syndrome and those who were not. A further smaller-scale study found axillary web syndrome was not associated with an increased risk of lymphoedema development.

Recommendations

QQ8.1 As there is emerging evidence that patients with axillary web syndrome are at higher risk of developing lymphoedema, additional vigilance is required in this cohort of patients.

Evidence Grade: C

Strength of recommendation: Strong

QQ8.2 Patients reporting symptoms of cording should be referred to their local oncology physiotherapist for management.

Evidence Grade: D

Strength of recommendation: Strong

QQ9: Does breast reconstruction surgery impact lymphoedema development in patients treated for breast cancer?

Evidence Summary

A large systematic review and meta-analysis addressed this question (Siotos et al., 2018). This review examined the association between breast reconstruction (BR) and the development of lymphoedema. In total 19 articles ($n = 7,501$) were included in the meta-analysis. Breast reconstruction was significantly associated with lower risk of developing lymphoedema ($p < 0.001$) compared to mastectomy-only or breast-conserving surgery. Between patients who received implant-based or autologous BR, rates of lymphoedema were not significantly different.

Recommendations

QQ9.1 Clinicians should be aware that breast reconstruction surgery does not appear to be associated with an increased risk of BCRL development and may reduce the risk of BCRL. Patients should consult with their surgeon regarding their own individual risk.

Evidence Grade: A

Strength of recommendation: Strong

OQ10: What is the evidence supporting the use of Complete Decongestive Therapy (CDT) for cancer patients?

Evidence Summary

Complete decongestive therapy (CDT) is the accepted standard of care for secondary lymphoedema in most developed countries (McLaughlin et al., 2017a). CDT involves manual lymphatic drainage (MLD), gradient compression bandaging, exercise, and dedicated skin care. Trials have shown CDT has been shown to be beneficial in the control of limb volume, fibrosclerotic tissue changes, infection risk and reducing disability burden. Subclinical lymphoedema is not treated with CDT, and efforts in this cohort of patients should focus on surveillance measures, with discretionary use of compression therapy. The use of MLD may be considered as a prophylactic measure. Stages 1-3 of lymphoedema progression should be treated with CDT as a first line measure. BCRL can be effectively managed with CDT in any stage, however some studies have suggested that CDT may be more effective in early-stage lymphoedema (Smile et al., 2018). Conversely, other trials have shown successful outcomes using CDT even at advanced stages of lymphoedema (Committee. 2011).

A review of available evidence concluded that owing to the heterogeneity of study designs and measurement methods, the evidence available is insufficient to recommend any specific CDT protocols, intensity or frequency of treatment at this time (Lasinski et al., 2012). A systematic review examining complementary and alternative therapies to CDT (e.g. laser, acupuncture, electrical stimulation and dietary changes) concluded that there is not enough robust evidence to recommend these therapies as CDT alternatives (Rodrick et al., 2014). A systematic review of current evidence examining the effectiveness of CDT in the treatment of early BCRL (defined as duration of symptoms < 1 year) yielded 7 studies. There was weak evidence supporting the use of CDT in the treatment of early BCRL. The authors concluded that the available evidence was insufficient to allow conclusions to be drawn regarding the most efficacious treatment for this cohort (Jefferis et al., 2018a).

A systematic review of the treatment of lymphoedema in patients following treatment for head and neck cancer identified 10 studies examining the efficacy of MLD and CDT in this cohort (Tyker et al., 2019). Seven of these studies were retrospective in nature and 6 out of these 7 studies showed that lymphoedema measurements decreased significantly at follow-up (3-12 weeks). Almost all of these studies have found a significant decrease in head and neck lymphoedema (HNL) after therapy, and the remaining studies demonstrate a non-significant decrease, suggesting CDT may be the most effective first-line treatment for HNL. However, it is worth considering the heterogeneity in treatment strategies defined as CDT in these studies, which makes it difficult to draw conclusions as many do not fully describe what physical therapy techniques were used. CDT is recommended as first line treatment in patients with BCRL by a number of international guidance documents (NICE, 2014, McLaughlin et al., 2017a, 2016).

Recommendations

OQ10.1 CDT has been shown to be effective in the treatment of lymphoedema for patients who have undergone treatment for cancer. See the [general section](#) of this guideline for specific guidelines on CDT treatment.

Evidence Grade: A

Strength of recommendation: Strong

OQ11: What is the evidence of safety and efficacy of CDT (Complete Decongestive Therapy) for cancer patients with advanced disease?

Evidence Summary

There is a paucity of evidence available to answer this question. MLD was previously thought to be contraindicated in patients with metastatic cancer, based on a fear that it may promote metastatic spread. An older review article postulated that CDT does not contribute to spread of cancer and hence should not be avoided in patients with metastatic disease (Godette et al., 2006). The authors argue that this is true based on our theoretical understanding of the "optimal microenvironment" required for metastasis to be established in distant sites.

There are very few studies examining manual lymphatic drainage in active cancer. The majority of these trials report no increased risk of spread in patients with metastatic cancer or active disease. However, these studies are of low quality (Pinell et al., 2008, Mena Flor, 2009) and therefore conclusions regarding the safety and efficacy of CDT in patients with advanced cancer cannot be drawn at this time.

NICE guidance on the management of lymphoedema in patients with advanced breast cancer (stage 4) recommends offering CDT as first-line management (NICE, 2014).

Recommendations

OQ11.1 Based on current available evidence, CDT is not contraindicated in metastatic disease.

Evidence Grade: C

Strength of recommendation: Strong

OQ11.2 Patients for whom there is a suspicion of metastatic disease should be referred to oncology services for assessment prior to commencement of CDT.

Evidence Grade: D

Strength of recommendation: Strong

OQ11.3 For patients with metastatic disease, the decision to commence CDT should involve MDT consultation.

Evidence Grade: D

Strength of recommendation: Strong

6. Lymphoedema in children and young people (CAYP)

Lymphoedema in children is rare, with an estimated prevalence of 1.15 cases per 100,000 persons aged < 20 years in a 1985 study (Mendez and Opitz, 1985, Smeltzer et al., 1985). Recent progress in genetics has provided better understanding of lymphoedema and has led to a new classification, with sporadic, familial and syndromic forms. These include gene associations, especially FLT4 (Milroy disease), FOXC2 (lymphoedema distichiasis syndrome), VEGFC (Milroy-like syndrome), CCBE1 (Hennekam syndrome), GATA2 (Emberger syndrome), SOX18 (hypotrichosis-lymphoedema-telangiectasia) and GJC2 (Meige syndrome) (Vidal et al., 2016). Not all lower limb swelling in children is true lymphoedema. Approximately one fourth of paediatric cases of lower limb swelling are mis-diagnosed with lymphoedema in place of other vascular malformations and abnormalities. History, physical examination, and often radiographic studies are required to differentiate lymphoedema from other conditions to ensure the child is managed appropriately (Schook et al., 2011).

Differential Diagnosis of Lower Limb Lymphoedema in children

The differential diagnosis lower limb lymphoedema in children includes*(Schook et al., 2011):

- Microcystic/macrocystic lymphatic malformation
- Non-eponymous combined vascular malformation
- Klippel-Trenaunay syndrome
- Capillary malformation
- Hemihypertrophy
- Posttraumatic swelling
- Parkes Weber syndrome
- Lipoedema**
- Venous malformation
- Rheumatic disease
- Infantile haemangioma
- Kaposiform haemangioendothelioma
- ipofibromatosis

*Note that some of these conditions may also be linked with lymphoedema

** Note that lipoedema usually begins in puberty (Dadras et al., 2017)

Charter of Care for Children and Young People with Lymphoedema

Clinicians may refer to the BLS and the Children's Lymphoedema Specialist Interest Group "[Charter of Care for Children and Young People with Lymphoedema](#)" for a thorough approach to managing children and young people with lymphoedema.

The themes of the charter are:

- **Diagnosis and Treatment** addresses the pathway to diagnosis and the requirements of the lymphoedema treatment service for specialist care suitable for the physical and psychological needs of a child or young person.
- **Communication, Involvement and Collaboration** addresses the need for close medical and social multiagency care with direct involvement of the child, young person, and their parent/guardian.
- **Environment** recognises the need for appropriate environment of the lymphoedema service.
- **School Nurse** – Transition addresses the collaboration and support required for the child and school once they reach school age and as the young person then progresses through it.

- **Transition to Adulthood** recognises the need to support the transition from child to adult services.
- **Evaluation** recognises the need for continued service improvement through comment and evaluation.
- **Support** recognises the child, young person, and parent/guardian need for support and where this can be sought.

PQ1: How should lymphoedema be diagnosed in children and young people (CAYP)?

Evidence Summary

A review of the literature and expert opinion document addressed this question. (Damstra and Mortimer, 2008)

CAYP with suspected primary lymphoedema should be examined for syndromic characteristics (Greene and Goss, 2018). Syndromic features may include:

- Lymphoedema distichiasis syndrome: extra eyelashes at birth, eyelid proptosis, cleft palate.
- Hypotrichosis-lymphoedema-telangiectasia syndrome: sparse hair, cutaneous telangiectasias.
- Hennekam syndrome: generalized oedema, visceral involvement, developmental delay, flat faces, hypertelorism, broad nasal bridge.
- Noonan syndrome: short stature, pectus excavatum, webbed neck, hypertelorism, low-set ears, and/or a flat nasal bridge.
- Turner syndrome: short stature, webbed neck, broad chest, and/or low set ears.

VASCERN guidelines (VASCERN, 2019) recommend that a thorough patient history should include the following:

- Age of onset
- Distribution
- Cellulitis history
- Systemic symptoms
- Cutaneous symptoms e.g. warts
- Segmental overgrowth
- Family history
- Associated problems
- Venous disease
- Surgical history

See figure 2 for the St. George's classification algorithm for primary lymphatic anomalies.

See [appendix II.III](#) for the paediatric assessment and review record templates

Expert opinion recommends that all CAYP presenting with swelling at birth or swelling for 3 months or more should be assessed and investigated for presence of lymphoedema

Recommendations

PQ1.1 All CAYP presenting with swelling at birth or swelling for 3 months or more should be assessed and investigated for presence of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PQ1.2 A complete history and clinical assessment may be sufficient to diagnose lymphoedema in CAYP. Additional investigations may be sought based on individual clinical presentation, as determined by the primary medical team.

Evidence Grade: D

Strength of recommendation: Strong

PQ1.3 If required, the choice of investigation depends on the clinical presentation of the CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ1.4 A specific paediatric assessment form should be used in the initial assessment of lymphoedema in CAYP. Please see the paediatric assessment form.

Evidence Grade: D

Strength of recommendation: Strong

PQ2: How should response to treatment be measured in CAYP with lymphoedema?

Evidence Summary

A number of different methods are used to measure response to treatment in CAYP with lymphoedema including circumferential measurement, volumetry, tissue dielectric constant (TDC), bioimpedance, quality of life reports and activity levels pre- and post- treatment. While limb circumference is frequently used as an outcome measure in adults, a study of 223 healthy CAYP found that limb circumference may measure more than fluid in CAYP when compared to bioimpedance spectroscopy and hence calls into the question the accuracy of this measurement technique in CAYP (Avila et al., 2015).

A prospective cohort of CAYP with primary lymphoedema (n = 155) measured response to treatment subjectively, considering whether volume increased, decreased or remained stable. Other subjective measures included whether there was greater or lesser suppleness or tightness of the skin, by the parents and/or the child where appropriate. A systematic review (Phillips and Gordon, 2014) examining the conservative treatment of lymphoedema in CAYP calls into question the efficacy of using the unaffected limb as a control to compare to the affected limb to monitor response to treatment. Comparisons made over time in paediatric lymphoedema are not useful due to the natural changes in limb length and circumference due to growth, rendering volumetry measurement inaccurate. Due to these challenges posed by growth, one study (Hassall et al., 2001) proposed a ratio of foot to thigh volume to assess change in limb volume in CAYP with bilateral lymphoedema. The method in this study was not published, rendering the validity of this measurement to be unsupported. The use of a ratio requires further investigation, as it is a plausible method to monitor change within a limb in a growing child or where bilateral lymphoedema is present.

Evidence Summary (cont.)

Outcome measures used in practice by lymphoedema therapists working with CAYP and young adults include:

- Subjective reports
- Activity levels
- Quality of life
- Circumferential measurements
- Volumetry measurements
- Bioimpedance

VASCERN guidelines (VASCERN, 2019) recommend that the following may be carried out in CAYP with suspected lymphoedema:

- Limb volume
- QoL
- Physical clinimetrics
- Tissue dielectric constant (TDC) / bioimpedance

NSW Child Health Network (2010) recommend the following to assess swelling in CAYP:

- Circumferential limb measurements
- Water displacement
- Perometry
- Bioimpedance

Expert opinion recommends that a specific paediatric assessment form should be used to measure outcomes in CAYP with lymphoedema (Refer to [appendix II.III](#)).

Recommendations

PQ2.1 Subjective reports, activity levels, quality of life as well as circumferential measurements, volumetry measurements, TDC and bioimpedance may all be used to assess response to treatment in CAYP with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PQ2.2 A specific [paediatric assessment form](#) should be used to measure outcomes in CAYP with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PQ3: What is the evidence for bioimpedance in CAYP with lymphoedema?

Evidence Summary

Currently there is conflicting evidence for the use of bioimpedance in CAYP. A comparative study of healthy CAYP (n = 223) examining bioimpedance in CAYP reported no statistically significant correlation between extracellular impedance ratio and difference in limb circumference, except in the case of arms in adolescents. These findings may suggest that limb circumference measures quantities other than fluid, suggesting that different elements of body composition are measured by bioimpedance and circumferential tape measure (Avila et al., 2015). Another study indicated that bioimpedance has been validated as a measure of normally hydrated weight and can assess fluid status in CAYP as young as 2 years old (Dasgupta et al., 2018). Bioimpedance has also been demonstrated as a valid measure of assessing body composition in overweight and obese CAYP (de-Mateo-Silleras et al., 2019, Pecoraro et al., 2003). The accuracy of bioimpedance as a method to assess body composition seems to decrease in severely obese adolescents (Verney et al., 2016).

Recommendation

PQ3.1 Bioimpedance may be considered as an adjunctive outcome measure in CAYP with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PQ4: What imaging techniques should be carried out in CAYP with swelling suspected to be lymphoedema?

Evidence Summary

VASCERN guidelines (VASCERN, 2019) suggest that the following imaging techniques may be part of the investigations carried out on CAYP with suspected swelling:

- Lymphoscintigraphy
- MRI
- MRL
- Intranodal lymphangiography
- Ultrasound – venous, soft tissue
- ICG
- Chest x-ray
- Abdominal ultrasound

NSW Child Health Network (2010) suggest the following imaging modalities be used when investigating CAYP with lymphoedema:

- Lymphoscintigraphy
- MRI
- CT
- Ultrasound

Evidence Summary (cont.)

The IUP consensus document (Lee et al., 2013a) recommends the following are essential tests that every patient with suspected primary lymphoedema should undergo:

- Plain x-ray
- Duplex ultrasonography
- MRI with or without contrast
- CT scan
- Radionuclide lymphoscintigraphy

The IUP guidance document recommends that the following imaging tests are optional:

- Ultrasonographic lymphangiography
- MR lymphangiography
- Microscopic fluorescent lymphangiography
- Indocyanine green imaging
- Indirect lymphography using water-soluble contrast agents
- Whole body blood pool scintigraphy
- Volumetry
- Bioimpedance Spectroscopy
- Air plethysmography

Lymphoscintigraphy

A retrospective review of patients with primary lymphoedema found that lymphoscintigraphic imaging findings and quantification can be characteristic in certain genetic forms of primary lymphoedema (Sarica et al., 2019). Lymphoscintigraphy may therefore be useful as an additional tool for in-depth phenotyping, leading to a more accurate diagnosis of aetiology of primary lymphoedema. Patients with Turner syndrome (TS) with signs of lymphatic dysplasia and those with TS with minimal or absent signs of lymphatic impairment could undergo lymphoscintigraphy (Bellini et al., 2009). A retrospective evaluation of CAYP with lymphoedema who underwent lymphoscintigraphy found that using both a qualitative and quantitative assessment of drainage patterns provides the best method of detecting abnormal lymphatic drainage (Wachsmann et al., 2013). Whole body lymphangioscintigraphy and SPECT/CT have also been proven safe and effective techniques for the initial evaluation of lymphatic abnormalities in CAYP with complex congenital heart disease (Kuo et al., 2019).

Lymphoscintigraphy versus indocyanine green lymphography

A study comparing lymphoscintigraphy versus indocyanine green lymphography (ICG) concluded that ICG could be used first as a screening test for primary lymphoedema (if available) and if results are positive, lymphoscintigraphy may be used for further assessment. ICG lymphography has been shown to demonstrate abnormal patterns in symptomatic CAYP with primary lymphoedema and normal patterns in asymptomatic limbs (Yamamoto et al., 2015).

Note that ICG is not currently widely available within the NHS or the HSE.

Recommendation

PQ4.1 The choice of investigation for paediatric lymphoedema depends on clinical presentation and may require onward referral to specialist vascular centres for appropriate imaging. The choice of investigation may also be dependent on availability.

Evidence Grade: D

Strength of recommendation: Strong

PQ5: What blood tests should be carried out for CAYP with swelling suspicious of lymphoedema?

Evidence Summary

VASCERN guidelines (VASCERN, 2019) recommend the following blood tests may be carried out as part of investigation for primary lymphoedema in CAYP:

- Albumin
- Full blood count
- Immunoglobulin stool
- Alpha-1 antitrypsin

NSW Child Health Network (2010) suggest the following blood tests may be carried out:

- Albumin
- Markers of immune function
- Renal profile

THE ILF (2010) recommend the following blood tests may be carried out:

- Albumin
- Markers of immune function
- B-type natriuretic peptide (BNP)
- Renal profile

Recommendation

PQ5.1 The choice of blood tests carried out in the investigation of paediatric lymphoedema depends on the clinical presentation of the patient and the subsequent differential diagnoses which must be ruled out. Refer to [St. George's Investigation Tool](#) for further guidance.

Evidence Grade: D

Strength of recommendation: Strong

PQ6: What should be included in the clinical assessment of CAYP with lymphoedema?

Evidence Summary

VASCERN guidelines (VASCERN, 2019) recommend that the following elements of a clinical assessment should be carried out in CAYP with swelling suspicious of lymphoedema:

- Family history and travel history
- Examination (Stemmer's sign / pitting oedema)
- Urinalysis to assess for evidence of albumin

NSW Child Health Network (2010) suggest the following elements should be assessed in CAYP with a clinical suspicion of lymphoedema:

Clinical history

- History and behaviour of swelling
- Symptoms such as heaviness, tightness, or hardness
- History of skin or nail infections
- Family history

Evidence Summary (cont.)

VASCERN guidelines (VASCERN, 2019) recommend that the following elements of a clinical assessment should be carried out in CAYP with swelling suspicious of lymphoedema:

- Family history and travel history
- Examination (Stemmer's sign / pitting oedema)
- Urinalysis to assess for evidence of albumin

NSW Child Health Network (2010) suggest the following elements should be assessed in CAYP with a clinical suspicion of lymphoedema:

Clinical history

- History and behaviour of swelling
- Symptoms such as heaviness, tightness, or hardness
- History of skin or nail infections
- Family history

Recommendations

PQ6.1 The clinical assessment of a CAYP with lymphoedema should include thorough assessment of the skin, objective measurement of the swelling and subjective symptoms, as well as a thorough family and developmental history if indicated.

Evidence Grade: D

Strength of recommendation: Strong

PQ6.2 Clinicians should be aware of and document evidence of dysmorphic features typical of syndromes associated with primary paediatric lymphoedema as part of a thorough clinical examination. Refer to Appendix [I.II](#) for the St. George's Algorithm for further guidance

Evidence Grade: D

Strength of recommendation: Strong

PQ7: At what age is lymphoscintigraphy suitable in CAYP?

Evidence Summary

Lymphoscintigraphy has been shown to be safe and useful in the diagnosis of primary lymphoedema in CAYP as young as newborns (Bellini et al., 2014, Bellini et al., 2008, Baulieu et al., 2003). Expert opinion recommends that lymphoscintigraphy may be more clinically appropriate in older CAYP

Recommendations

PQ7.1 Lymphoscintigraphy is safe from birth and may be considered as an investigation in CAYP, however it may be more clinically appropriate in older CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ8: How should intestinal lymphangiectasia be diagnosed in CAYP?

Evidence Summary

Intestinal lymphangiectasia is a rare condition which affects the lymph vessels supplying the lining of the small intestine and may result in malabsorption.

St. George's Hospital published a Paediatric Investigation Pathway for Primary Lymphoedema in Childhood based on expert consensus (CLSIG, 2016) which recommends that patients being investigated for intestinal lymphangiectasia should have the following investigations carried out:

- FBC / Albumin / alpha-1 antitrypsin
- Faecal alpha-1 antitrypsin and calprotectin

They also recommend that patients with suspected intestinal lymphangiectasia need to be referred to expert dietitian to discuss a medium chain triglyceride (MCT) diet.

Expert opinion recommends that CAYP with intestinal lymphangiectasia should be referred to a gastroenterologist for review.

Recommendations

PQ8.1 CAYP being investigated for intestinal lymphangiectasia should have the following investigations carried out:

- FBC
- Albumin
- Alpha-1 antitrypsin
- Faecal alpha-1 antitrypsin
- Faecal calprotectin

Evidence Grade: D

Strength of recommendation: Strong

PQ8.2 CAYP with suspected intestinal lymphangiectasia should be referred to a dietitian to discuss an MCT diet.

Evidence Grade: D

Strength of recommendation: Strong

PQ8.3 CAYP with suspected intestinal lymphangiectasia may require referral to gastroenterology as part of an MDT approach to care.

Evidence Grade: D

Strength of recommendation: Strong

PQ9: What genetic tests are indicated for investigation of primary lymphoedema in CAYP?

Evidence Summary

St. George's Hospital published a Paediatric Investigation Pathway for Primary Lymphoedema in Childhood based on expert consensus (CLSIG, 2016) which recommends the following:

If child appears dysmorphic or has learning difficulties:

- Carry out array CGH (detailed chromosome analysis)

Congenital lower limb lymphoedema:

- VEGFR3 for suspected Milroy disease
- KIF11 if microcephaly present
- Turner syndrome if female
- Noonan panel if dysmorphic

Congenital generalised lymphoedema (e.g. hydrops fetalis, chylous effusions, ascites, intestinal lymphangiectasia, pericardial effusions and widespread lymphoedema):

- Consider CCBE1/ FAT4 / PIEZO1 / Noonan gene panel.

Childhood onset of bilateral lower limb lymphoedema (after the age of 1 year):

- FOXC2 (especially if distichiasis present)
- GATA2 (especially if genital involvement, low monocyte count)
- GJC2 (especially if hands are swollen too)
- Noonan panel (if dysmorphic and other associated features)
- Full blood count- refer to Haematology if any concerns.

Multi-segmental lymphoedema with evidence of overgrowth:

- Consider taking a skin biopsy for PIK3CA gene testing.

Isolated genital lymphoedema:

- Consider Noonan syndrome (gene panel test).
- Consider anogenital granulomatosis: take skin biopsies of scrotum and/or penis looking for granulomas within the dermis.
- Refer to gastroenterology for consideration of endoscopy and biopsy looking for Crohn's disease.

A small case series (n = 3) concluded that neurological assessment including electroencephalography (EEG) be carried out on CAYP with generalised lymphoedema and facial involvement to identify generalized lymphoedema associated with neurologic signs (GLANS) syndrome (Berton et al., 2015). VASCERN guidelines (VASCERN, 2019) recommend that where appropriate CAYP should be referred to a genetic specialist for genetic testing.

Recommendation

PQ9.1 Genetic tests required for the investigation of primary lymphoedema in CAYP depend on the clinical presentation of the patient.

Evidence Grade: D

Strength of recommendation: Strong

PQ10: Which CAYP with lymphoedema should be referred for genetic testing?

Evidence Summary

A classification system for primary lymphoedema has been proposed which may assist in highlighting which patients require referral for genetic testing (Connell et al., 2010). CAYP with any of the following features should undergo genetic testing:

- Syndromic Features
- Systemic/visceral involvement (e.g. chylous, pericardial/pleural effusions, ascites and pulmonary/intestinal lymphangiectasia)
- Altered growth
- Cutaneous Features
- Vascular Abnormalities
- Congenital onset lymphoedema
- Distichiasis

The International Lymphoedema Framework (ILF) (2010) recommend that CAYP with lymphoedema and dysmorphic features and/or learning difficulties should be referred for consideration of genetic testing.

Recommendation

PQ10.1 CAYP with lymphoedema and any of the following features should undergo genetic testing:

- Syndromic Features
- Congenital onset lymphoedema
- Early or late onset lymphoedema
- Systemic/visceral involvement (e.g. chylous, pericardial/pleural effusions, ascites and pulmonary/intestinal lymphangiectasia)
- Altered growth
- Cutaneous features
- Vascular abnormalities
- Distichiasis
- Learning difficulties
- Family history of lymphoedema

Evidence Grade: D

Strength of recommendation: Strong

PQ11: Should all CAYP with primary lymphoedema be offered genetic counselling?

Evidence Summary

The CAYP's Lymphoedema Special Interest Group (CLSIG) Charter of Care for CAYP and Young People with Lymphoedema recommend that in cases where a genetic cause is suspected the child or young person should have access to genetic counselling ((CLSIG), 2016). The ILF state that CAYP with inherited forms of lymphoedema should undergo genetic counselling (2010).

Patients with lymphoedema-distichiasis syndrome (LDS) should undergo genetic counselling as it is inherited in an autosomal dominant manner (Mansour et al., 1993). If the FOXC2 pathogenic variant is identified in an affected family member, prenatal testing may be possible and foetal echocardiography is recommended as there is an increased risk of CHD.

Patients with Milroy disease should undergo genetic counselling as MD also demonstrates an autosomal dominant pattern of inheritance (Brice et al., 1993). Ultrasonography carried out during pregnancy may detect oedema of the dorsum of the foot, pleural effusions and rarely more extensive oedematous states such as hydrops fetalis. Prenatal testing in at-risk families may be completed but is rarely requested.

Recommendation

PQ11.1 CAYP and their families with a suspected genetic cause of their lymphoedema should be offered genetic counselling.

Evidence Grade: D

Strength of recommendation: Strong

PQ12: How should compression garments be prescribed in CAYP with lymphoedema?

Evidence Summary

VASCERN guidelines recommend compression garments and bandages be a part of the initial and maintenance phase of lymphoedema management in CAYP (Paton et al., 2020). A clinical guideline (New South Wales Agency for Clinical Innovation, 2018) in compression therapy recommends the following in relation to the care of CAYP with lymphoedema :

“There is very limited evidence regarding treatment techniques in paediatric lymphoedema. An experienced paediatric therapist should consider the....typical lymphoedema treatment options in adults....and make necessary and appropriate modifications considering the age, size, growth, understanding, and requisite monitoring of their patient.”

A prospective cohort study (n = 155) of CAYP with primary lymphoedema used multilayer low-stretch bandages and instructed CAYP and/or their carers to cover their lymphoedema affected area for the longest duration possible during a 24 hour period, at night for ambulant CAYP and for full 24 hours otherwise. Ambulant CAYP (n = 128, 83%) were prescribed the constant wearing of custom-made and circular-knit elastic garment (20–36 mmHg). The garments always had a closed toe, to avoid worsening of toe lymphoedema and potential lymph oozing (Vidal et al., 2016). At median follow up of 38 months, 97% of CAYP or their carers reported improved

Evidence Summary (cont.)

lymphoedema symptoms. A small study (n = 5) examined the use of homemade compression stockings and shoes in the management of primary congenital lymphoedema (de Godoy et al., 2010). The material used was a cotton/polyester blend and had a low-elasticity, allowing low-stretch compression. This study found a positive association between duration of daytime use of compression and improvements in limb volume.

Pneumatic Compression

A systematic review (Phillips and Gordon, 2019) of available evidence revealed that there is low-level evidence from moderate quality studies showing significant positive outcomes with dosage times of 45-60 minutes of IPC in adults and CAYP, with pressures in the range of 30-60 mmHg. However, owing to the methodological limitations of studies, conclusions are difficult to draw. A retrospective review of CAYP (n = 16) with primary and secondary lymphoedema who were treated with pneumatic compression found a trend towards significance in terms of improvement in limb volumes ($p > 0.05$) (Hassall et al., 2001).

Vascular Malformations

A systematic review (Langbroek et al., 2018) examining the effectiveness of compression therapy in the management of congenital low-flow vascular malformations (venous malformations or Klippel-Trenaunay syndrome), concluded that there is currently a lack of high-quality evidence to validate its use in this population. However, wearing compression may lessen intravascular coagulation, improve symptoms and limb appearance, reduce oedema, and protect against trauma. The authors highlight the need for prospective comparative trials with standardised outcome measures to better understand the benefits and risks of this treatment option.

Ambulation and compression

Expert opinion recognises that the act of ambulation may increase lymphatic clearance secondary to muscle pump action. Therefore swelling may improve in infants once they become mobile. This may be taken into consideration when deciding whether or not to use compression garments or bandaging with infants.

Recommendations

PQ12. 1 Compression garments and bandages should be part of the initial and maintenance phase of lymphoedema management in CAYP.

Evidence Grade: C

Strength of recommendation: Strong

PQ12.2 The lowest level of compression possible should be applied in CAYP with lymphoedema to achieve the desired clinical outcome.

Evidence Grade: D

Strength of recommendation: Strong

PQ12.3 Appropriate modifications should be made to compression in CAYP, considering the age, size, growth, engagement and health literacy of the CAYP and the patient/family preference.

Evidence Grade: D

Strength of recommendation: Strong

PQ12.4 Pneumatic compression may be considered as an adjunctive treatment in the management of paediatric lymphoedema.

Evidence Grade: A

Strength of recommendation: Strong

PQ13: What is the evidence for night compression for CAYP with lymphoedema?

Evidence Summary

A systematic review examining the conservative treatment of lymphoedema in CAYP concluded that currently there is insufficient evidence to recommend specific parameters for any treatment modality in CAYP (Phillips and Gordon, 2014).

Expert opinion recognises the potential benefit of night compression for CAYP with lymphoedema.

Recommendation

PQ13.1 Night compression may be beneficial for CAYP and may be considered as part of lymphoedema management.

Evidence Grade: A

Strength of recommendation: Strong

PQ14: How often should compression garments be remeasured in CAYP with lymphoedema?

Evidence Summary

The International Lymphoedema Framework (ILF) (2010) recommend that, taking into account growth and the type of activities CAYP engage in, CAYP require regular compression fittings and two to three sets every 6 months. A guidance document produced by the NSW agency for clinical innovation recommend that CAYP require garment replacement more often than every 6 months and the timing of remeasurements will depend on the growth rate of the child as well as with wear and tear (Innovation, 2018). Expert opinion is that children younger than 18 months may need to be measured every 4 months due to the rapid rate of growth at this age. Similarly, teenagers may need to be remeasured more frequently than six-monthly.

Recommendations

PQ14.1 Paediatric garment replacement may be considered more frequently than six monthly. This may be four monthly and will be dependent on the growth rate of the CAYP as well as wear and tear of the garments.

Evidence Grade: D

Strength of recommendation: Strong

PQ14.2 It is recommended that parents/guardians liaise with their clinicians when new compression garments are required so that they can be issued in a timely manner.

Evidence Grade: D

Strength of recommendation: Strong

PQ15: How many garments should CAYP with lymphoedema receive in a year?

Evidence Summary

The International Lymphoedema Framework (ILF) (2010) recommend that, taking into account growth and the type of activities CAYP engage in, they require regular compression fittings and more than two sets of compression garments every 6 months. A guidance document produced by the NSW agency for clinical innovation recommends that CAYP be issued with two sets of garments, however more sets may be required if the child is toilet training for example (Innovation, 2018).

Recommendations

PQ15.1 CAYP with lymphoedema should be supplied with at least two sets of compression garments per affected body part every 6 months, and more sets may be required on a case-by-case basis.

Evidence Grade: D

Strength of recommendation: Strong

PQ15.2 CAYP with lymphoedema may require more than two sets of compression garments per affected body part every 6 months because of growth and activity levels. Quarterly reviews with garment supply may be more relevant during periods of rapid growth.

Evidence Grade: D

Strength of recommendation: Strong

PQ16: Is there evidence to support bandaging (full or modified) in CAYP with lymphoedema?

Evidence Summary

There appears to be a lack of studies examining the efficacy of bandaging in the management of lymphoedema. A retrospective study of CAYP with lymphoedema (n = 86) found that 68% of cases were managed with multi-layered bandaging (Watt et al., 2017). A systematic review examined the conservative treatment of lymphoedema in CAYP and concluded that currently there is insufficient evidence to recommend specific parameters for any treatment modality in CAYP (Phillips and Gordon, 2014).

Expert opinion recognises that the act of ambulation may increase lymphatic clearance secondary to muscle pump action. Therefore swelling may improve in infants once they become mobile. This may be taken into consideration when deciding whether or not to use compression garments or bandaging with infants.

Recommendations

PQ16.1 Bandaging may be considered in conjunction with other modalities in the treatment of lymphoedema in CAYP.

Evidence Grade: C

Strength of recommendation: Strong

PQ16.2 Bandaging may be more appropriate when CAYP become ambulatory, unless there is another clear clinical indication for compression before the child is mobile.

Evidence Grade: D

Strength of recommendation: Strong

PQ17: What type of bandaging should be applied to CAYP with lymphoedema?

Evidence Summary

While short-stretch bandages are recommended to reduce limb volumes in adults with lymphoedema, little data exists to support their use in CAYP with lymphoedema. A retrospective study of CAYP (n = 48) with lower limb lymphoedema assessed the efficacy of short-stretch bandages in this cohort (Benoughidane et al., 2018). Results showed intensive lymphoedema-treatment based on short-stretch bandaging led to significant volume reduction (unilateral and bilateral), with no difference in outcomes between sexes. Expert opinion is that the lowest level of bandaging possible should be applied to CAYP with lymphoedema. Cohesive bandaging systems may stay in place better in active CAYP. Clinical response and arterial/vascular symptoms should be closely monitored throughout treatment.

Recommendations

PQ17.1 The lowest effective compression level of bandaging should be applied to CAYP with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PQ17.2 Cohesive bandaging systems may stay in place better for highly active CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ17.3 Clinical response and arterial/vascular symptoms should be closely monitored throughout treatment.

Evidence Grade: D

Strength of recommendation: Strong

PQ18: Should patients and their guardians be trained in bandaging to enhance outcomes in CAYP with lymphoedema?

Evidence Summary

There is a lack of research examining self-management and its impact on outcomes in CAYP with lymphoedema. VASCERN guidelines (VASCERN, 2019) recommend that patient and/or guardian education should be a component of the initial self-management of CAYP with lymphoedema. Expert opinion recommends that patients and/or guardians can be trained in self-bandaging and assessed for competency.

Recommendations

PQ18.1 Patients or their parents/guardians should be trained in bandaging techniques and competency assessed if this is part of the agreed treatment package.

Evidence Grade: D

Strength of recommendation: Strong

PQ19: What is the role of MLD/SLD in CAYP with lymphoedema?

Evidence Summary

There are reports that MLD treatment for lower limb lymphoedema may lead to reduction of swelling of other untreated regions of the body such as the upper limb, suggesting systemic effects of MLD (Pereira de Godoy et al., 2018). A small prospective study (n = 15) examining the efficacy of MLD in paediatric limb lymphoedema found that MLD significantly reduced limb circumference and dermal thickness, but not limb volume (Habnoui et al., 2020). This study also found that MLD is well accepted in CAYP and led to improvements in their reported well-being. MLD appears to be very common practice in the management of lymphoedema in CAYP, with one retrospective chart review (n = 86) demonstrating that 97% of cases were managed with MLD (Watt et al., 2017). A systematic review examined the conservative treatment of lymphoedema in CAYP and concluded that currently there is insufficient evidence to recommend specific parameters for any treatment modality in CAYP with lymphoedema (Phillips and Gordon, 2014). Expert opinion recommends that CAYP and/or parents/guardians be taught simple lymphatic drainage (SLD) techniques.

Recommendations

PQ19.1 MLD may be used as part of a combined treatment approach to assist in the reduction of swelling and may lead to improvements in the well-being of CAYP with lymphoedema.

Evidence Grade: C

Strength of recommendation: Strong

PQ19.2 CAYP and/or their parents/guardians should be taught simple lymphatic drainage techniques.

Evidence Grade: D

Strength of recommendation: Strong

PQ20: What is the optimal frequency for CDT in CAYP with lymphoedema?

Evidence Summary

A systematic review examined the conservative treatment of lymphoedema in CAYP and concluded that currently there is insufficient evidence to recommend specific parameters for any treatment modality in CAYP (Phillips and Gordon, 2014). A clinical guideline (Innovation, 2018) in compression therapy recommends the following in relation to the care of CAYP with lymphoedema:

“There is very limited evidence regarding treatment techniques in paediatric lymphoedema. An experienced paediatric therapist should consider the....typical lymphoedema treatment options in adults....and make necessary and appropriate modifications considering the age, size, growth, understanding, and requisite monitoring of their patient.”

Recommendations

PQ20.1 As there is currently insufficient evidence to recommend a frequency for CDT, clinical judgement should be used to decide optimal frequency and appropriate modifications to CDT for CAYP, considering the age, size, growth, understanding and required monitoring.

Evidence Grade: D

Strength of recommendation: Strong

PQ21: What is the indication for prophylactic antibiotics in CAYP with lymphoedema?

Evidence Summary

There have been no studies to date assessing the use of prophylactic antibiotics in CAYP with lymphoedema.

Recommendations

PQ21.1 The decision to prescribe antibiotic prophylaxis for CAYP with lymphoedema should be made by the specialist medical team after thorough clinical assessment.

Evidence Grade: D

Strength of recommendation: Strong

PQ22: What antibiotics are recommended for CAYP with cellulitis?

Evidence Summary

As in adults, cellulitis in CAYP may present with local symptoms of pain, discomfort, redness or swelling with or without general ill health and malaise. It is important to treat early, and recognise that CAYP who present with systemic symptoms of infection or have deteriorating local signs should be seen in hospital and treated aggressively with intravenous antibiotics.

As for adults there appear to be no trials examining the efficacy of antibiotics in the treatment of cellulitis in CAYP living with lymphoedema. The ILF recommend that adult protocols for the treatment of cellulitis/erysipelas be followed with dosage adjusted according to the size of the child (International Lymphoedema Framework, 2010).

Recommendation

PQ22.1 There are several antibiotic regimens available to treat cellulitis in patients with lymphoedema. Clinicians should be aware of the [BLS guidance](#) on this topic however they should consult their local antimicrobial guidelines in the first instance.

Evidence Grade: D

Strength of recommendation: Strong

PQ23: How should skin care be managed in CAYP with lymphoedema?

Evidence Summary

VASCERN guidelines recommend that skin care should form part of the initial and maintenance phase of treatment for CAYP with lymphoedema (VASCERN, 2019). NSW Child Health Network (2010) recommend the following as part of skin and limb care:

- Daily inspections for breaks in skin integrity (e.g. cuts, bites or scrapes)
- Regular moisturising
- Referral for assessment if any signs of infection
- Regular nail care
- Sensible footwear and clothing
- Avoiding sunburn or overheating
- Avoiding the affected limb if repeated blood pressure measurements and/or injections are required

Recommendation

PQ23.1 Skin care management of lymphoedema in CAYP should include:

- Daily inspections for breaks in skin integrity (e.g. cuts, bites or scrapes)
- Regular moisturising
- Referral for assessment if any signs of infection
- Regular nail care
- Sensible footwear and clothing
- Avoiding sunburn or overheating
- Avoiding the affected limb if repeated blood pressure measurements and/or injections are required

Evidence Grade: D

Strength of recommendation: Strong

PQ23.2 The choice of emollient used in CAYP should be based on the skin integrity and level of dryness of the skin

Evidence Grade: D

Strength of recommendation: Strong

PQ24: Is surgery suitable for CAYP with lymphoedema?

Evidence Summary

A systematic review addressed this question (Kanth et al., 2019). This review concluded that overall evidence examining surgical management of lymphoedema is lacking and hence definitive conclusions regarding its efficacy cannot be drawn. Surgical outcomes were positive for genital lymphoedema in CAYP. Excisional surgical procedures appear to be successful in the treatment of extremity lymphoedema. Physiological procedures had mixed outcomes in the small population studied.

Head and Neck Lymphatic Malformations

A systematic review of 41 articles (n = 1,205) concluded that lymphatic malformations of the head and neck may be treated with surgery or sclerotherapy and currently there is insufficient evidence to draw firm conclusions on which is superior (Adams et al., 2012).

Genital Lymphoedema

A small retrospective study (Schook et al., 2014) of male CAYP with genital lymphoedema found that patients who underwent surgical contouring had sustained improvement in their symptoms after a median follow up time of 4.2 years.

Lymphaticovenous Anastomosis (LVA)

In a retrospective study (Hara et al., 2015), LVA was shown in the under 11 age group to be effective only in select cases of patients diagnosed with primary lymphoedema. The procedure was, however, found to be effective in patients who developed lymphoedema after the age of 11.

Microsurgery

In a retrospective study of CAYP with primary lymphoedema, microsurgery significantly improved cellulitis episodes and QoL without the need for compression garments (Cheng and Liu, 2020).

Quality of Life

A qualitative study (n = 109) assessed the QoL of CAYP who had undergone surgery for primary lymphoedema. Results from this survey reported that surgery for severe lymphoedema improved QoL at early assessment but that these results may not be sustained. Genital lymphoedema reduction appeared to lead to greater perceived benefit and increase in QoL compared to limb reduction (Ogunbiyi et al., 2009).

Recommendation

PQ24.1 In select cases under specialist guidance, surgery may be considered in the treatment of lymphoedema in CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ25: What specific types of physical activity should be advised for CAYP with lymphoedema?

Evidence Summary

There do not appear to be any trials published comparing exercise types for CAYP living with lymphoedema. VASCERN guidelines recommend that exercise be a component of both the initial and maintenance phase of lymphoedema management in CAYP (VASCERN, 2019).

The BLS Children's Lymphoedema Specialist Interest Group (CLSIG) run a bi-annual 'Lymphaletics' event which is free for families to attend. This award winning one-day event encourages physical activity as one of its key aims, alongside peer support and teaching.

Recommendation

PQ25.1 Physical activity should be an integral component of both the initial and maintenance phases of lymphoedema management in CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ25.2 New exercise activities should be introduced gradually, and progressed slowly. CAYP and their parents/carers should monitor their condition closely after initiating a new form of physical activity, and continue to monitor as their activity progresses.

Evidence Grade: D

Strength of recommendation: Strong

PQ25.3 There is currently insufficient evidence to recommend one exercise type over another so choice of physical activity should be based on the CAYP's preference and ability.

Evidence Grade: D

Strength of recommendation: Strong

PQ25.4 Clinicians should consider recommending that CAYP and their families/carers investigate the 'Lymphaletics' events. Services should also consider organising local events to enhance local family peer support.

Evidence Grade: D

Strength of recommendation: Strong

PQ26: Should all CAYP with lymphoedema be referred to a specialist paediatric lymphoedema centre?

Evidence Summary

The Charter for Care for CAYP and Young People with Lymphoedema was developed by the CAYP's Lymphoedema Special Interest Group (CLSIG), a group of healthcare professionals involved in the care of CAYP living with lymphoedema aligned to the BLS and LSN. This charter states that all CAYP with lymphoedema should be referred to a lymphoedema centre ((CLSIG), 2016). This expert group recommend that: "CAYP can be seen in an Adult Lymphoedema service that has provision for CAYP. The service should offer the child or young person and their families an opportunity to meet others with lymphoedema and should provide access to other services such as paediatrics, genetics and play therapists. The delivery of treatment should be conducted in a setting that meets their physical and psychological needs and the environment should be conducive to their developmental stage of learning and comprehension".

The ILF recommend that CAYP with lymphoedema be referred to a specialist or advanced lymphoedema practitioner (2010).

Recommendation

PQ26.1 CAYP with lymphoedema should ideally be assessed in a national specialist clinic with dedicated staff, which will ensure standardised care and psychological support including meeting other CAYP with lymphoedema, in partnership with the local care provider.

Evidence Grade: D

Strength of recommendation: Strong

PQ26.2 Once lymphoedema has been diagnosed, management may be carried out in a specialist centre or a service with appropriate resources to cater for the needs of CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ26.3 Ongoing concurrent management of lymphoedema can continue while investigations are being carried out, once serious pathology has been ruled out by the medical team.

Evidence Grade: D

Strength of recommendation: Strong

PQ26.4 It is recommended that clinicians treating lymphoedema in CAYP should engage in ongoing continuous professional development (CPD) specific to paediatric populations. Please see the [education section](#).

Evidence Grade: D

Strength of recommendation: Strong

PQ27: What is the most effective method of providing information to CAYP with lymphoedema?

Evidence Summary

A clinical practice guideline (Innovation, 2018) on compression recommends the following in relation to education in CAYP with lymphoedema: "For CAYP, education of the patient as well as the parents/carers and siblings should be included in basic management. This education must be age appropriate and consider the patient's understanding, capacity to self-manage and adherence levels where capable. Other community carers (e.g. school, sports coaches) may need education about lymphoedema and its management."

One study examined the role of an educational camp in promoting self-efficacy in CAYP with lymphoedema (Moffatt et al., 2019). A book aimed at CAYP living with lymphoedema "The Big Book of Lymphoedema" by Jacqueline Todd won the patient information for CAYP award from the British Medical Association. The book has been translated into many different languages and is the first book of its kind to be rolled out globally in hospitals to assist CAYP in understanding lymphoedema. The book can be ordered from [Lymphshop](#).

The Lymphoedema Support Network (LSN) provide free membership for CAYP with lymphoedema and offer a range of information for the child and the family.

Recommendations

PQ27.1 Education provided to CAYP with lymphoedema must be age appropriate and consider the CAYP's understanding and capacity to self-manage.

Evidence Grade: D

Strength of recommendation: Strong

PQ27.2 Families and/or carers should be included in lymphoedema education as part of the management of lymphoedema in CAYP.

Evidence Grade: D

Strength of recommendation: Strong

PQ27.3 If CAYP are competent in its use, the use of electronic information and communication should be considered e.g. via websites, e-groups etc. All information should be provided in an accessible format.

Evidence Grade: D

Strength of recommendation: Strong

PQ28: What age should transition to adult services commence for CAYP with lymphoedema?

Evidence Summary

CAYP's Lymphoedema Special Interest Group (CLSIG) recommend that when the young person is approaching adulthood, a discussion regarding transfer of their care to adult services should occur ((CLSIG), 2016). International Lymphoedema Framework (ILF) (2010) state that the transition to adult services may not necessarily be age dependent. The age at which a person is considered a child or young person can vary from agency to agency. Currently the Department of Health in the UK considers anyone under the age of 19 years a child or young person. The ILF recommend that for CAYP with access to small, expert units, the transition is generally seamless as they continue to see the same health professionals. This may be achieved by the same clinician continuing in the role of lead care provider, with care supplemented with clinics for young adults which provide social interaction, education and support. A clinical guidance document on compression recommends that care referrals be individualised for young people with lymphoedema (Innovation, 2018). Patients generally should be referred to adult services when they turn 18. Patients who present after the age of 16 may attend an adult service rather than starting treatment in a paediatric service.

Recommendation

PQ28.1 CAYP attending a paediatric service generally should be referred to adult services when they turn 18. If CAYP present with lymphoedema after the age of 16, they may attend an adult service rather than starting treatment in a paediatric service.

Evidence Grade: D

Strength of recommendation: Strong

PQ29: Which HCPs should be members of the MDT treating CAYP with lymphoedema?

Evidence Summary

Dutch Guidelines recommend that CAYP with lymphoedema should be treated by a team including a paediatrician, paediatric physiotherapist, geneticist, lymphoedema specialist, dermatologist and nurse (Damstra and Halk, 2017). The International Lymphoedema Framework (2010) recommend that a team of health professionals is required to fully meet the needs of a child with lymphoedema. This team should comprise a specialist/advanced lymphoedema practitioner, a paediatrician, a geneticist, a clinical psychologist and a play therapist. Other members may include dermatologist, physiotherapist, occupational therapist, surgical fitter, surgeon and dietician.

Recommendations

PQ29.1 Members of the MDT involved in the care of CAYP with lymphoedema depend on the clinical presentation of the CAYP and may include lymphoedema practitioners, dermatologists, physiotherapists, lymphoedema therapists, occupational therapists, nurses, dietitians, podiatrists, play therapists, psychologists and paediatricians.

Evidence Grade: D

Strength of recommendation: Strong

PQ29.2 There should be identified referral pathways for all MDTs involved in the care of CAYP with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PQ29.3 Community healthcare staff (e.g. school nurse, health visitor, home help or carers) should also be included in the MDT treating CAYP with lymphoedema, and formal communication links should be established.

Evidence Grade: D

Strength of recommendation: Strong

PQ29.4 Where appropriate, links should be made with charitable organisations related to CAYP's specific conditions e.g. BLS Children's Lymphoedema Special Interest Group (CLSIG) and LSN children's group.

Evidence Grade: D

Strength of recommendation: Strong

7. Lymphoedema in people living with obesity (PWO)

Obesity affects one third of the population in the United States, 27% of adults in Northern Ireland and 23% of adults in the Republic of Ireland.

A threshold may exist for lymphoedema development in PWO with a suggested tipping point of a BMI of between 50-60 kg/m² (Greene et al., 2015). At this stage, an increased risk of lower extremity lymphatic function appears to present. Green et al. (2015) performed lymphoscintigraphy in 15 people with severe obesity (BMI > 40 kg/m²) with no prior history of lymphoedema. The average BMI of those in the study with lymphoedema (70.1 kg/m²) was significantly greater than the BMI of those without lymphoedema (42.0 kg/m²). All patients with a BMI above 59 kg/m² had evidence of lymphoedema, whereas every patient with a BMI less than 54 kg/m² had normal lymphatic function.

Lymphoedema-like swelling can affect the lower limbs of people living with severe obesity, despite normal lymphoscintigraphy. Abnormal lymphatic and/or venous drainage in PWO can impact limb movement and consequently both physical function and physical activity levels, which in turn reduce lymphatic flow. Obesity affects skin barrier function, lymphatic function, collagen function, wound healing and vascular health which may explain the increased propensity for PWO to develop cellulitis (Savetsky et al., 2014).

Obesity as a Risk Factor

A BMI of greater than 25 kg/m² has been associated with a greater risk of developing lymphoedema. The oedema component in obesity is associated with increased lymph production due to the increased ultrafiltration and overburdening of the lymphatic system, rather than a structural impairment. In a prospective cohort study (n = 486) of women who developed lymphoedema after cancer treatment, pre-morbid obesity was significantly associated with lymphoedema symptoms.

PWO may be at risk of developing lymphoedema because they have compromised lymphatic function at baseline, abnormal inflammatory responses that can negatively impact the lymphatic system, and have impaired ability to regenerate damaged lymphatics after injury.

Massive Localised Lymphoedema

Massive Localised Lymphoedema (MLL) is a non-malignant condition that can clinically mimic a variety of soft tissue tumours. Patients with MLL are typically females with BMI > 40 kg/m² who present with non-specific symptoms. The diagnosis of MLL is challenging, and its aetiology and treatment are not extensively described in the literature. MLL is a consequence of obesity-related Lymphoedema and affects approximately 60% of PWO with lower-extremity lymphatic dysfunction. BMI > 56 kg/m² significantly increases the risk of MLL. PWO should be referred to a bariatric weight-loss centre before their BMI reaches a threshold for obesity-related lymphoedema or MLL to develop.

OBQ1: How should obesity-related lymphoedema be diagnosed?

Evidence Summary

A review of obesity-related lymphoedema (Greene, 2016) recommends that lymphoscintigraphy should be used to diagnose this condition. A thorough history (including family history) and clinical examination should be completed to investigate for primary lymphoedema and to assess for other secondary causes of lymphoedema.

Lymphoscintigraphy provides a definitive diagnosis of obesity-related lymphoedema if after thorough assessment, the diagnosis remains unclear. Where resources allow, patients with possible obesity-related lymphoedema should undergo this study to determine whether or not they have lymphatic dysfunction.

Studies have shown the prevalence of obesity-related lymphoedema-like swelling to be as high as 31.5% (O'Malley et al., 2015) and up to 18.7% of PWO have an associated lifetime prevalence of cellulitis (Corcoran et al., 2020). People living with obesity should not have treatment delayed in the absence of access to lymphoscintigraphy.

Subclinical Lymphoedema

A small cross-sectional study (n = 30) examined the diagnosis of subclinical lymphoedema in patients living with obesity (de Godoy, 2019). The study used a thorough history, physical examination, and measurement of intracellular and extracellular fluid levels via bioelectrical impedance to diagnose subclinical lymphoedema in patients living with obesity. The authors concluded that subclinical lymphoedema occurs earlier in patients who go on to develop clinical lymphoedema

Refer to the [oncology section](#) of this guideline for further details on the diagnosis of subclinical lymphoedema.

Recommendation

OBQ1.1 Obesity-related Lymphoedema should be diagnosed using subjective and objective means via a standardised assessment template. Refer to appendix II for assessment templates. If the diagnosis remains unclear, lymphoscintigraphy may be considered to confirm pathology.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2: What additional considerations should be made in the assessment of people living with obesity and lymphoedema?

Evidence Summary

Obesity is a chronic disease. Similarly to lymphoedema, it necessitates a systematic and comprehensive approach to its diagnosis, assessment and treatment. Obesity Canada recommend a non-judgmental, stigma-free approach to be an essential component of an effective assessment of a person living with obesity (Rueda-Clausen et al., 2020).

Causes of obesity and obesity complications, in addition to potential barriers to treatment, should be identified by taking a comprehensive patient history and clinical examination. Obesity screening should include the measurement (with patient consent) of body mass index (BMI) and waist circumference (in individuals with a BMI 25-35 kg/m²). Further appropriate screening tests include: blood pressure measurement in both arms, fasting glucose, glycated haemoglobin, lipid profile, liver profile to screen for non-alcoholic fatty liver disease. The use of the Edmonton Obesity Staging System should be considered to determine the severity of obesity and to guide clinical decision making.

The assessment process should include the establishment of treatment goals, the provision of helpful resources or onward referral to specialist services as appropriate. The utilisation of the 5As framework, to structure assessment and treatment interactions, is indicated when providing care to people living with obesity (Refer to the [5As for adults](#)).

A review of lymphoedema in people living with obesity addressed this question (Fife and Carter, 2008). The best screening tool is a thorough patient history and clinical examination. Physical exam should focus on determining sites of lymphoedema, excluding ascites and jaundice, and observation for any features of myxoedema. Laboratory tests if ordered should be chosen based on history and exam. They may include: serum creatinine, albumin, electrolytes, thyroid panel, liver profile and FBC. If patient presentation or history is suggestive of cardiac disease, an echocardiogram may be considered to assess the ejection fraction before commencing compression therapy, and cardiology referral arranged if appropriate.

Recommendations

OBQ2.1 Clinicians should ensure they use a non-judgmental approach when assessing PWO and conversations regarding weight should be conducted in a sensitive and respectful manner.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2.2 Clinicians should assess PWO readiness to adopt changes and assess confidence in making change.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2.3 Clinicians should ensure they use equipment that meets a safe working load as well as ensuring they follow appropriate manual handling guidelines when assessing people living with obesity.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2.4 Clinicians should complete an assessment of the individual barriers (medical, physical, psychological, socioeconomic as appropriate) affecting people living with obesity.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2.5 Clinicians should complete training in motivational interviewing to enhance communication skills for patient health behaviour support. Refer to the [HSE Health Behaviour Change training course](#) – Making Every Contact Count (MECC), [HSC Public Health Agency “Making Lives Better 2012-23” guidance document](#) and Behaviour Change Training <https://bctonline.co.uk/courses/>.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2.6 Clinicians should be aware of the complications of obesity (e.g. type 2 diabetes, hypertension, cardiovascular disease, osteoarthritis, musculoskeletal pain, dyslipidaemia and sleep apnoea) and ensure these complications are screened for and managed appropriately as part of a multidisciplinary team.

Evidence Grade: D

Strength of recommendation: Strong

OBQ2.7 Clinicians should recognise the role of waist circumference measurement with regard to health risks.

Evidence Grade: D

Strength of recommendation: Strong

Evidence Summary (cont.)

Additionally, NICE suggest that adults with a BMI of 35 or more who have been diagnosed with type 2 diabetes (within the past 10 years) be offered an expedited referral for bariatric surgery assessment. Bariatric surgery may improve QoL and reduce premature mortality in people living with both obesity and type 2 diabetes (diagnosed less than 10 years prior), by improving glycaemic control and reducing/delaying the need for medication to control diabetes. Expedited referral refers to the fact that patients do not need to have tried non-surgical measures before they are referred for bariatric assessment.

Please see NICE guidance for further information on referral to bariatric and weight management services.

The HSE Model of Care for Management of Overweight and Obesity recommends referral to Level 3 & 4 services for adults with BMI >30 and significant and uncontrolled obesity-related complications. <https://www.hse.ie/eng/about/who/cspd/ncps/obesity/model-of-care/>

Recommendations

OBQ3.1 Clinicians treating people living with lymphoedema should be aware of referral criteria and pathways to local weight management and bariatric services.

Evidence Grade: D

Strength of recommendation: Strong

OBQ3.2 Referral criteria to bariatric services include:

- Body Mass Index (BMI) >50 kg/m² * and >30 kg/m² in the ROI to level 3 & 4 services.
- BMI > 35 kg/m² plus type 2 diabetes diagnosed in the last 10 years*
- BMI of 30 or more for whom tier 2 interventions have been unsuccessful have a discussion about the choice of alternative interventions for weight management, including tier 3 services

*Referral criteria will most likely change as evidence-based recommendations encourage a shift away from BMI-centric definitions of obesity

Evidence Grade: D

Strength of recommendation: Strong

OBQ3.3 Patients may not be suitable for referral if they:

- Have active psychiatric disease that would impact on their ability to adhere to a programme
- Do not wish to participate in a medical weight management programme
- Are unable to travel to the service on a regular basis

Evidence Grade: D

Strength of recommendation: Strong

OBQ3.4 Bariatric services should employ a lymphoedema specialist to treat PWO with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

OBQ3.5 Clinicians should refer PWO to local HSE/NHS weight management services.

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Clinicians should ensure the following elements are addressed when assessing PWO with lymphoedema:

- Questions regarding PWO view of their weight and the diagnosis, and possible reasons for weight gain
- Explore eating patterns and physical activity levels
- Explore any beliefs about eating and physical activity and weight gain that are unhelpful if the person wants to lose or manage weight
- Awareness that people from certain ethnic and socioeconomic backgrounds may be at greater risk of obesity, and may have different beliefs about what is a healthy weight and different attitudes towards weight management
- History of weight management attempts and how successful they have been, and what they learned from the experience
- Physical function, balance, and a falls assessment

OBQ3: What are the referral criteria for bariatric and weight management services?

Evidence Summary

NICE guidance (2016) recommends that adults with a BMI above 50 should be offered a referral for bariatric surgery assessment.

Additionally, adults with a BMI of 30 or more for whom tier 2 interventions have been unsuccessful have a discussion about the choice of alternative interventions for weight management, including tier 3 services.

OBQ4: What is the recommended treatment for lymphoedema in people living with obesity?

Evidence Summary

There is a paucity of original research examining lymphoedema in PWO, hence conclusions specific to this population are difficult to draw. A review of existing literature recommends that CDT, including manual lymph drainage (MLD), compression bandaging, and skin care should be the mainstay of treatment of lymphoedema for PWO. This review also reports that semi-rigid devices may be more suitable than rigid compression garments which frequently don't fit the varied limb shapes and abdominal girth typically found in this population.

A prospective clinical study addressed this question (Duyur Cakit et al., 2019). The efficacy of CDT was decreased in PWO with BCRL, compared to those without obesity. Based on these findings the authors recommend that early CDT treatment should be encouraged, before fat accumulation and fibrosis occur. Obesity has been shown to decrease the effectiveness of CDT. A small prospective trial of patients with unilateral BCRL who underwent CDT found PWO had less reduction in limb volume after one year of follow up when compared to patients without obesity, suggesting obesity may exert a deleterious effect on CDT. It is unclear at precisely which BMI level this effectiveness decreases.

Recommendations

OBQ 4.1 Patients should be referred to weight management services concurrently with lymphoedema management according to the referral criteria.

Evidence Grade: D

Strength of recommendation: Strong

OBQ 4.2 CDT is safe and appropriate in the management of lymphoedema for PWO, however expectations should be managed with consideration that effectiveness of MLD reduces with increasing BMI and needs to be considered on a case by case basis.

Evidence Grade: C

Strength of recommendation: Strong

OBQ4.3 A risk assessment should be carried out before treatment for PWO and lymphoedema to ensure safety of the patient and the treating clinician.

Evidence Grade: D

Strength of recommendation: Strong

OBQ4.4 Clinicians should consider CDT for unilateral lymphoedema when BMI is > 40. If lymphoedema is bilateral and BMI > 40, modified CDT should be considered.

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

The following should be considered before commencing outpatient lymphoedema treatment in PWO:

- The patient should be able to safely transfer with minimal assistance on and off the therapy table
- The patient should be able to ambulate independently
- The patient should not have any comorbidities that affect the safety of MLD
- The patient should be encouraged to maintain a constant weight, or lose weight, during the course of MLD

OBQ5: What is the role of physical activity in people living with obesity and lymphoedema?

Evidence Summary

A systematic review of the literature including 42 exercise trials found that patients with overweight and obesity can exhibit psychological benefits from a single acute exercise session. Aerobic exercise improves well-being and reduces psychological distress, but it remains unclear which forms of exercise (aerobic, resistance or combined) lead to superior psychological outcomes. Healthcare professionals should use exercise as a tool to increase long-term participation in these patients (Elkington et al., 2017).

A systematic review of 13 trials examined adherence to exercise in PWO (Fonseca-Junior et al., 2013). The best predictors of adherence to exercise in PWO include early weight loss, lower BMI, better baseline mood, male gender and older age.

Cancer related lymphoedema

A large RCT (n = 351) of PWO with BCRL, assessing the efficacy of a weight loss programme compared to home-based programmes (The WISER Survivor RCT), addressed this question (Schmitz et al., 2019). The trial consisted of a year-long home based exercise programme of twice weekly resistance training and 180 minutes of walking per week, a 20 week programme of meal replacements and 52 weeks of lifestyle modification counselling, and a combination of the two programmes. The trial found that weight loss, home-based exercise, and combined programmes did not improve BCRL. Results suggest a supervised centre-based programme for exercise may be more beneficial than a home-based programme in improving lymphedema outcomes in patients with excess weight.

A systematic review of RCTs examined adherence to exercise in PWO with previous endometrial or breast cancer diagnosis (Rossi et al., 2018). Theory-based physical activity interventions (e.g. based on social cognitive theory) were found to be safe and feasible in this cohort. Interventions that consisted of a centre-based approach in addition to home-based activity were superior to home based interventions alone and showed moderate to large effect sizes in relation to adherence to physical activity. A Cochrane review examining the effectiveness of interventions for weight reduction in PWO with endometrial cancer concluded that there is insufficient evidence to determine effect of weight loss interventions in women with obesity and endometrial cancer (Kitson et al., 2018).

Refer to [Irish National guidance](#) and [UK National Guidance](#) on physical activity. Refer to physical activity recommendations from [Obesity Canada](#) for further guidance on physical activity.

Recommendations

OBQ5.1 Physical activity is a key component in the management of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

OBQ5.2 Physical activity prescription in the management of lymphoedema in PWO should consider patient preferences and ability.

Evidence Grade: D

Strength of recommendation: Strong

OBQ5.3 Consider group-based activity programmes in addition to home-based activities in the management of lymphoedema in PWO.

Evidence Grade: D

Strength of recommendation: Strong

OBQ6: Is there any evidence to support specific diets in the management of lymphoedema in PWO?

Evidence Summary

Nutrition is important for everyone regardless of body size, weight or health status (Brownell et al., 2010). Nutritional interventions for obesity-related lymphoedema should be nutritionally adequate, culturally acceptable and affordable for long-term adherence. Health care providers should adapt nutrition interventions and/or adjuvant therapy to meet their patients' individual values, preferences and treatment goals. However, to date, it appears that there is 'no one-size-fits-all' nutritional intervention for obesity-related lymphoedema (Koliaki et al., 2018). Nutritional interventions should be based on a collaborative care approach with a registered dietitian who has experience in obesity management and medical nutritional therapy. Dietitians can support people living with obesity who also have other chronic diseases, malnutrition, food insecurity or disordered patterns of eating (Williams et al., 2019).

Individualised medical nutritional therapy for obesity-related lymphoedema should promote a healthy relationship with food, consider the social context of eating and promote eating behaviours that are sustainable and realistic for the individual (Puhl and Heuer, 2010, Brownell et al., 2010, Ramos Salas et al., 2019). Systematic reviews and meta-analyses of RCTs have shown that individualised nutrition consultations by a registered dietitian decreases weight by an additional -1.03 kg and BMI by -0.43 kg/m² in participants with a BMI \geq 25 kg/m² compared with usual care (Williams et al., 2019).

Significant calorie restrictions can achieve short-term reductions in weight (i.e. < 12 months) but have not been shown to be sustainable long-term (i.e. > 12 months). Caloric restriction may in some individuals lead to pathophysiological drivers to promote weight gain via exaggerated hunger, appetite and decreased satiety. In addition, caloric restrictions may impair skeletal health and muscle strength, supporting the role of individualised nutritional interventions that are safe, effective and meet the values and preferences of the patient with obesity-related lymphoedema.

To date there appears to be no single-best nutritional intervention to sustain weight loss long-term, and literature continues to support the value of long-term adherence, regardless of the intervention. It is worth noting that obesity-related lymphoedema may cause irreversible lymphatic dysfunction which may not resolve with weight loss. Nevertheless, systematic reviews and meta analyses of RCTs assessing weight loss interventions for the treatment and prevention of BCRL have found that dietary advice to reduce energy intake can reduce BCRL (Schmitz, 2010).

Consequently, nutritional interventions for obesity-related lymphoedema should emphasise individualised eating patterns, food quality and a healthy relationship with food. Such interventions may consider mindfulness-based eating practices that may lower food-cravings, reduce reward-driven eating, improve body satisfaction and improve awareness of hunger and satiety (Williams et al., 2019). Future research should assess nutrition-related outcomes, health-related behaviour changes in addition to weight and body composition outcomes instead of weight loss outcomes alone across all weight ranges (Wharton et al., 2020). Due to the lack of scientific evidence it is not possible to recommend "a single best nutritional intervention plan" for people living with obesity-related lymphoedema (Koliaki et al., 2018, Williams et al., 2019, Johnston et al., 2014).

Recommendations

OBQ6.1 There is currently not enough evidence to recommend a single-best nutritional intervention plan for PWO living with lymphoedema. A patient with obesity-related lymphoedema should receive individualised medical nutritional therapy provided by a registered dietitian according to individual values, preferences and treatment goals to support a dietary approach that is safe, nutritionally adequate and can improve health-related outcomes. It is recommended to choose dietary patterns and/or food-based approaches that support their best long term adherence.

Evidence Grade: D

Strength of recommendation: Strong

OBQ7: What skin care advice is recommended for people living with obesity and lymphoedema?

Evidence Summary

PWO with lymphoedema are at increased risk of skin complications (Fife and Carter, 2008). There appears to be a lack of studies examining skin care practices in lymphoedema patients living with obesity. Therefore, the general skin care principles in lymphoedema management should be employed in the management plan for this cohort.

A review (Fife and Carter, 2008) on the subject recommends the following:

- Patients with fissured skin may benefit from products containing lactic acid. These may be helpful in treating desquamation.
- Interdry silver complex is a wicking fabric which reduces moisture and bioburden. This product may be useful in patients with multiple folds.

The All Wales Lymphoedema Obesity Policy Group (2014) guidance document recommends the following:

- Nail care should be included in the management plan for all patients. Patients who are unable to care for their nails should be referred to a podiatrist or a chiropodist.
- If a patient cannot adhere to their skin care plan of washing and applying emollients by the time of their next review (or do not have carers to do so), they should be discharged from the lymphoedema service. If and when they feel ready to comply with the treatment, they should be re-referred to the service.

Recommendations

OBQ7.1 People living with obesity should be encouraged to wash, dry and moisturise their skin daily, with particular emphasis on skin folds, as per guidance in the [general section](#) of this guideline.

Evidence Grade: D

Strength of recommendation: Strong

OBQ7.2 If a patient cannot adhere to a basic skin care plan of washing and applying emollients, they should be referred to community-based nurses or carers for additional support, and/or be referred to OT for assessment of ADLs. A referral to social services or to psychology may be beneficial where there are further concerns.

Evidence Grade: D

Strength of recommendation: Strong

OBQ8: What is the evidence for using multilayer bandaging for people living with obesity and lymphoedema?

Evidence Summary

There is a paucity of evidence available to answer this question. A case report accompanied by a brief review of the literature addressing this question was identified (Lister and Noble-Jones, 2017). The author of this paper recommended that incorporating an intense period of layered compression bandaging may improve the shape and appearance of moderate/severe lymphoedema in PWO. A dual-component compression system was just as effective as MLLB at reducing limb volume in PWO with lower limb lymphoedema.

Recommendations

OBQ8.1 MLLB should be considered in the treatment of lymphoedema in PWO if the person can tolerate it, and it does not negatively impact their QoL.

Evidence Grade: D

Strength of recommendation: Strong

OBQ 8.2 If there is evidence of vascular involvement, a vascular assessment should be carried out prior to applying MLLB. Refer to the [BLS position paper](#) (2018) on requirements for vascular assessment.

Evidence Grade: D

Strength of recommendation: Strong

OBQ9: What evidence is there to support the use of compression garments in people living with obesity and lymphoedema?

Evidence Summary

A review of the literature on obesity and lymphoedema addressed this question (Fife and Carter, 2008). The use of semi-rigid materials is suggested in patients with complex obesity. The review also recommends that an echocardiogram be carried out for any PWO with a history or physical findings suggestive of cardiac disease before the initiation of compression.

The Queensland Health Guidelines recommend that compression garments be replaced more frequently in PWO (Queensland Health, 2014). The STRIDE guidance document (Bjork and Ehmann, 2019) suggests that successful compression can be achieved in even the most complex patients with multi-morbidity, if selection of compression garments is based on the clinical presentation of lymphoedema. The same document recommends custom-made compression garments for people living with complex obesity. They also suggest use of products featuring “one-handed, side bending closure” for PWO who may not be able to bend forward to adjust regular garment straps.

Recommendations

OBQ9.1 Compression garments are safe to use in the management of lymphoedema in PWO.

Evidence Grade: D

Strength of recommendation: Strong

OBQ9.2 Compression garments should be replaced more frequently than manufacturer instructions for PWO, due to increased wear and tear of garments and size/fit changes as needed.

Evidence Grade: D

Strength of recommendation: Strong

OBQ9.3 If there is evidence of cardiovascular disease in PWO, an echocardiogram should be completed and patients with evidence of dysfunctional ejection fraction should be reviewed by a cardiologist prior to commencement of compression.

Evidence Grade: C

Strength of recommendation: Strong

OBQ 9.4 If there is difficulty wearing compression garments or donning and doffing, then suitable options should be considered e.g. semi-rigid wrapping devices, and referral to an MDT for additional support if required.

Evidence Grade: C

Strength of recommendation: Strong

OBQ10: Does CDT reduce lymphoedema in people living with obesity and lymphoedema?

Evidence Summary

A small prospective study (n = 59) compared the effectiveness of CDT in PWO compared to patients/people who are not living with obesity with BCRL and found that overall, obesity was negatively associated with the effectiveness of CDT (Duyur Cakit et al., 2019).

The above trial examined the effectiveness of CDT which comprised of MLD, IPC, multilayer compression bandaging, lymphoedema exercises, and skin care. The CDT was performed for 1 hour per day, 5 days a week for a total of 3 weeks. After 15 sessions of CDT, there was a non-statistically significant reduction in lymphoedema volume in the group of obese patients (p = 0.013), with a significant decrease in percentage excess volume (p = 0.002). In the non-obese group, after CDT there was a statistically significant reduction in both volume (p < 0.0001) and percentage excess volume (p < 0.0001). After 1 year all the patients in the group of PWO had returned to their baseline extremity volumes, whereas the group without obesity could maintain their post-CDT values of extremity volumes. The authors recommended that based on these findings, early treatment before fat accumulation and fibrosis develops should be the primary goal in the treatment of BCRL.

While the effectiveness of MLD appears to reduce significantly with increased weight, a review (Fife and Carter, 2008) of the management of lymphoedema in PWO found that MLD is safe and may be beneficial in these patients.

Recommendation

OBQ10.1 Obesity appears to significantly reduce the effectiveness of CDT and hence, should only be used in PWOs, on a case-by-case basis in combination with compression therapy, exercise and dietary management and only after a thorough risk assessment is completed including assessment of equipment.

Evidence Grade: D

Strength of recommendation: Strong

OBQ11: For PWO with lymphoedema, do any practices increase service-engagement?

Evidence Summary

There does not appear to be any evidence available to answer this question. A review of the literature on adherence to treatment in patients with BCRL found that barriers to self-management of BCRL included: complexity of treatment, symptom burden, lack of education and lack of support. Only eight studies included outcome measures of adherence to BCRL treatments, which is a major limitation for evidence in this field of study. Larger trials with greater numbers of patients are needed to establish an adequate evidence base for recommending best practice standards for improving adherence to BCRL treatment regimens (Ostby and Armer, 2015).

Recommendations

OBQ11.1 The provision of a non-judgmental, stigma-free environment is required for effective management of lymphoedema in PWO.

Evidence Grade: C

Strength of recommendation: Strong

OBQ11.2 Assessment and treatment practices should align with a behavioural treatment model in the management of lymphoedema in PWO. Clinicians should complete training in motivational interviewing to enhance communication skills for patient health-behaviour support.

Evidence Grade: C

Strength of recommendation: Strong

OBQ11.3 Lymphoedema services should work in partnership with local weight management services to ensure timely weight management, particularly for those with a BMI < 50 kg/m² who have the potential to improve their lymphoedema through weight loss.

Evidence Grade: C

Strength of recommendation: Strong

OBQ11.4 Weight management services should be aware of the influence of BMI on lymphatic dysfunction and the potential for permanent lymphatic damage as a result of obesity.

Evidence Grade: C

Strength of recommendation: Strong

OBQ12: What is the evidence supporting psychological interventions in the treatment of lymphoedema in people living with obesity?

Evidence Summary

A systematic review and meta-analysis of available literature reported that behavioural treatment strategies (e.g. goal setting, motivational interviewing, relapse prevention and cognitive restructuring) can improve adherence to lifestyle intervention programmes for people living with obesity (Burgess et al., 2017).

Recommendations

OBQ12.1 Behavioural treatment strategies should be considered part of the treatment of lymphoedema in PWO.

Evidence Grade: D

Strength of recommendation: Strong

OBQ12.2 Referral to psychology services (or GP services where appropriate) may be necessary in certain people living with lymphoedema and obesity and should be considered on a case-by-case basis.

Evidence Grade: D

Strength of recommendation: Strong

OBQ12.3 Lymphoedema services should have funded access to psychology services. Refer to appendix [LVI](#) for the All Ireland psychology pathway for patients living with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong All-Ireland Psychological

OBQ13: What is the effect of pharmacological weight loss treatment on lymphoedema?

Evidence Summary

We were unable to identify any studies directly examining the impact of pharmacological weight loss treatment on lymphoedema. However, weight loss medications can be effective in supporting weight management, which may have a positive effect on lymphoedema. Among PWO, orlistat, lorcaserin, naltrexone-bupropion, phentermine-topiramate, and liraglutide, compared with placebo, were each associated with achieving at least 5% weight-loss after 52 weeks (Khera et al., 2016). Phentermine-topiramate and liraglutide were associated with the most promising evidence of achieving at least 5% weight loss. Currently liraglutide, naltrexone-bupropion and orlistat are the only medications licenced in Ireland and the UK. Refer to the [general section](#) of this guideline for recommendations regarding the impact of pharmacological weight loss treatment on the management of lymphoedema.

Recommendation

OBQ13.1 Where indicated and licenced, weight loss medications may provide an effective method of supporting weight management in PWO with lymphoedema.

Evidence Grade: C

Strength of recommendation: Strong

OBQ14: What is the role of bariatric surgery in people living with lymphoedema and obesity?

Evidence Summary

The role of surgery in PWO and lymphoedema is best described in patients with MLL (Refer to OBQ15 and OBQ16). The authors of a large systematic review examining the surgical treatment of lymphoedema claim that obesity-related lymphoedema is a result of excessive weight and hence its treatment should focus on weight loss and recommend referring these patients for bariatric surgery. If patients continue to have lymphoedema after weight loss then other procedures may be considered and in these cases, the surgical risk would be lower due to reduced BMI (Carl et al., 2017).

NICE guidelines recommend consideration for bariatric surgery in those with:

- BMI \geq 40 kg/m², or BMI 35-40 kg/m² with other significant disease (e.g. type 2 diabetes or high blood pressure) which could be improved by weight loss
- Failure to achieve or maintain adequate, clinically beneficial weight loss after trial of all appropriate non-surgical measures
- Access to intensive management in a tier 3 service
- Medical fitness for anaesthesia and surgery
- Acceptance and understanding for the requirement of long-term follow-up

There are cases reported in the literature in which obesity-related lymphoedema was found to be non-reversible following massive weight loss post bariatric surgery (Greene et al., 2015a).

Recommendations

OBQ14.1 In PWO with lymphoedema who do not respond to conservative or medical treatment for obesity, bariatric surgery may be associated with substantial weight loss and should be considered.

Evidence Grade: B

Strength of recommendation: Strong

OBQ14.2 PWO with lymphoedema should be referred early for bariatric surgery as lymphoedema may be irreversible in those with BMI > 50 kg/m².

Evidence Grade: C

Strength of recommendation: Strong

OBQ15: How should massive localised lymphoedema (MLL) be diagnosed?

Evidence Summary

Massive localised lymphoedema (MLL) is a type of pseudosarcoma. It is a rare, benign, pseudoneoplastic complication of obesity. MLL has been described in multiple areas of the body in patients with obesity, with the medial thigh being the most common. A 2018 review of the literature on MLL yielding 53 articles (n = 105) concluded that currently there exists a low quality of evidence on MLL. The authors recommend that MLL is a clinical diagnosis and does not require biopsy in the vast majority of cases (Shavit, 2018). In cases with concern for malignancy, particularly rapidly growing masses, biopsy may be indicated (Hou et al., 2019). This viewpoint is upheld by several other case reports, suggesting the diagnosis can be made clinically by recognition of the typical presentation, underpinning the importance of increasing awareness of this condition (Tenhagen et al., 2014, Hou et al., 2019).

Other authors recommend that diagnosis of MLL requires correlation between clinical and histological findings in order to enable distinction between MLL and its various mimics (Lee et al., 2013b, Manduch et al., 2009). Magnetic resonance imaging (MRI) has also been used to diagnose MLL (Khanna et al., 2011, Hou et al., 2019) with typical presentation being described as “sharply demarcated, pedunculated mass consisting of fat partitioned by fibrous septae surrounded by a thickened dermis. There is oedema both within the mass and tracking along the subcutaneous septae in a “lace-like” fashion outwards from the pedicle, outlining large lobules of fat”. Computed tomography (CT) has also been used to exclude vascular malformations or malignancy (Hou et al., 2019).

Recommendations

OBQ15.1 If MLL is suspected, referral to a surgical team for further investigations should be considered.

Evidence Grade: B

Strength of recommendation: Strong

OBQ16: How should massive localised lymphoedema (MLL) be treated?

Evidence Summary

A number of small studies have examined the efficacy of surgery to treat massive localised lymphoedema (MLL). Positive outcomes after surgery were observed in these patients, in terms of functional ability and quality of life, with authors suggesting MLL is best treated with surgical excision (Cintra Júnior et al., 2014, Jabbar et al., 2015, Wisenbaugh et al., 2018, Siegel et al., 2016, Machol et al., 2014, Tenhagen et al., 2014, Modolin et al., 2006). Surgical resection of MLL appears to improve patient quality of life, functional capacity and optimises engagement of PWO with rehabilitation services (Cintra Júnior et al., 2014). A review of MLL including 53 reports concluded that surgery appears to be the best treatment option for MLL and may be curative but it is not without risk of complications, both intra- and post-operatively (Shavit, 2018). Standard lymphoedema treatment (e.g. compression garments and skin care) may also be beneficial in terms of improving QoL in patients living with MLL.

Recommendation

OBQ16.1 People with MLL should be referred for assessment for surgery. If surgery is not indicated, then conservative treatment should be offered to improve QoL.

Evidence Grade: C

Strength of recommendation: Strong

8. Lymphoedema in Palliative Care

The World Health Organisation (2004) has defined 'Palliative Care' as: "An approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illnesses, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychological and spiritual."

Palliative care is an approach that improves the quality of life of people facing the problems associated with life-limiting illness and supports their families. The palliative care approach focuses on the prevention and relief of suffering by means of assessing and treating pain and other physical, psychosocial or spiritual problems. The aim of palliative care is to enhance quality of life and, wherever possible to positively influence the course of illness. Palliative care also extends support to families to help them cope with their family member's illness and their own experience of grief and loss.

Lymphoedema in palliative care is a significant problem. In non-cancer patients an oedema prevalence of 85% is reported near the end of life. This may occur months prior to death and may be amenable to management during that time. The International Lymphoedema Framework (2010) states that oedema in palliative care patients is thought to represent approximately 5%-10% of all lymphoedema referrals, but this is considered an underestimate. A 2016 regional audit (Northern Ireland) found that palliative lymphoedema referrals accounted for 4.42%-6.5% of the dedicated lymphoedema team referrals, and additionally 19.96%-26.93% of the specialist palliative care physiotherapy referrals (Public Health Agency, 2018). A study in the Republic of Ireland (Real, 2015) found an incidence of 10.5% of lymphoedema at end of life. The 2019 Specialist Palliative Care Workforce Review projects that by 2024, 5,747 people in Northern Ireland will require specialist palliative care services and that there will a year-on-year increase of 49 patients.

The total number of palliative patients in the Republic is not currently available as the data is linked to service rather than the individual and patients can access many palliative services. There were 3,019 patients admitted to 15 hospice settings in 2016 which would equate to approximately 300 palliative patients with lymphoedema in a hospice setting each year in Ireland (HSE, 2018).

Lymphoedema due to advanced cancer or oedema at the end of life

Patients with lymphoedema due to advanced disease and who require palliative care can have complex needs. Lymphoedema can produce distressing and debilitating symptoms. Extensive generalised oedema affecting more than half of the body may be due to multiple factors such as immobility, advanced disease and hypoproteinaemia. Swelling of the limbs can be severe with the involvement of adjacent areas such as the trunk, genitalia, and digits. Other symptoms such as severe skin changes, fragile skin and lymphorrhoea affect lifestyle and function. Patients with advanced disease may not be able to tolerate a full programme of assessment and treatment. A modified palliative approach may be more appropriate in which assessment techniques are modified and individual treatments are selected to ease specific symptoms.

Oedema may primarily be from lymphatic blockage or from a variety of medical causes including venous hypertension and hypoproteinaemia (often termed chronic oedema) or a mixture of both (mixed aetiology). Symptoms may develop rapidly and cause acute distress to the patient or oedema may develop more slowly. In most cases, it is a symptom that can be relieved and actively managed even where disease-related treatments have been exhausted.

A multi-disciplinary approach which encompasses the needs and preferences of the patient is vital. Lymphoedema practitioners must work in tandem with other health care professionals,

negotiating a shared plan of care and treatment goals. Supportive treatment and intervention can help reduce the distressing and often debilitating symptoms that affect the patient's functional ability and quality of life, regardless of the disease status. Particular individual risks, such as increased risk of cellulitis where lymphorrhoea is present should be emphasised.

Patients who present with advanced disease may not always tolerate full decongestive therapy, and therefore, the assessment, intervention, and overall management of lymphoedema in palliative care may be modified according to the needs of the patient. As the overall status of the patient in the palliative phase can potentially deteriorate quickly at any given time, the lymphoedema management must be implemented in a timely manner and regularly reviewed by a specialist lymphoedema therapist.

PALQ1: How should oedema be diagnosed in palliative care patients?

Evidence Summary

Three reviews (Beck et al., 2012, Cheville et al., 2014, Towers, 2010) addressed this question. There does not appear to be one recommended method of diagnosing oedema in patients in the palliative care setting. Oedema is often multifactorial in patients with advanced cancer so a thorough assessment is required to assess the cause of the oedema, which in turn will assist in determining appropriate treatment. The following causes of oedema should be considered when diagnosing oedema in palliative care patients:

- Malignant involvement or infiltration of lymphatic structures
- Lymphatic insufficiency
- Venous obstruction (thrombosis, compression by tumour)
- Decreased albumin (anorexia/cachexia of advanced cancer, ascites with repeated paracentesis)
- Renal or hepatic failure
- Cardiac failure
- Dependent limb, immobility, neurological deficit
- Effects of drug or cytotoxic chemotherapy interventions (e.g. docetaxel)
- Infection
- Previous surgery or radiotherapy

In terms of assessment tools, in a retrospective chart review of palliative care patients with lymphoedema, circumferential measurement was the most commonly used assessment tool (Cobbe et al., 2017). The next most commonly used assessment tool in this cohort was skin descriptors e.g. clinician description of skin colour and texture. Photographs, while not used commonly, are very useful clinically for monitoring changes in limb volume and skin appearance.

Recommendations

PALQ1.1 The diagnosis of oedema in patients with palliative care needs should involve MDT discussion.

Evidence Grade: D

Strength of recommendation: Strong

PALQ1.2 A thorough medical assessment, as tolerated by the patient, should be carried out to identify underlying causes of oedema in palliative care patients.

Evidence Grade: D

Strength of recommendation: Strong

PALQ1.3 There is no single recommended method of diagnosing oedema in palliative care patients. Circumferential measurements and skin descriptors (photos, written descriptions of condition) may be used in the measurement of the extent of oedema and resulting sequelae. Please see the [general section](#) for advice on the comprehensive assessment of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

PALQ1.4 The methods and extent of assessment should be determined by the clinical presentation of the patient and their individual prognosis.

Evidence Grade: D

Strength of recommendation: Strong

PALQ2: How should oedema be treated in palliative care patients?

Evidence Summary

A systematic review (Beck et al., 2012), a prospective pilot study (Cobbe et al., 2018), a cross-sectional study (Gradalski, 2019) and a retrospective study (Cobbe et al., 2017) addressed this question. Limb oedema in patients with advanced cancer can be treated occasionally with decongestive or supportive physiotherapy. The choice of therapy employed should depend on the patient prognosis, symptoms, stage of oedema and disease progression (Gradalski, 2019).

A review of the treatment of lymphoedema in palliative care patients (Towers, 2010) outlines the recommended adaptations to treatment in this group. The following amendments to bandaging in palliative patients are outlined below.

Standard bandaging	Palliative bandaging
Full standard pressure	Reduced pressure
Multilayer bandages	Consider fewer layers
24-hour bandaging during intensive phase	Intensive treatment for lymphorrhoea may require frequent reapplication of bandages
Foam padding used	Soft padding generally better tolerated
Transition to compression garments	May transition to lighter support bandaging, or to compression garments or continue palliative bandaging

A review of the treatment of lymphoedema in palliative care patients (Towers, 2010) outlines the recommended adaptations to treatment in this group. The following amendments to bandaging in palliative patients are outlined below.

Standard CDT	Palliative CDT
Goals to reduce swelling, transition to garments, life-long maintenance, self-care	Goals to provide comfort, support, relief of symptoms, maintain function, include caregivers in care
Four elements of CDT	CDT elements may be modified or omitted
Two distinct phases of treatment	Less distinction between phases of treatment
Definitive contraindications to treatment	Contraindications now relative

Recommendations

PALQ2.1 The treatment of oedema in palliative care patients should be based on their clinical presentation, prognosis and the individual patient's main concerns, priorities and goals.

Evidence Grade: D

Strength of recommendation: Strong

PALQ2.2 Modification of CDT should be considered in palliative care patients, based on their clinical presentation.

Evidence Grade: D

Strength of recommendation: Strong

PALQ2.3 Clinicians should consider the burden of treatment versus the benefits when considering treatment options for oedema in palliative care.

Evidence Grade: D

Strength of recommendation: Strong

PALQ2.4 When treating lower limb or abdominal oedema in palliative care patients, clinicians should monitor for evidence of genital and abdominal oedema, and lymphorrhoea.

Evidence Grade: D

Strength of recommendation: Strong

PALQ2.5 Clinicians should consider and prioritise skin care and positioning, and liaise with nursing and tissue viability nursing (TVN) as required. Refer to GQ41-44 on skin care for further guidance.

Evidence Grade: D

Strength of recommendation: Strong

PALQ2.6 Clinicians may consider all treatment options available, including wraps, laser, pneumatic pumps and oscillators. Refer to the [general section](#) of this guideline for a complete list of treatment options.

Evidence Grade: D

Strength of recommendation: Strong

PALQ3: Are abdominal binders effective for reducing oedema in palliative care patients with abdominal tumours?

Evidence Summary

There were no studies identified which addressed this question. There are studies that show the benefit of abdominal binders post major abdominal surgery, which show increased ambulation/mobility and reduced post-operative pain, however these results cannot be extrapolated to support the use of binders in the palliative care setting.

The use of abdominal binders has been anecdotally shown to provide symptomatic relief in palliative care patients with abdominal oedema, when used by experienced lymphoedema therapists. Patients with certain medical co-morbidities specifically cardiac and blood pressure conditions may not be suitable candidates for abdominal binders.

A review of the treatment of lymphoedema in palliative care patients (Towers, 2010) outlines the recommended adaptations to treatment in this group. The following amendments to bandaging in palliative patients are outlined below.

Recommendation

PALQ3.1 Abdominal binders may be considered in the treatment of abdominal oedema in palliative care patients, and should only be applied by experienced lymphoedema clinicians.

Evidence Grade: D

Strength of recommendation: Strong

PALQ4: What treatments are recommended for palliative care patients with head and neck swelling secondary to cancer?

Evidence Summary

There was no trial evidence available to answer this question. Expert opinion is that modified CDT should be considered in the treatment of palliative care patients with head and neck swelling. Other treatment modalities may be considered including Kinesio Taping and compression garments.

Clinicians should consider that steroid-induced head and neck swelling is not amenable to CDT.

Refer to the [BLS Head and Neck education document](#) for further guidance.

Recommendations

PALQ4.1 Modified CDT should be considered in the treatment of palliative care patients with head and neck swelling. Other treatment modalities may be considered including positioning, Kinesio taping, compression garments, LymphTouch and laser therapy. Refer to the general section of this guideline for full a selection of treatment options.

Evidence Grade: D

Strength of recommendation: Strong

PALQ4.2 Steroid-induced head and neck swelling is not amenable to CDT, but skin condition and comfort may be improved with light massage and Kinesio tape.

Evidence Grade: D

Strength of recommendation: Strong

PALQ5: Is there evidence that multilayer compression bandaging is effective in the treatment of lymphorrhoea in palliative care patients?

Evidence Summary

While there is a distinct lack of trial evidence available to answer this question, case studies as well as expert opinion support the use of multilayer compression bandaging in patients with lymphorrhoea, and it is generally considered safe and effective for these patients (Towers, 2010). In some patients with severe lymphorrhoea, lower limb bandaging may be helpful even if it may cause some proximal swelling, a reported side effect of treatment. It is therefore important to involve the patient (and medical team) in decision-making. Lymphorrhoea usually responds well to continuous compression bandaging. Frequent changes in bandaging may be required, often more than once a day (Regnard et al., 1997). Non-adherent dressing materials such as paraffin-impregnated gauze may be beneficial at the leaking area (Renshaw, 2007).

Each patient should be assessed individually and the level of compression should be determined by the clinical findings and the priorities of the patient and the agreed goals of management. Pressure levels applied may need to be reduced or more gradually increased compared to standard care. It may be advisable to use fewer bandage layers and lighter materials for bandaging e.g. tubigrip. Community lymphoedema therapists in some regions may not have access to all types of dressing materials and may need to involve hospital or community and tissue viability nurses.

Recommendations

PALQ5.1 Short-stretch or inelastic compression bandaging is effective in the treatment of lymphorrhoea in palliative care patients and should be considered as a first-line treatment, except when actively dying. Refer to [Lymphoedema Network Wales Lymphorrhoea Pathway](#) for further advice on lymphorrhoea management. (Appendix I.V)

Evidence Grade: D

Strength of recommendation: Strong

PALQ5.2 Short stretch or inelastic compression bandaging should be commenced as soon as possible in the treatment of lymphorrhoea in palliative care patients to prevent skin maceration.

Evidence Grade: D

Strength of recommendation: Strong

PALQ5.3 Clinicians should consider liaising with the multidisciplinary team when treating palliative care patients with lymphorrhoea.

Evidence Grade: D

Strength of recommendation: Strong

PALQ5.4 Clinicians should consider liaising with tissue viability nurses when treating palliative care patients with lymphorrhoea, who have compromised skin integrity or with open wounds.

Evidence Grade: D

Strength of recommendation: Strong

PALQ6: Is radiotherapy effective in reducing limb swelling in palliative care patients with lymphoedema?

Evidence Summary

There does not appear to be any trial evidence available to answer this question. A review of the management of lymphoedema in palliative care patients recommends that palliative chemotherapy and radiotherapy may reduce limb swelling in select patients by reducing metastatic tumour burden (Towers, 2010).

Some centres in Ireland have adopted radiotherapy where tumours/metastases are invading lymphatics.

Recommendations

PALQ6.1 Palliative radiotherapy may reduce lymphatic obstruction attributable to tumour burden, so may be considered as a treatment option. This should be discussed with the MDT.

Evidence Grade: D

Strength of recommendation: Strong

PALQ7: Is MLD and/or compression bandaging safe in patients with open wounds?

Evidence Summary

While there do not appear to be any trials examining the safety of MLD or bandaging in patients with open wounds, one review (Towers, 2010) states that MLD and bandaging should be adapted in cases where tumours infiltrate the skin or subcutaneous tissue in palliative care patients. Open wounds, however, do not constitute an absolute contraindication to MLD or compression bandaging in these patients.

Recommendations

PALQ7.1 Open wounds do not constitute an absolute contraindication to MLD or compression bandaging in palliative care patients. Modified MLD should be considered in the treatment of oedema in palliative care patients with open wounds. Clinicians should evaluate the affected area regularly and observe for any localised deterioration.

Evidence Grade: D

Strength of recommendation: Strong

PALQ7.2 Clinicians should liaise with tissue viability services and/or nursing staff in the treatment of palliative care patients with open wounds.

Evidence Grade: D

Strength of recommendation: Strong

PALQ8: What is the role of physical activity in the management of palliative patients with lymphoedema/oedema?

Evidence Summary

There were no trials available to answer the question. When designing exercise protocols for palliative care patients, consideration should be given to pain, strength limitations, presence of bony metastasis, risk of pathological fracture and reduced exercise tolerance (Towers, 2010). Patients should be encouraged to move within their own limitations and comfort levels. Active and active-assisted exercises may be beneficial.

Recommendations

PALQ8.1 Prescribed physical activity may be beneficial for palliative care patients with oedema within the limits of patient pain and tolerance.

Evidence Grade: D

Strength of recommendation: Strong

PALQ8.2 Assistive exercise devices, mobility aids or carer assistance may be required by patients when carrying out physical activity.

Evidence Grade: D

Strength of recommendation: Strong

PALQ9: What is the effectiveness of paracentesis in the treatment of lower limb oedema?

Evidence Summary

There was no trial evidence available to answer this question. Expert opinion supports the use of abdominal paracentesis for the treatment of ascites which may provide an additional treatment benefit by reducing lower limb oedema.

Recommendation

PALQ9.1 Paracentesis as a treatment for ascites may provide a secondary treatment benefit of reducing lower limb oedema.

Evidence Grade: D

Strength of recommendation: Strong

PALQ10: How should genital oedema in palliative care patients be treated?

Evidence Summary

While there was no trial evidence available to answer this question, expert opinion supports the use of modified manual techniques, positioning, compression and Kinesio taping in the treatment of genital oedema for palliative care patients.

Recommendations

PALQ10.1 Any patient presenting with lower limb oedema/ascites should be asked specifically about genital oedema. Refer to appendix [II.IV.I](#) - [II.IV.III](#) for the Genital Oedema Assessment forms.

Evidence Grade: D

Strength of recommendation: Strong

PALQ10.2 The use of modified MLD techniques, positioning, compression, skin care and Kinesio taping should be considered in the treatment of genital oedema in palliative care patients.

Evidence Grade: D

Strength of recommendation: Strong

PALQ10.3 In the case of complex genital oedema, the treating clinician should always discuss treatment options with the MDT.

Evidence Grade: D

Strength of recommendation: Strong

PALQ11: What outcome measures should be used to monitor response to treatment in palliative care patients with lymphoedema?

Evidence Summary

There was no specific evidence examining outcome measures in palliative care patients with lymphoedema. Expert opinion supports the use of symptom assessment scales, QoL assessment scales, limb volume and skin integrity in measuring outcomes.

Examples of palliative care outcome measures:

- McGill Quality of Life Questionnaire (MGQOL)
- Edmonton Symptom Assessment System (ESAS)
- Palliative Care Outcome Scale (POS)
- Palliative Care Performance Scale
- Karnofsky Performance Scale
- Palliative Care Outcomes Collaboration (PCOC)

Recommendations

PALQ11.1 In monitoring treatment response, the use of symptom assessment scales and QoL assessment scales and assessment of limb volume and skin integrity should be considered.

Evidence Grade: D

Strength of recommendation: Strong

PALQ11.2 The choice of outcome measure should depend on patient medical status and treatment goals.

Evidence Grade: D

Strength of recommendation: Strong

PALQ12: Can oedema volume be used as a prognostic indicator for palliative patients?

Evidence Summary

The presence of oedema, but not oedema volume, is a well-established prognostic factor for palliative care patients (Cui et al., 2014, Kalpakidou et al., 2018, Chiang et al., 2009). While there are no studies examining the impact of oedema volume on prognosis, the Chuang Prognostic Score (CPS) includes limb oedema volume in palliative care patients (Stone and Lund, 2007). While this score does not incorporate specific volumes as predictors of outcome, it assigns a score of 1-3 for pitting oedema of < 1-2 finger breadth, ½-1 finger breadth or > 1 finger breadth.

Recommendations

PALQ12.1 The presence of oedema in palliative care patients is a negative prognostic factor, however the degree of oedema is not currently considered prognostic.

Evidence Grade: D

Strength of recommendation: Strong

9. Lymphoedema Education

All health care professionals who come into contact with patients with, or at risk of developing lymphoedema should be aware of the signs and symptoms of lymphoedema, the relevant diagnostic and current treatment pathways, and availability of local resources (Yarmohammadi et al., 2021). Risk reduction education can reduce the risk of developing lymphoedema and allow the creation of pre-treatment lymphoedema educational protocols. Early diagnosis and referral will optimise outcomes. Relevant healthcare providers should receive education regarding lymphoedema and risk reduction, and should communicate this knowledge to patients on a consistent basis both in verbal and in written formats. This includes all at risk groups; older people's services, ulcer/wound clinics, obesity services as well as oncology.

Pre-treatment (e.g. pre cancer surgery, neoadjuvant chemotherapy or radiotherapy) lymphoedema education may improve patient recall of information received (Ridner, 2006). The format could be a cancer-related prehabilitation programme for those with known risk of lymphoedema, or a self-management lymphoedema service adjunct (in group or individual formats) to support lymphoedema management for all patients already diagnosed with the condition and prior to starting their next phase of treatment.

It is internationally recognised that education on the lymphatic system and lymphoedema is poorly addressed in health professional undergraduate education curricula. Undergraduate education is essential to promote risk reduction and an understanding of lymphoedema management and pathways. From a Republic of Ireland perspective, a review of seven universities in 2017 revealed that only four undergraduate health professional courses (three physiotherapy and one occupational therapy) had specific lymphoedema content in their curriculum, varying from 1 hour to 18 hours (Health Service Executive, 2018). Current education standards leading to nurse registration also did not include lymphoedema education as a component, however one oncology post-graduate nursing course included a one-hour lecture in oncology-related lymphoedema.

The Lymphoedema Network Northern Ireland (LNNI) introduced an education strategy that included several training levels, including specialist training opportunities for lymphoedema clinicians, general training for nursing and allied health professional undergraduate staff (via the Ulster University) regarding prevention and diagnosis of lymphoedema. Undergraduate awareness training remains to be implemented in medical school undergraduate curricula. For further details please see the [LNNI website](#).

The LNNI education strategy was credited with the early identification of many cases, resulting in an increase in referrals with consequent better patient outcomes due to early intervention. The increased awareness also resulted in more non-oncology referrals and subsequent early initiation of treatment. The composition of patient referrals changed from 90% oncology-related lymphoedema referrals in 2008 to 54% in 2020, achieving a balanced mix commensurate with population health planned predictions. The benefits and learning from the Northern Ireland approach should be explored with a view to transferring these to the health service in the Republic of Ireland.

To address this education deficit, the International Lymphoedema Framework has [identified Lymphoedema Education Benchmark Statements \(LEBS\)](#) that reflect international consensus regarding knowledge, that any person with, or at risk of lymphoedema, might reasonably expect from newly qualified health professionals. Implementation of these standards to relevant All-Ireland undergraduate healthcare professional programmes would foster global consistency and governance in relation to lymphoedema education. Within this framework, there are broadly three levels of education required to ensure that lymphoedema is diagnosed, treated and maintained, according to best practice guidelines.

Education Level	Health care professional	Education required
Level 1	All health care professionals (HCPs)	Basic awareness education for all HCPs including those working in care homes, should be provided at undergraduate level. There are education benchmark statements for lymphoedema produced by the ILF (2017). These are available on the ILF website for reference and are a simple and inexpensive way to integrate the information into a curriculum.
	GPs	Early diagnosis and knowledge of appropriate pathways for lymphoedema, lipoedema and cellulitis. There are currently online GP education tutorials that can be assessed through e-learning modules.
	Oncology services	Education on screening tools, basic preventative care and referral pathways.
	HCPs in other high prevalence areas	Basic preventative care, awareness and knowledge of the referral pathway.
Level 1	HCPs working in primary care clinics	One-day introductory course to include; measurement of limb volumes, prescription of compression garments and provision of support and education.
	HCPs working in wound care or chronic oedema	One-day course to manage and treat chronic oedema to include assessment, compression bandaging and exercise for the lower limb.
Level 3	Lymphoedema therapist	All therapists must have completed and passed an internationally recognised certification course including a minimum of 135 hours duration. Supervision and professional CPD will dictate varied competency requirements.

Level 1 is for all HCPs for basic awareness of lymphoedema
Level 2 is for HCPs treating non-complex lymphoedema
Level 3 is certification for HCPs to become lymphoedema clinicians

Level 3 is an internationally recognised certification course of a minimum of 135 hours duration. There are positive opportunities for Level 3 clinicians to provide teaching and mentoring for staff to be trained to Levels 1 and 2. This would help to improve professional working relationships and provide consistency in content and continuity as changes in staff occur. Refresher courses may be required for therapists that are working in specific areas of lymphoedema, especially in the case of lone practitioners, to enable them to work in all areas of the service. Initially new staff will require a formal update after two years of practice but thereafter working in a multi-professional team with peer support, future reviews may be 3-5 yearly. The competencies and methods of learning for each level of education should be agreed with the development of the standards of care and can be adapted from the recommendations of the ILF (Sneddon, 2007). Partnerships between universities and lymphoedema services should continue to be established to identify research needs and education opportunities for the development of services.

As healthcare professionals play a pivotal role in meeting the needs of individuals with lymphoedema, the focus on patient education should include identification of risk factors, early detection, risk reduction measures and self-care activities using appropriate and innovative technology and strategies to deliver timely information. The service user LNNI education level was directed to inform and empower patients who were at risk, and includes tailored information

available via the [LNNI website and LNNI App](#). There are many educational resources available for people living with lymphoedema including those from the formal support groups, but there is a lack of consistency. Utilising a standardised approach in lymphoedema education, especially in the area of risk reduction, is advocated and is reflective of current developments to promote ongoing evidence based practice.

EQ1: What are the educational needs of healthcare staff in relation to lymphoedema?

Evidence Summary

There appears to be a lack of evidence examining teaching strategies in the education of health professionals treating patients with lymphoedema. According to a descriptive institutional report (Schaverien et al., 2020) in order to build a comprehensive academic programme, institutional support and involvement of key stakeholders is essential. Modern academic centres should strive to establish MDT clinical programmes and develop coordinated care pathways and conduct clinical research including clinical trials.

There is a strong body of evidence in education that change-in-practice requires there to be an assessment of understanding and competence, not just knowledge. This supports the recommendation for a substantial assessed course for practitioners, but it also suggests that short, non-assessed provision will have little impact.

Recommendations

EQ1.1 All health care professionals should have an awareness of lymphoedema in order to be able to:

- Recognise/diagnose lymphoedema
- Ensure appropriate referral of patients with suspected lymphoedema
- Provide simple advice in relation to lymphoedema management

Evidence Grade: D

Strength of recommendation: Strong

EQ1.2 For non-specialist healthcare staff treating patients with non-complex lymphoedema, comprehensive education and training programmes including management of skin, exercise and compression should be developed.

Evidence Grade: D

Strength of recommendation: Strong

EQ1.3 For specialist staff treating patients with lymphoedema, an approved comprehensive education and training programme, as recognised by the ILF and BLS, should be completed.

Evidence Grade: D

Strength of recommendation: Strong

EQ1.4 Lymphoedema clinicians have a professional requirement to maintain CPD training, and undergo appraisal and goal setting as a part of ongoing supervision and coaching programmes in lymphoedema care, in line with their own professional governing body.

Evidence Grade: D

Strength of recommendation: Strong

EQ1.5 The use of motivational interviewing and coaching is recommended as an integral component of a comprehensive lymphoedema assessment.

Evidence Grade: D

Strength of recommendation: Strong

EQ2: What teaching strategies assist in meeting the educational needs of patients with lymphoedema?

Evidence Summary

It has been shown that knowledge impacts on a patient's ability to carry out self-management tasks. Therefore, a lack of knowledge regarding rationale of treatment, and expected treatment outcomes, inevitably affects perceived treatment benefit. Lymphoedema education to date has focused on advice to at-risk patients and practical aspects of treatment, with little mention of expected treatment outcomes or how to monitor symptoms. Findings from a qualitative study (Jefferies et al., 2016) suggest that different types of knowledge are required at different stages of the lymphoedema journey e.g. novice patients require information about how and why they should carry out treatment. Once the basic concepts of lymphoedema management are gained, knowing how to recognise and interpret symptom changes, and adjust treatment accordingly appears more beneficial.

Self-Management Approaches

A small pilot study (Ridner et al., 2014) (n = 39) tested the psychological hypothesis of self-regulation theory, which states that objective self-measurement of physiological conditions is necessary to promote self-regulation/self-care. The self-monitored group had more days of garment use (p = 0.005) and more candidates who remained stable after the self-monitoring period ended. This study concluded that objective self-monitoring of arms using BIS is possible, and self-monitoring may positively impact self-care behaviours. Highly symptomatic patients may require coaching or other psychological support to improve their self-care. Studies that combine cognitive behavioural therapy components along with self-measurement should be considered as potential interventions to impact lymphoedema self-care.

One prospective study (Sherman and Koelmeyer, 2013) highlighted the importance of underlying beliefs as determinants of whether a patient, who is informed and knowledgeable about lymphoedema risk and its management, will undertake the recommended risk management actions. In addition to raising lymphoedema awareness and its risk management, health professionals should instil positive beliefs in patients regarding the control they have over their condition through education, early detection and early treatment approaches.

A small pilot study (Gradalski and Ochalek, 2020) examined the efficacy of trained lymphoedema therapists educating lay carers (n = 24) to bandage patients with non-complex lymphoedema. The results of this small study showed no statistically significant difference in treatment outcomes in the group who had bandages fitted by lay carers versus the group who had their bandages fitted by therapists. Educating lay carers may therefore provide a simple clinically effective solution for non-complex lymphoedema management with lower associated healthcare costs.

Technological/E-Health Approaches

Various educational models have recently been trialled to assist patients with lymphoedema management. The utility of a web-based multimedia intervention (WBMI) compared to an educational pamphlet for educating patients on BCRL was assessed in one RCT (Ridner et al., 2020). The WBMI was perceived by patients as providing better self-care information than educational pamphlets. Statistically significant improvements in bio-behavioural symptoms (e.g. mood), were also reported by those using the web-based intervention. A second RCT (Omidi et al., 2020) compared group-based education and social network-based self-management education in the clinic and via Telegram™ messenger channel, respectively. This study reported that despite the social network-based education method being effective, the group-based education method appeared more beneficial in patients with BCRL. The authors highlight the need to carry out cost-effectiveness studies before implementing these educational modalities in patients with BCRL.

A smaller prospective study (Okutsu and Koiyabashi, 2014) of female genital cancer surgery patients (stages I and II) compared routine self-care support group (control group) to a mobile telephone-assisted support group (intervention group). Compared with routine self-care support, the mobile telephone-assisted support appeared to lead to statistically significant improvements in lymphoedema patients' QoL and mental health status, as well as improvements in their self-care behaviours.

A prospective cohort study (Fu et al., 2016) examined the efficacy of an educational system called The-Optimal-Lymph-Flow health IT system (TOLF) in patients (n = 355) with lymphoedema. TOLF is a patient web-and-mobile-based educational and behavioural mobile health intervention focusing on safe, innovative, electronic assessment and self-care strategies for lymphoedema management. Patients were very satisfied with the mobile health self-care interventions with 90% rating the system as having no problems with usability; and the majority of participants (96.6%) strongly agreed that the system was easy to use and effective in assisting patients to learn about lymphoedema, symptoms and self-care strategies.

Recommendations

EQ2.1 Patient education should be tailored to patients' individual learning styles and communication ability, taking into consideration transition from novice, to expert, and adjusting to living with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.2 Patients who have the capacity and capability should be directed to access recommended [available online resources](#) to assist in the management of their condition.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.3 A group education approach should be available to support patients living with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.4 Patients who have the capacity and capability should be encouraged to utilise phone applications (e.g. [LNNI Lymphoedema app](#)) to support self-management of lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.5 Virtual contact (e.g. telephone/video calls) should be available during working hours to provide assistance to support patients' self-care interventions where feasible.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.6 Further investigation of existing online resources should be undertaken to support self-monitoring and supported care, and consideration should be given to embedding this into practice.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.7 Patient education materials should be available in multiple languages and translation services should be available to all patients presenting with lymphoedema. Alternative formats should be available for those additional accessibility needs.

Evidence Grade: D

Strength of recommendation: Strong

EQ2.8 Easy-read patient information documents should be available to all lymphoedema service users. Refer to [appendix I.VIII](#) for "Easy read; 5 key points of lymphoedema care".

Evidence Grade: D

Strength of recommendation: Strong



Good Practice Point

Patient education delivery

Written information should be provided to patients prior to attending appointments with lymphoedema clinicians if possible.

Patients should be provided with written patient information and available supports and resources to assist in the management of their condition.

There should be gender-neutral educational information available to suit all patient needs.

There should be easy-read options available to suit all patient needs.

EQ3: What are the educational barriers to concordance to treatment for patients with lymphoedema?

Evidence Summary

A number of qualitative studies have assessed barriers to concordance in patients with lymphoedema (Ostby et al., 2018, Radina et al., 2014, Kwan et al., 2012, Ridner et al., 2011, Alcorso and Sherman, 2016). Findings from these studies identified physiological, psychological, and psychosocial factors as barriers to successful lymphoedema self-management. Lack of education regarding lymphoedema treatment and risk-reduction activities was identified as one of the main barriers. In addition, more than half of patients surveyed defined support as "prescriptions" and "referrals"; therefore, it is unclear whether patients were exposed to support other than medical treatment.

Recommendations

EQ3.1 Clinicians should be aware of the barriers to concordance to lymphoedema treatment that exist and should address these barriers where possible.

Evidence Grade: D

Strength of recommendation: Strong

EQ3.2 Educational material provided should be adapted to meet patient's individual needs and should be tailored to maximise concordance based on individual barriers identified. (Refer to [LNNI website](#) for intellectual disability easy-read documents).

Evidence Grade: D

Strength of recommendation: Strong

EQ3.3 Risk reducing activities should be addressed at select moments such as after a cancer diagnosis or when treating patients awaiting bariatric surgery and these opportunities must be optimised by clinicians. Prehabilitation education opportunities should be developed to include risk reduction information for tumour-specific groups.

Evidence Grade: D

Strength of recommendation: Strong

EQ4: What additional training should lymphoedema clinicians have prior to treating children with lymphoedema?

Evidence Summary

While no specific research is available to answer this question, the Children's Lymphoedema Special Interest Group (2016) has published the following recommendations, which this guideline development group endorses:

- All HCPs involved in care of a child or young person with lymphoedema must have completed Level 3 Child Safeguarding Training with consideration to additional training regarding consent, parental responsibility, Fraser guidelines, confidentiality and, if providing treatment for children with communication or learning difficulties, communication skills training.
- HCPs who are experienced lymphoedema practitioners need not have a specific paediatric professional qualification but must work within the limits of their knowledge, skills, and competence.
- The lymphoedema service should provide specialist assessment conducted by a HCP who is an experienced lymphoedema practitioner with a specialist lymphoedema qualification.
- The service must provide coordinated multi-agency care for investigations or medical treatment associated with the lymphoedema if required.

Recommendations

EQ4.1 All lymphoedema clinicians must have completed child safeguarding training prior to treating children with lymphoedema.

Evidence Grade: D

Strength of recommendation: Strong

EQ4.2 All lymphoedema clinicians who work with children should work within their competence and if necessary, collaborate and upskill with an experienced practitioner.

Evidence Grade: D

Strength of recommendation: Strong

EQ5: What education is required by HCPs to apply compression garments?

Evidence Summary

A recent multicentre study (Hall et al., 2019) aimed to assess if non-complex compression garment selection, fitting and monitoring could be safely and successfully carried out by generalist clinicians without specific and recognised lymphoedema training for this task i.e. by physiotherapists and occupational therapists. The provision of these clinical tasks was found to be safe, effective and regarded in a positive light by staff and patients. The authors report that their service model, which included resources and training, improved access to lymphoedema services particularly in rural areas.

Recommendations

EQ5.1 Education programmes should be developed for non-specialist healthcare professionals to undergo training in non-complex compression garment measurement and assessment techniques, inclusive of supervised practice and continued competency assessment.

Evidence Grade: D

Strength of recommendation: Strong

EQ5.2 All healthcare professionals treating non-complex lymphoedema should work as part of a multidisciplinary team and have access to a lymphoedema specialist.

Evidence Grade: D

Strength of recommendation: Strong

EQ5.3 Education programmes should be developed for healthcare professionals to enable them to prescribe compression garments. All services should ensure that staff progress towards this qualification.

Evidence Grade: D

Strength of recommendation: Strong

EQ5.4 Advanced skills are required for prescribing compression garments for complex presentations and should be carried out by a lymphoedema clinician.

Evidence Grade: D

Strength of recommendation: Strong

EQ6: Which HCPs should re-measure patients for replacement compression garments?

Evidence Summary

A recent study (Hall et al., 2019) found that generalists such as physiotherapists and occupational therapists, without certified lymphoedema qualifications, were safe and efficient in selecting, fitting and monitoring compression garment use in patients with stable and non-complex lymphoedema, when supplied with adequate resources and education. These findings are in line with guidance from the Queensland Health Guideline which recommends that "All clinicians prescribing initial and changed compression garments are recommended to have level one lymphoedema training with additional continuing professional development in lymphoedema as outlined by the National Lymphoedema Practitioner".

Recommendations

EQ6.1 Healthcare professionals can re-measure compression garments for stable and non-complex oedema, once they have completed an approved measurement education session.

Evidence Grade: D

Strength of recommendation: Strong

EQ6.2 In cases where measurements change significantly, re-referral should be made to the specialist lymphoedema centre by the treating healthcare professional.

Evidence Grade: D

Strength of recommendation: Strong

EQ6.3 All complex and non-resolving cases should be referred to the specialist lymphoedema centre for garment re-measurement.

Evidence Grade: D

Strength of recommendation: Strong

EQ7: Do certified lymphoedema therapists (CLT) require a background in a health profession?

Evidence Summary

There is no literature available to answer this question, however the New South Wales (NSW) lymphoedema guideline recommends that: "People identified with symptoms or signs of lymphoedema should receive a timely clinical assessment by an appropriately trained health professional: a medical practitioner or an accredited lymphoedema practitioner (such as a registered nurse, physiotherapist or occupational therapist) who has completed an accredited lymphoedema training course."

Several international bodies have issued guidance on the topic. The Australasian Lymphology Association (ALA) require that accredited Lymphoedema Practitioners must have a range of health professional backgrounds. This body states that "Undergraduate and/or post-graduate qualification in a relevant health profession - the ALA Constitution deems eligible professions as: medical practitioners, nurses (Division 1), occupational therapists, physiotherapists and remedial massage therapists."

The ALERT Education programme, issues similar guidance stating those wishing to partake in a Lymphoedema accreditation course should have: "Australian Qualification Framework (AQF) level 7 bachelor's qualification in a health degree or recognised equivalent. For Registered Nurses that received their qualification as "Registered Nurse" via a certificate prior to University Nursing programmes being commonplace, this qualification is accepted. Nurses must be Division 1 or Registered Nurses. ALERT welcomes interdisciplinary learning, each individual health professional needs to ensure that they practice within their scope of practice. Please seek guidance from your professional board and from your insurance company in regards to which modalities you are covered to deliver."

The National Lymphoedema Network position statement on the training of lymphoedema therapists (2013) states their minimum requirements for training in lymphoedema treatment include:

"• Practitioners treating patients with lymphoedema will successfully have completed a minimum of 135 hours of Complete Decongestive Therapy coursework. The CDT entry level curriculum should be presented in no more than four integrated courses from a single training programme. Unrelated review, advanced or supplemental courses do not satisfy the entry level requirement of intentional course linkage.

• It is required that one-third (1/3) of the training hours, a minimum of 45 hours, should be theoretical instruction. Two-thirds (2/3) of the training hours, a minimum of 90 hours, should be practical, hands-on, face-to-face laboratory instruction. It is expected that the course work would include ongoing measures of student competency such as exams after completion of independent study unit and evaluation of skills competency.

• Didactic instruction can be delivered in the classroom or by distributed education, which is defined as the teacher and the student being separated by time and/or space. Typically, distributed education involves technology such as the internet, interactive television, or videotape. Review time (independent study) and homework are not recognised as interactive instruction and will not be counted as contact hours.

• Proof of satisfactory completion of 12 credit hours of college-level human anatomy, physiology, and/or pathophysiology from an accredited college or university.

• Have current unrestricted licensure in a related medical field (PT, PTA, OT, COTA, MT, SLP, RN, MD, DO, DC, PA, ATC). These criteria are consistent with the Lymphology Association of North America (LANA) standards that have been put forth in an effort to establish basic minimum standards to certify adequate competency in the treatment of lymphoedema. Advanced education in Complete Decongestive Therapy is necessary to achieve these basic criteria. Patients and health care providers are advocating for advanced training to adequately meet the needs of this specialised population. It is the position of the NLN that therapists treating patients with lymphoedema meet the above criteria as a basic minimum standard to ensure that an appropriate level of care is being provided to this population."

Recommendations

EQ7.1 All certified lymphoedema therapists (CLT) employed by the health service should have a health profession qualification and be registered with their relevant professional body.

Evidence Grade: D

Strength of recommendation: Strong

EQ8: Is lymphoedema education necessary to be taught as part of the curriculum in undergraduate health professions?

Evidence Summary

To address the undergraduate lymphoedema educational deficit, the International [Lymphoedema Framework has identified Lymphoedema Education Benchmark Statements \(LEBS\)](#), which reflect an international consensus regarding what any person with, or at risk of lymphoedema, might reasonably expect from newly qualified health professionals. Implementation of these standards to All-Ireland undergraduate healthcare professional programmes would foster global consistency and governance in relation to lymphoedema education. Within this framework, there are broadly three levels of education required to ensure that lymphoedema is diagnosed, treated and maintained, according to best practice guidelines.

Level 1 is for all HCPs and pertains to basic awareness of lymphoedema.

Level 2 is for HCPs treating non-complex lymphoedema.

Level 3 is for certified lymphoedema clinicians.

Recommendations

EQ8.1 All relevant undergraduate health professional degrees should include a basic lymphoedema education as part of their curriculum as per the ILF Lymphoedema Education Benchmark Statements.

Evidence Grade: D

Strength of recommendation: Strong

PART B: Guideline Development

1.0 INITIATION

1.1 Purpose

The purpose of the All-Ireland Lymphoedema Guidelines 2021 is to provide an evidence-based, standardised approach for lymphoedema management in Ireland. This guideline will facilitate safe and holistic patient care, to those who access healthcare across Ireland. It will also influence the HSE/HSC to develop services according to evidence-based best practice.

1.2 Scope

While these guidelines and the general principles of lymphoedema management largely apply to all patients with lymphoedema, this document places particular emphasis on the patient groups most commonly encountered in routine clinical practice, and those that pose particular challenges to clinicians. The guideline is organised into the following sections:

- Lymphoedema definitions and background
- General lymphoedema management
- Chronic oedema management
- Primary lymphoedema management
- Management of lymphoedema in people living with obesity
- Management of oncology-related lymphoedema
- Management of palliative care-related lymphoedema
- Management of lymphoedema in children and young people
- Surgical management options for lymphoedema
- Education for people living with lymphoedema and HCPs

The needs of these special populations with lymphoedema are addressed in the relevant sections.

1.2.1 Target user

The guideline is a clinical awareness and commissioning resource for all in the HSE and HSC: clinicians, managers and commissioners, and is a guide for academic development and research in this specialist area.

1.2.2 Target population

For the purpose of this document healthcare professionals (HCPs) are defined as Health and Social Care Professionals (HSCP), Allied Health Professionals (AHPs), Social Work Staff, Nursing and Medical Staff. This document is aimed at healthcare staff and social care professionals involved in the care of people, (adults and children) living with lymphoedema. This population is multi-professional in accordance with the causative factors and sequelae of the wider condition and includes those involved in the physical and mental health management of people living with lymphoedema. Each member of the multidisciplinary team is clinically and professionally accountable for implementing the recommendations relevant to their discipline.

The relevant CEO, Clinical Director and the Director of HCPs/Chief Allied Health Profession Officer (CAHPO) and health service providers have corporate responsibility for the implementation of the recommendations in this guideline. This guideline is relevant for academic and research institutions for future education and research, as highlighted in the guideline recommendations.

This document is also relevant for service user support and to those who provide specific resource support e.g. Lymphoedema Ireland and the Lymphoedema Support Network.

1.3 Aims and Objectives

1.3.1 Aims

To provide current evidenced-based recommendations for lymphoedema diagnosis, assessment and management for both adults and children.

1.3.2 Objectives

To promote a standardised approach to lymphoedema management across all care settings in Ireland.

1.4 Outcomes

It is anticipated that this guideline will enhance or improve patient outcomes, education and research opportunities.

1.5 Guideline Development Group

1.5.1 The Guideline Development Group (GDG)

The GDG, directed by two project leads, undertook a comprehensive review of the existing literature and regulation to inform this guideline. The cross-border group collaborated extensively on this resource, which was then circulated nationally and internationally for external consultation and peer-review.

1.5.2 Membership of the Guideline Development Group

The guideline development group and the work-stream groups (WSGs) comprised of professional clinical experts representing various lymphoedema pathways and health disciplines from both Northern Ireland (N.I.) and the Republic of Ireland (RoI). The project leads worked with all resources to undertake and implement the project. The WSGs were responsible for providing expert advice, support and assistance to the project leads. All project management plans were reviewed and approved by the project team.

Refer to [appendix VI](#) for details of the membership of the GDG.

1.5.3 Conflict of Interest

As indicated by the completed 'conflict of interest forms' (appendix VII) no conflicts of interest were noted.

1.5.4 Funding Body and Statement of Influence

The guideline was commissioned and funded by the HSE and the HSC (via the Lymphoedema Network Northern Ireland (LNNI)). Formatting was funded by the HSE and printing was provided by the Office of the Revenue Commissioners Ireland. This process was fully independent of lobbying influence. All recommendations were based on the best research evidence integrated with clinical expertise.

1.6 Governance Group

The Office of the Director of Primary Care Strategy and Planning and the Public Health Agency (via the LNNI Board) commissioned this project, and had the authority and responsibility for managing and executing the project according to the project plan. The Project Leads (who report to The Office of the Director of Primary Care Strategy and Planning and the LNNI Board (chaired by Assistant Director of AHPs and PPI, Public Health Agency)), managed, coordinated and administered the process. The GDG is grateful to the health service organisations and members of Higher Education Institutions (HEIs) whose practice-development staff and clinical personnel gave their time and expertise to this project. The GDG views the implementation of this guideline, at all levels of responsibility, as fundamental to the success of patient-centred care being delivered by the organisation.

1.6.1 Membership of the Approval Governance Group

Refer to [appendix VIII](#) for Membership of the Approval Governance Group.

1.7 Supporting Evidence

References can be found in Section 8.0. Other supporting evidence is located within the appendices

1.7.1 Legislation and other related Policies

- Health Service Executive (2011) Standards and Recommended Practices for Healthcare Records Management
- Health Service Executive (2016) National Framework for developing Policies, Procedures, Protocols and Guidelines (PPPGs)
- Health Service Executive (2017) Integrated Risk Management Policy
- National Guideline for Oedema Compression Garments for the Prevention and Management of Chronic Oedema/Lymphoedema 2022

These were the current versions of these documents at the time of publication of this guideline.

1.7.2 Guidelines being replaced by this guideline

The CREST (2008) Guidelines for the diagnosis, assessment and management of lymphoedema.

1.7.3 Related PPPGs

Currently there are no other PPPGs related to this guideline.

1.8 Glossary

Refer to [appendix XIII](#) for a full glossary.

2.0 GUIDELINE DEVELOPMENT

2.1 The Clinical Questions

The clinical questions informing the revision of the 2008 CREST guideline, determined the need for a robust literature search, to identify the most current evidence underpinning the areas of lymphoedema diagnosis and management discussed in this guideline.

2.2 Literature Search Strategy

A comprehensive literature review was undertaken which included national and international publications. Specific research questions were formulated by the GDG and WSGs using PICO methodology, and a literature search was undertaken to answer the questions posed. All results were reviewed by the work streams and helped in the generation of recommendations presented in this document. All searches and screening were conducted independently by each work stream, each with at least 4 reviewers which increased our confidence that all relevant and current evidence was identified for the review. A second search was completed in Spring 2021 to capture subsequent additions to the literature.

Refer to [appendix X](#) for full search strategy including databases and online search resources used for.

2.3 Evidence Appraisal

2.3.1 Data Extraction

The following data was extracted using a bespoke data extraction tool: author, title, source; date of study, country of origin; care setting; inclusion and exclusion criteria; baseline participant characteristics; study design details; specific initiative under investigation (with definitions); duration of follow-up; loss to follow-up and outcomes data.

2.3.2 Data Analysis

The literature review was performed according to international standards by following the Cochrane Guidelines (<http://handbook.cochrane.org/>) and the following PRISMA Guidelines (<http://www.prisma-statement.org/>). This review followed the Cochrane guidelines and specifications set out as a requirement of a thorough, objective and reproducible search of a range of sources to identify as many relevant studies as possible. Transparent and complete reporting of the literature review followed the PRISMA guidelines (Moher et al., 2009) and AGREE II instrument (Brouwers et al., 2010) for reporting on clinical guidelines.

2.3.3 Quality Appraisal

Each included study was quality-appraised using the evidence-based literature critical appraisal checklist devised by Glynn (2006). This checklist appraised each study under the following domains:

- Population
- Data collection
- Study design
- Results

The critical appraisal checklist has a number of subcategories, and each is assessed with a generic list of closed questions, resulting in a yes (Y), no (N), unclear (U), or a not-applicable (N/A) answer. Calculation for each section's validity according to the checklist is as follows: $T = Y + N + U$, if $Y/T < 75\%$, or if $(N + U)/T > 25\%$, then you may safely conclude that the section identifies significant omissions and has questionable validity. The Glynn calculation for overall study validity was performed for each study assessed in this guideline and is as follows: $T = Y + N + U$, if $Y/T \geq 75\%$ or if $(N + U)/T \leq 25\%$ then it can be concluded that the study is valid. The critical appraisal tool provided a thorough, generic list of questions that one would normally ask when attempting to determine the validity, applicability and appropriateness of a study, either qualitative or quantitative, since the tool allows for the use of non-applicable for questions which are not relevant to the particular study under examination.

2.4 Grading of recommendations

The recommendations in this guideline originate either directly from existing guidelines or were formulated by members of the GDG, based on evidence gathered in response to PICO questions proposed. As per ADAPTE (2009) guidance for documents of this nature, an original grading scheme was developed and used to grade all recommendations.

Recommendations not originating from existing guidelines were formulated by the GDG, based on evidence derived from PICO searches. The process used for grading the evidence throughout this guideline is as follows:

Level of Evidence	Source of the Evidence
A	Data derived from multiple randomised clinical trials or meta-analysis
B	Data derived from a single randomised clinical trials or large non-randomised studies
C	Recommendation comes directly from an existing guideline
D	Consensus of expert opinion and/or small studies, retrospective studies, registries

This grading system was devised by members of the GDG, and recommendations graded using this system were denoted as "HSE Recommendation Evidence Grade: A, B, C or D".

Strength of recommendations (Adapted from GRADE working group 2013); recommendations are graded either "Strong" or "Weak". The strength of recommendation reflects the balance of the following items:

- The quality of the body of evidence
- The balance between benefit and harm to patient
- Patient preferences and values
- Resources/cost

2.5 Summary of the Evidence

Using a systematic approach to searching, screening and appraisal, this review has identified a number of evidence-based recommendations for lymphoedema management, adapted to reflect care in the All-Ireland healthcare setting. The findings of this review should be viewed alongside the following limitation: inclusion of exclusively English-language studies potentially limits the scope of our search and we cannot exclude the possibility that we have missed some significant portion of scientific literature. This limitation is somewhat offset however, by the use of explicit inclusion criteria, PICO, and a broad search strategy including guideline databases.

2.6 Resources

A budget impact analysis was not undertaken however the resources required to implement the guideline recommendations have been considered in the HSE Lymphoedema Model of Care and the LNNI Health Needs Assessment. These recommendations update current practice and include changes that will result in an increase in resource consumption, which needs to be analysed further upon publication of this guideline.

3.0 GOVERNANCE AND APPROVAL

3.1 Governance

The Office of the Director of Primary Care Strategy and Planning (RoI) and Assistant Director of AHPs and PPI, Public Health Agency (PHA) (N.I.) commissioned this project. The Project Leads (who report to the Director of the Primary Care Strategy and Planning and PHA Assistant Director) coordinated and administered the process.

A multidisciplinary project team undertook the guideline development process and the GDG was chaired by the Project Leads. Membership of the GRG included service users and clinicians from across disciplines representing a range of clinical settings and from Higher Education Institutes, and from across the island of Ireland. Consultation with chairs of each National Clinical Care Programme and other national stakeholders was undertaken. Details of the governance arrangements, the GRG membership and each of the guideline Work Stream Group members are available in Membership of the PPPG Development Group .

When necessary, wider consultation was undertaken with topic-specific experts to ensure that all available evidence was included. Final approval was sought and issued from the sponsors of the project. Governance of the guideline development process was provided by a multidisciplinary project team, and the Guideline Development Group (GDG) was chaired by the Project Leads.

3.2 Method for assessing the guideline as per the HSE national framework for developing PPPGs and the HSC GAIN / Regulation and Quality Improvement Authority

The Policies, Procedures, Protocols and Guidelines Checklist was reviewed in conjunction with the final revised guideline to ensure compliance with the standards outlined in the “HSE National Framework for developing Policies, Procedures, Protocols and Guidelines (PPPGs) 2016”. National and international expert peer review of the guideline was completed. This feedback was used to finalise the document.

3.2.1 National Stakeholder and International Expert Review

National and international expert peer review of the guideline was completed in March/April 2021. Reviewers were requested to comment on the presentation, process of development, robustness of the search, comprehensiveness of the evidence used, content of the recommendations and implementation. Feedback was submitted with supporting evidence on a form provided. All feedback received was reviewed by GDG and incorporated, as appropriate, into the final document. A log was maintained of all submissions and amendments from the national and international expert review process.

3.3 Copyright/Permission Sought

Copyright and permissions were sought from the organisations or authors of texts included in this guideline, where necessary. Refer to [appendix XII](#) for a full list of copyright/permissions sought.

3.4 Approval and Sign-Off

The completed Lymphoedema Management Guideline 2022 was submitted for approval to the HSE Chief Clinical Officer Forum and the Public Health Agency (PHA) Northern Ireland. This was accompanied by the signed PPPG Checklist (refer to [appendix IX](#) Approved Policies, Procedures, Protocols and Guidelines Checklist) to confirm that all the required stages in the revision of the guideline had been completed and met the “HSE National Framework for developing Policies, Procedures, Protocols and Guidelines (PPPGs) 2016”. The guideline was approved in April 2022.

4.0 COMMUNICATION AND DISSEMINATION

4.1 Communication and Dissemination Plan

It is important that the guideline is disseminated as soon as it has been completed. This approach ensures that it can be implemented immediately to support clinicians. The Communication and Dissemination Plan will be implemented to achieve maximum circulation to inform all stakeholders that this guideline supersedes all previous lymphoedema management guidelines. The following activities will be undertaken by the HSE/HSC to ensure all relevant stakeholders are informed of the updated guidelines:

- Utilise the master list of all relevant stakeholders
- All relevant stakeholders to receive a copy of the guideline (insofar as is possible)
- Use of communication links including healthcare organisations, professional bodies, associated charitable bodies and educational groups
- The identification of local champions to promote the new guideline
- Uploading the policy to relevant webpages
- Dissemination via lymphoedema groups and special interest organisations

5.0 IMPLEMENTATION

5.1 Implementation of the All-Ireland Lymphoedema clinical Guideline 2022
Implementation of the guideline will follow communication and dissemination.

5.1.1 Barriers and facilitators to implementation

It is recommended that each local clinical setting to which this guidelines applies should determine what resources are necessary for its implementation. In the Republic of Ireland, the HSE model of care for lymphoedema services should be referred to for workforce planning etc. In N.I., the 2019 LNNI Health Needs Assessment provides detail regarding current service provision and development requirements. The implementation of the guideline can be facilitated by ensuring that all clinicians understand and appreciate the degree to which this guideline contributes to the quality and safety of patient care.

5.2 Education

It is recommended that each local clinical setting will identify the educational needs that are necessary to implement this guideline in practice. Lymphoedema specialists must complete a recognised lymphoedema certification course prior working in this field. The level of education requirement for all team members will vary from in-service, continuing professional development to stand-alone modules and postgraduate education programmes.

5.3 Responsibility for Implementation

All stakeholders involved in lymphoedema management have a responsibility for the implementation of this guideline.

5.3.1 Organisational Responsibility

The corporate responsibility for the implementation of this guideline in each local health service provider lies with the CEO, Clinical Director and the Director of Nursing and/or Midwifery and/or Allied Health Profession Lead. Each member of the multidisciplinary team is responsible for the implementation of the guideline recommendations relevant to their discipline.

5.4 Roles and Responsibilities

Senior managers:

- Assign personnel with responsibility, accountability, and autonomy to implement the guideline
- Ensure local policies and procedures are in place to support its implementation
- Facilitate education to all relevant clinical staff to ensure they have the knowledge and skills to implement the guideline
- Monitor the implementation of this guideline
- Ensure audit processes are in place

Heads of department:

- Ensure all relevant staff members are aware of this guideline
- Ensure staff are supported to undertake education programmes and related training as appropriate

All clinical staff:

All clinical staff should comply with this Guideline and related policies, procedures and protocols. Clinical staff should adhere to their professional scope of practice, guidelines, and maintain competency. In using this guideline, clinicians must be aware of the role of appropriate delegation. Refer to appendix V for a copy the signature sheet. This should be signed to record that all clinicians have read, understood and agree to adhere to this guideline.

6.0 MONITORING, AUDIT AND EVALUATION

6.1 Governance

It is anticipated that these guidelines will promote and enhance evidence based practice in lymphoedema management in Ireland. This guideline positively impacts on patient care, it is important that it is audited to support continuous quality improvement in relation to its implementation. The audit process should be undertaken from a multidisciplinary perspective.

6.1.1 Monitoring

The CEO, Clinical Director and Director of Nursing and/or Midwifery and/or Allied Health Profession Lead in each local health service provider have corporate responsibility for monitoring the implementation of this guideline.

The MDT should monitor the implementation recommendations specific to their practice.

All clinicians with responsibility for the care of patients who are at risk of or who have developed lymphoedema should:

- Adhere to their professional code of conduct and scope of practice.
- Utilise this guideline and any related procedures or protocols.
- Maintain their competency for the management and treatment of patients with lymphoedema.

6.1.2 Audit

Audit practice, using key performance indicators, should be undertaken to provide evidence to support continuous quality improvement.

6.1.3 Evaluation

Evaluation of the effectiveness and associated costs of the guideline should be undertaken locally to support its implementation and sustainability.

7.0 REVISION/UPDATE

7.1 Procedure for Revising the Guideline

The All Ireland Lymphoedema Guideline 2022 will be reviewed on a 5-yearly basis and updated to incorporate any relevant new evidence.

7.2 New evidence

As new evidence emerges that requires change in practice, a surveillance of the literature will be undertaken so that the guideline will maintain its relevance and currency.

7.3 Version control

The original CREST Guidelines for the diagnosis, assessment and management of lymphoedema were issued in 2008. Therefore the revised “All Ireland Lymphoedema Guideline” is the second version and will be due for revision in June 2027. The guideline will be available on the [HSE](#) and [LNNI](#) websites, and will be shared with national and international stakeholders.

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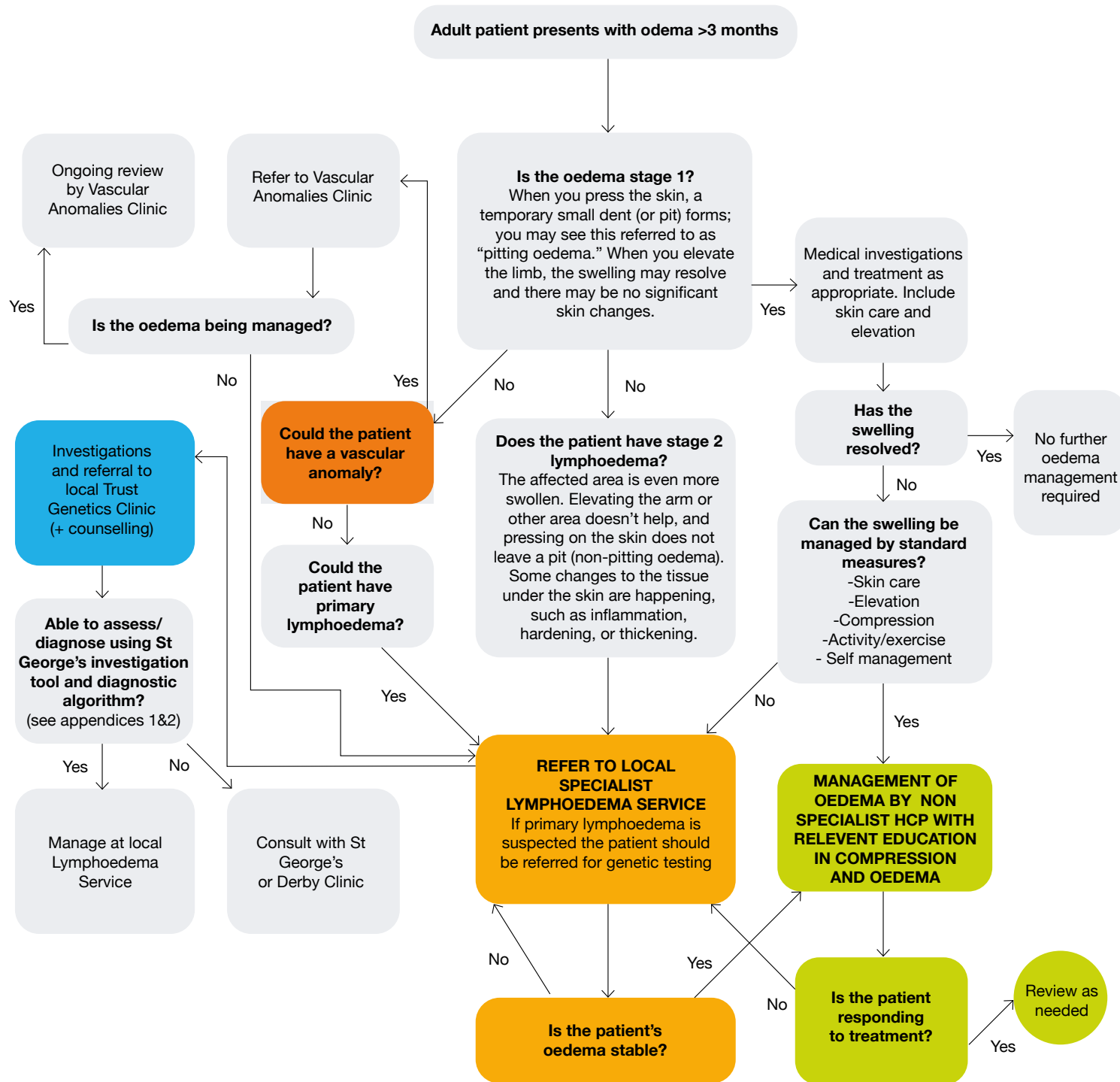
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9.0 APPENDICES

Appendix I. Lymphoedema Care Pathways

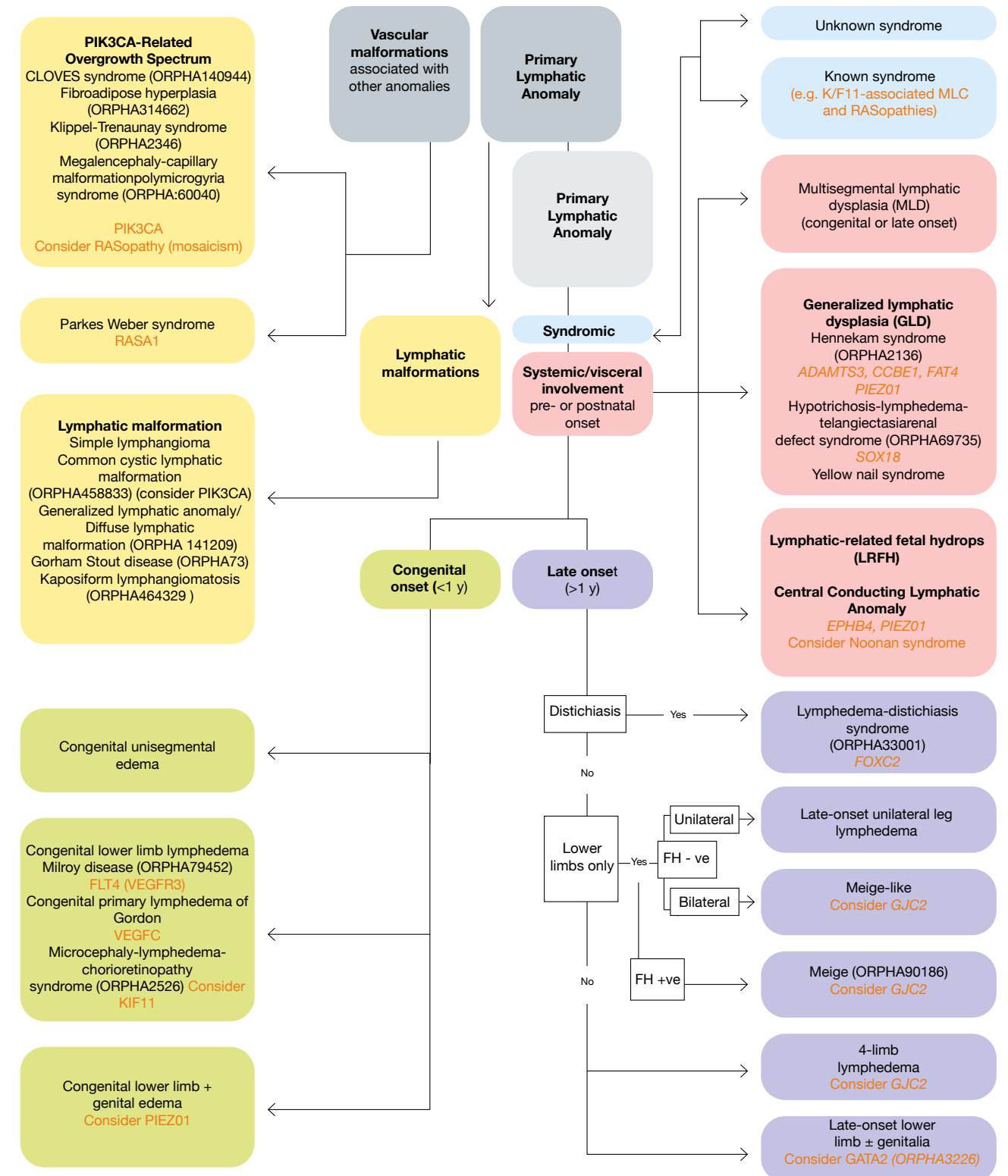
I.I Adult Lymphoedema Care Pathway for Stage 1 Lymphoedema and above



Liaise with relevant Health Care Professionals:
podiatry, footwear clinic, occupational therapy, dietetics, physiotherapy, psychosocial etc.



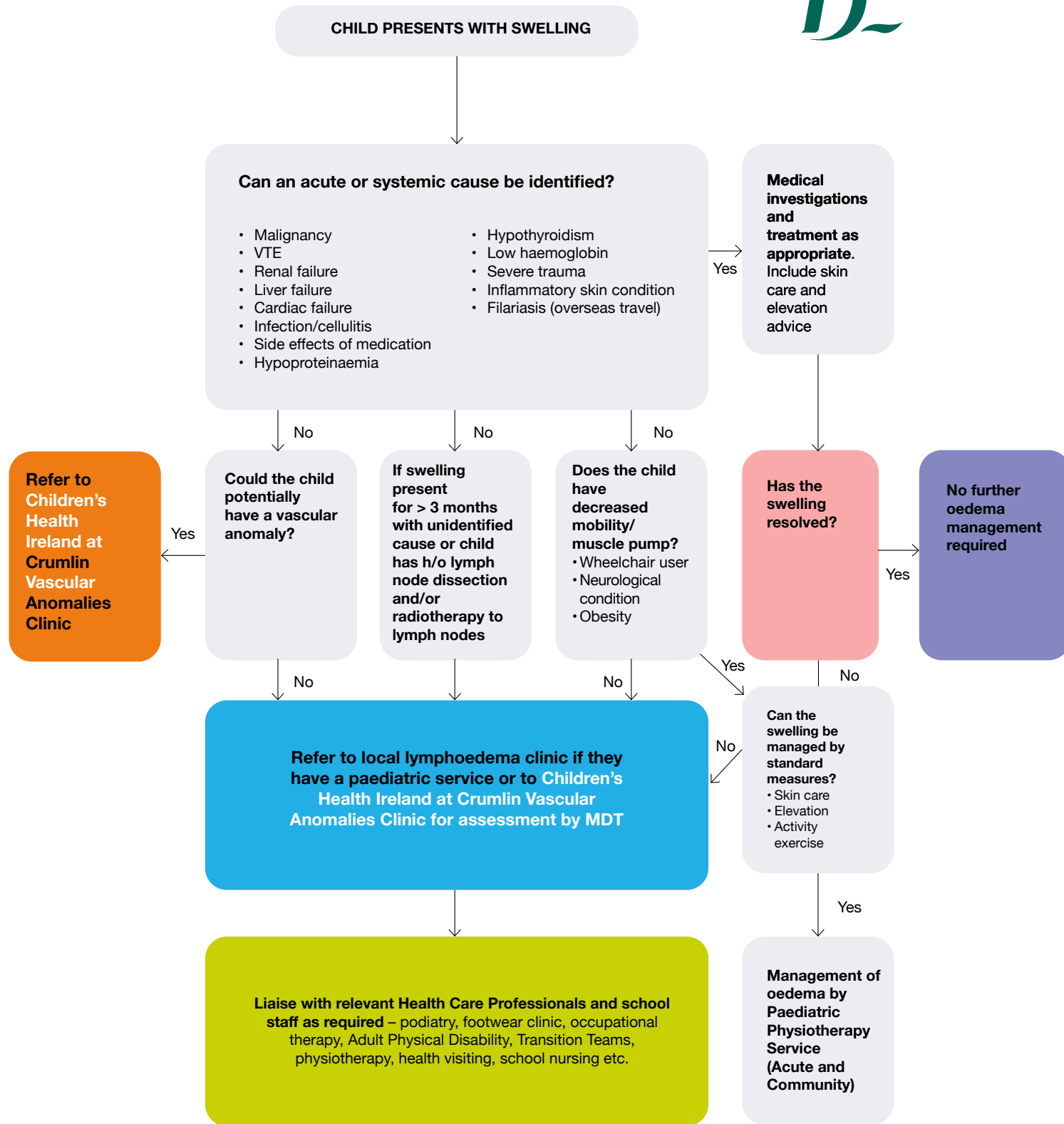
I.II Primary Lymphoedema Care Pathway: St. George's Classification Algorithm for Primary Lymphatic Anomalies (Gordon et al., 2021)



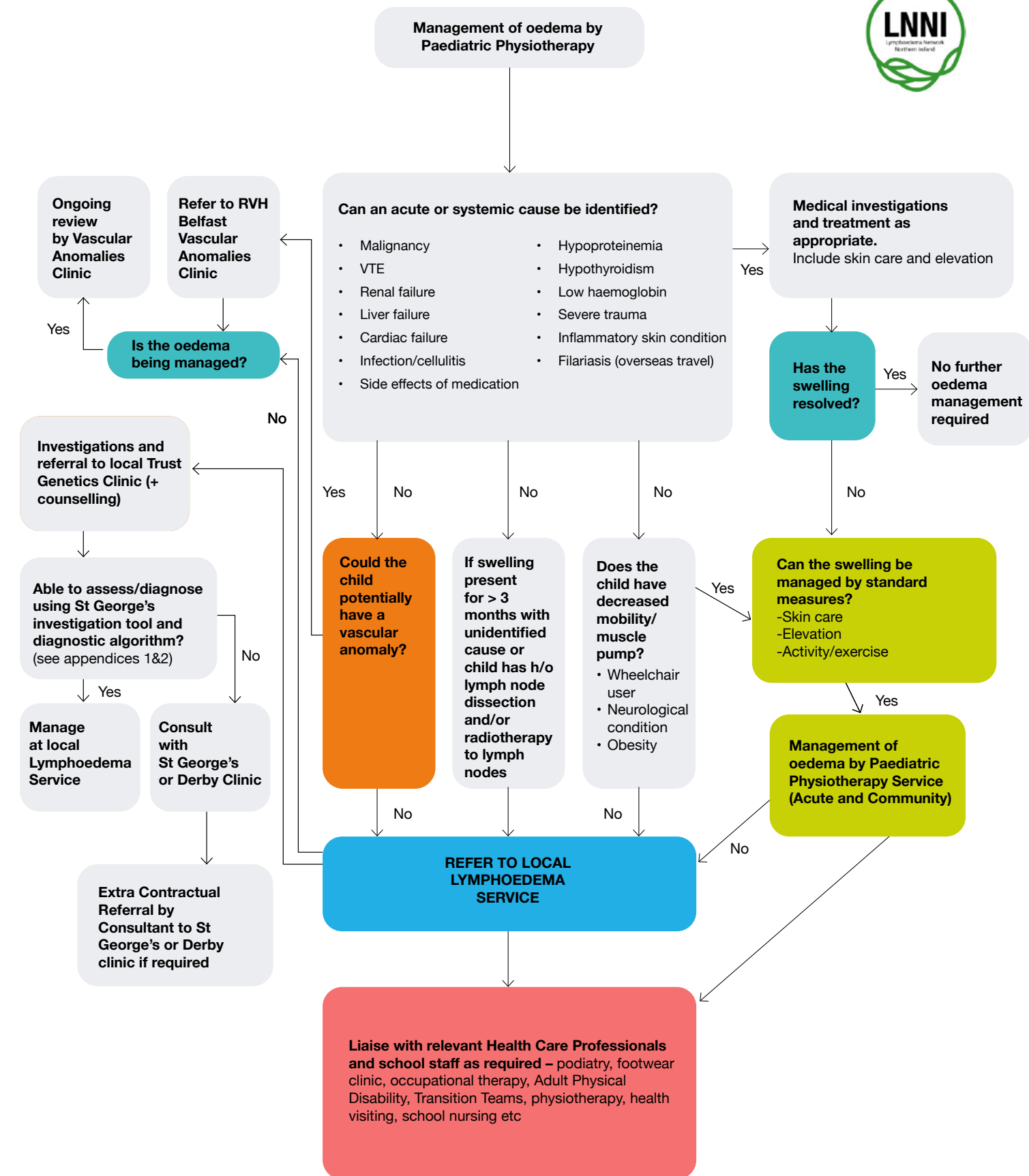
Note. Reprinted from "The St. George's Classification Algorithm of Primary Lymphatic Anomalies," by K. Gordon, 2021, *Lymphat Res Biol*, 19 (1), 25-30. Copyright (2022) by, Mary Ann Liebert Inc. publishers.

I.III Children And Young People Pathways

I.III.I HSE Pathway for Children and Young People with Lymphoedema

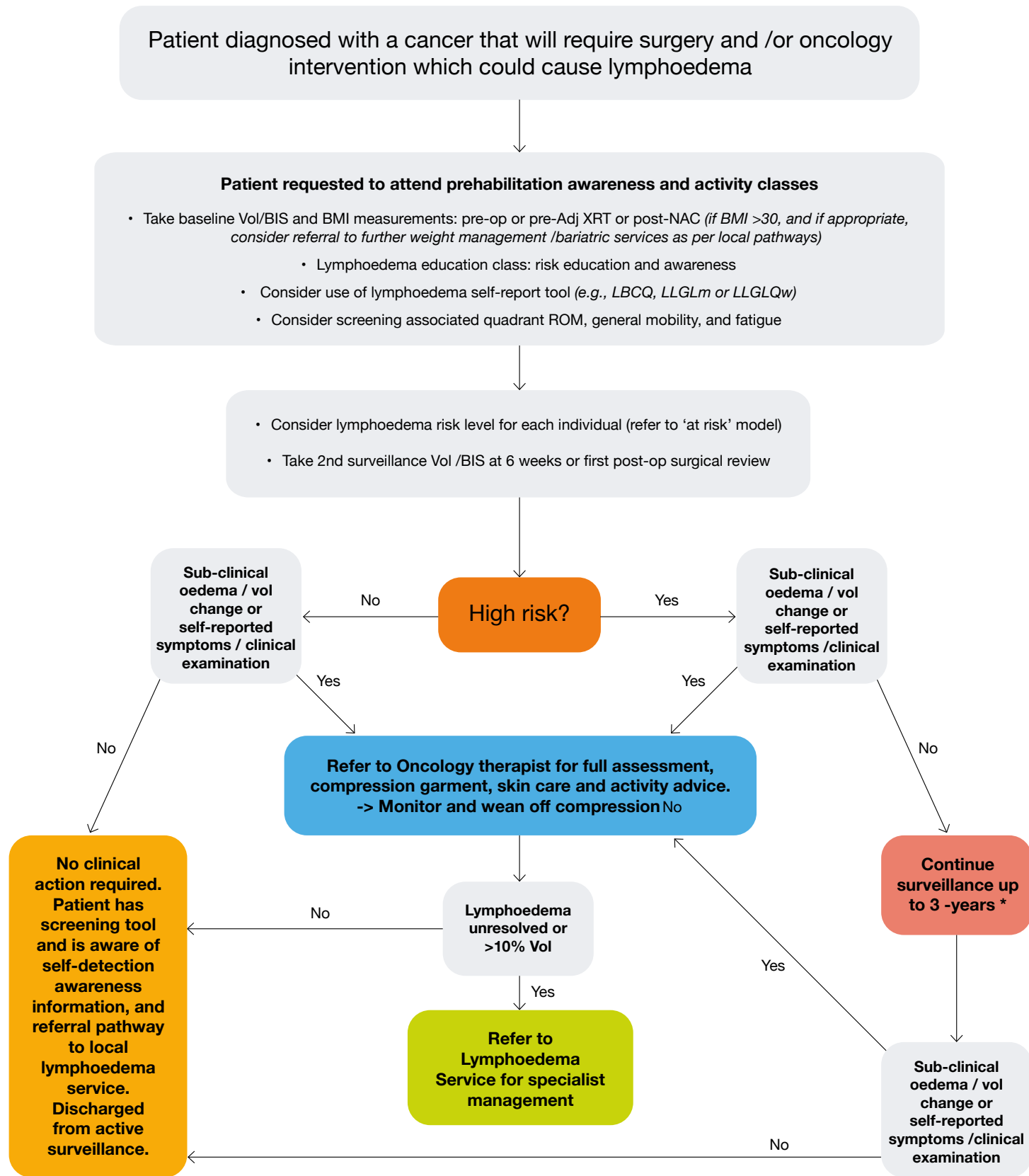


I.III.II LNNI Pathway for Children and Young People with Lymphoedema



I.IV Oncology Screening and Surveillance and Management Pathways

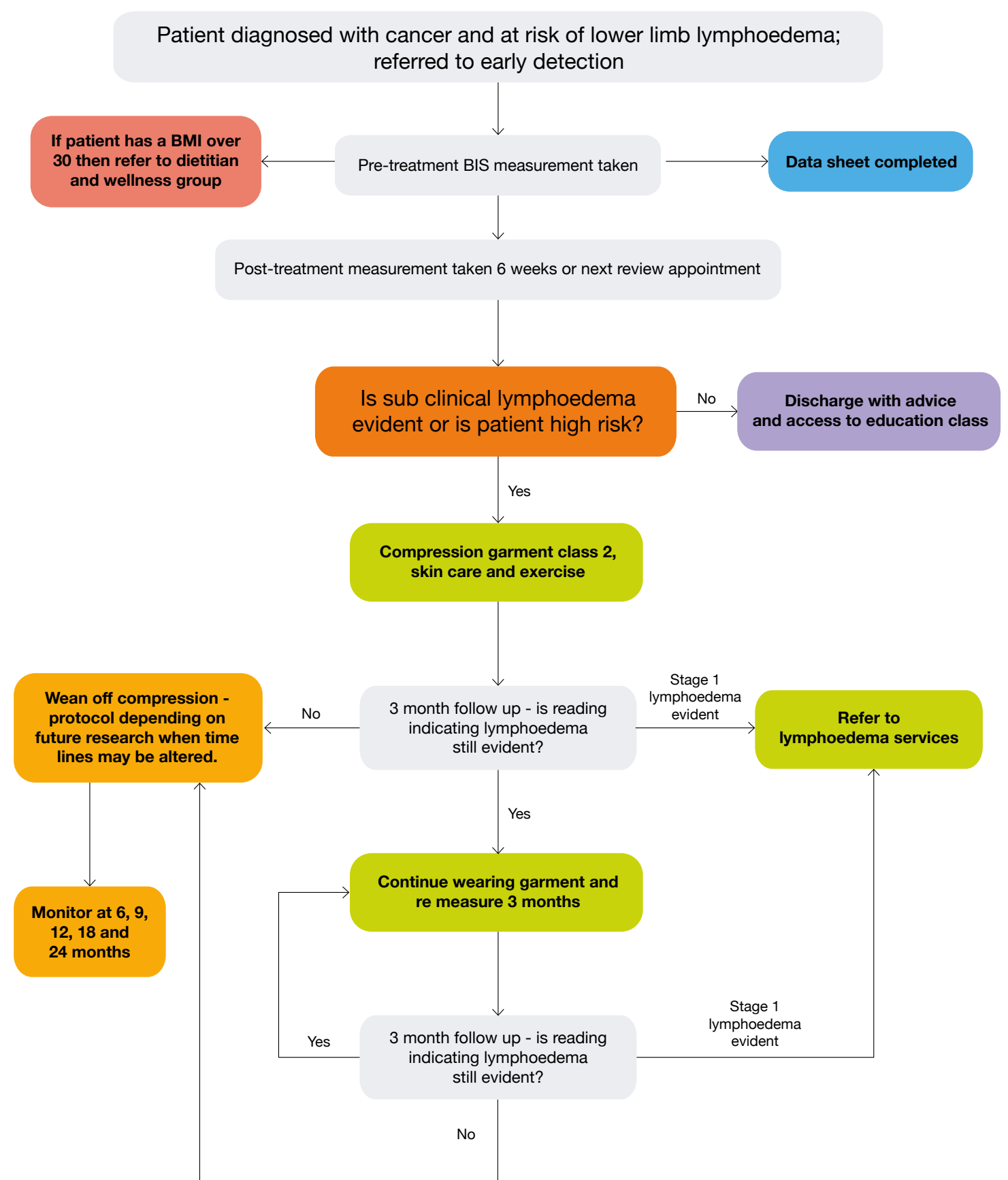
I.IV.I Guideline Screening Model

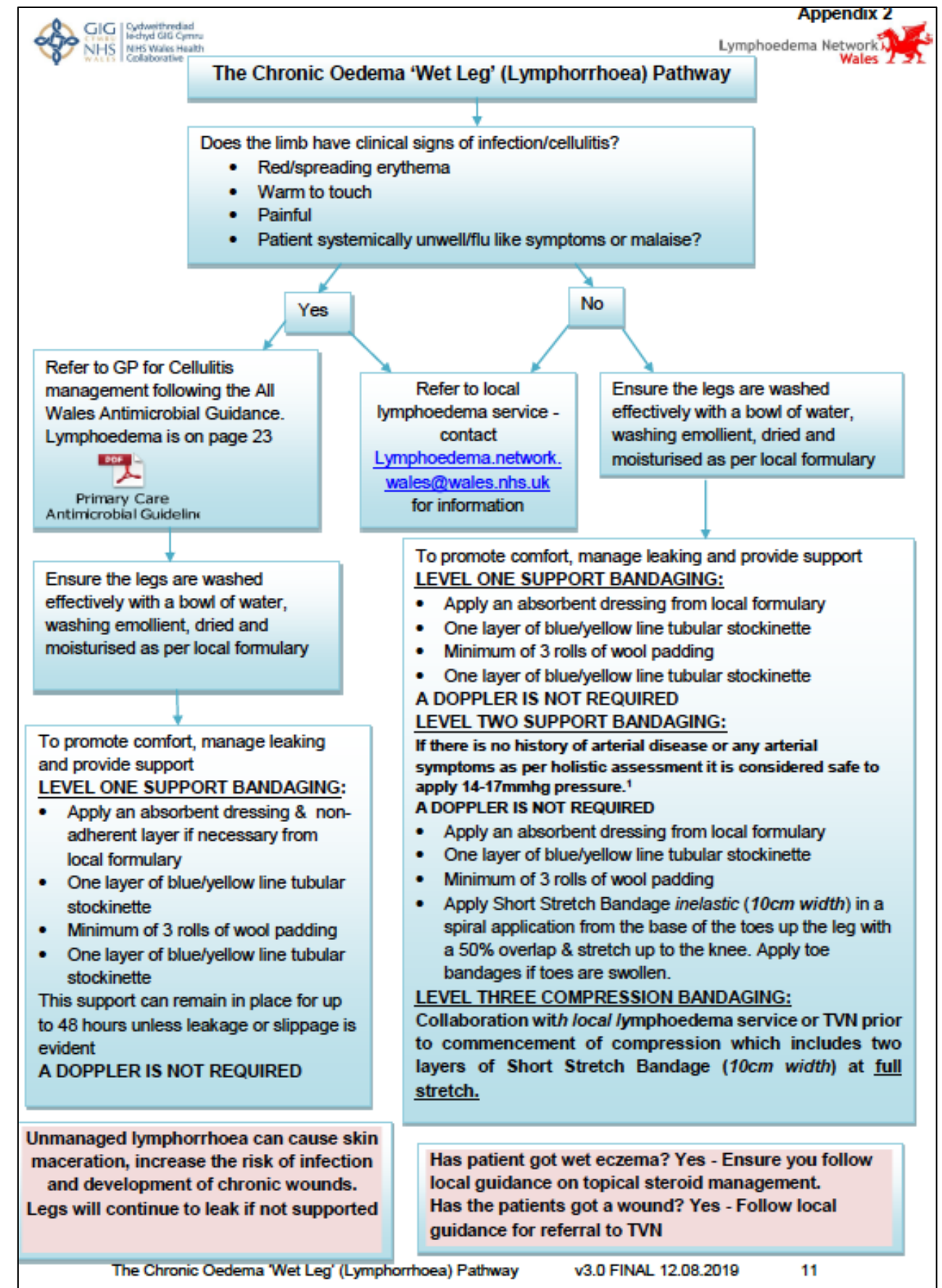
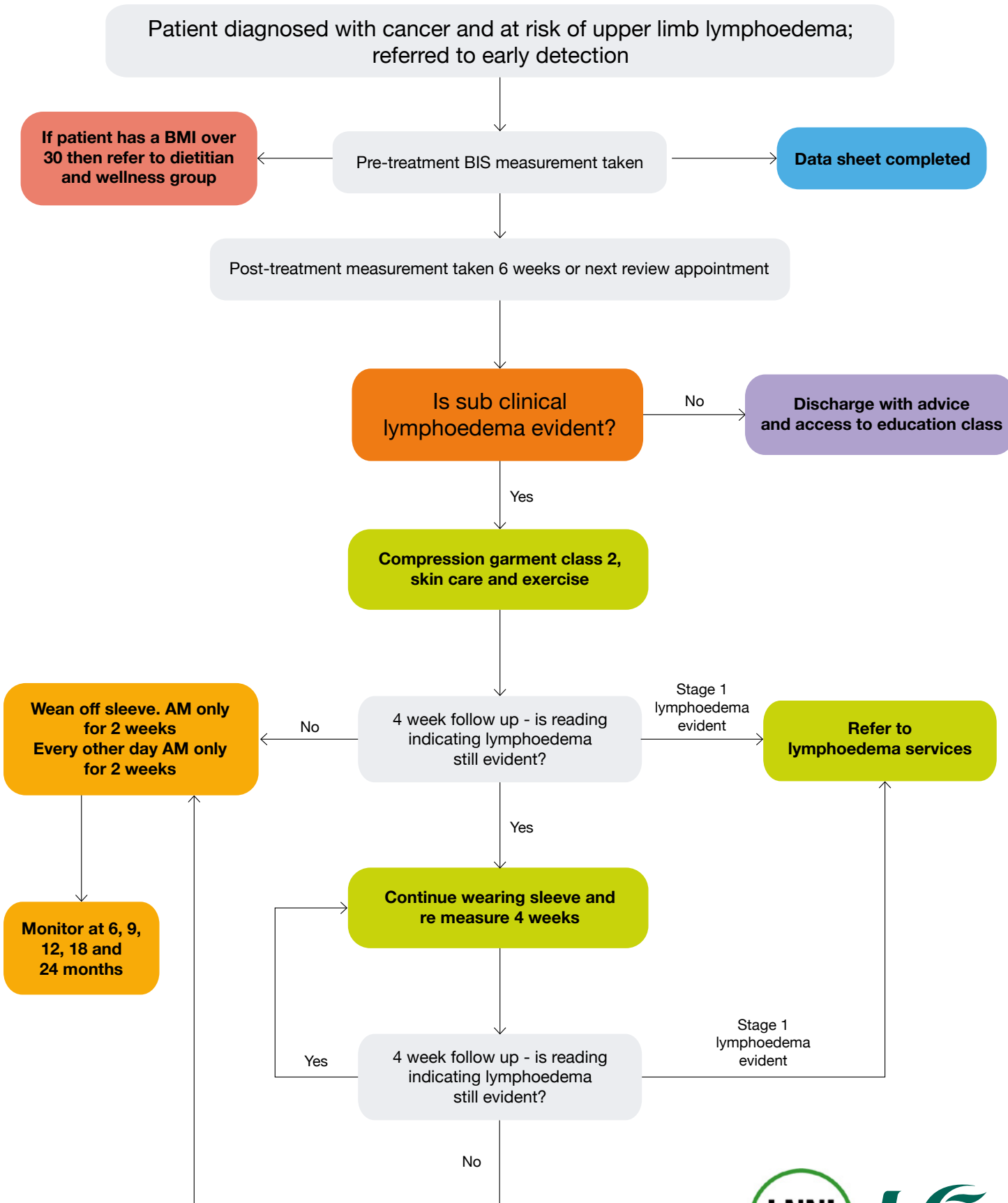


*Patients at high risk of cancer related lymphoedema should be on the screening and surveillance pathway and should be monitored at baseline (pre-op), surgical review (6-8 weeks post op), 9 months, and thereafter at 1, 1.5, 2, 2.5 and 3 years. Relative volume change, TDC or BIS should be undertaken at these points to assess for lymphoedema.

LBCQ -Lymphoedema Breast Cancer Questionnaire; LLGLQm - Lower Limb and Genital Lymphoedema Questionnaire - for men; LLGLQw - Lower Limb and Genital Lymphoedema Questionnaire - for woman

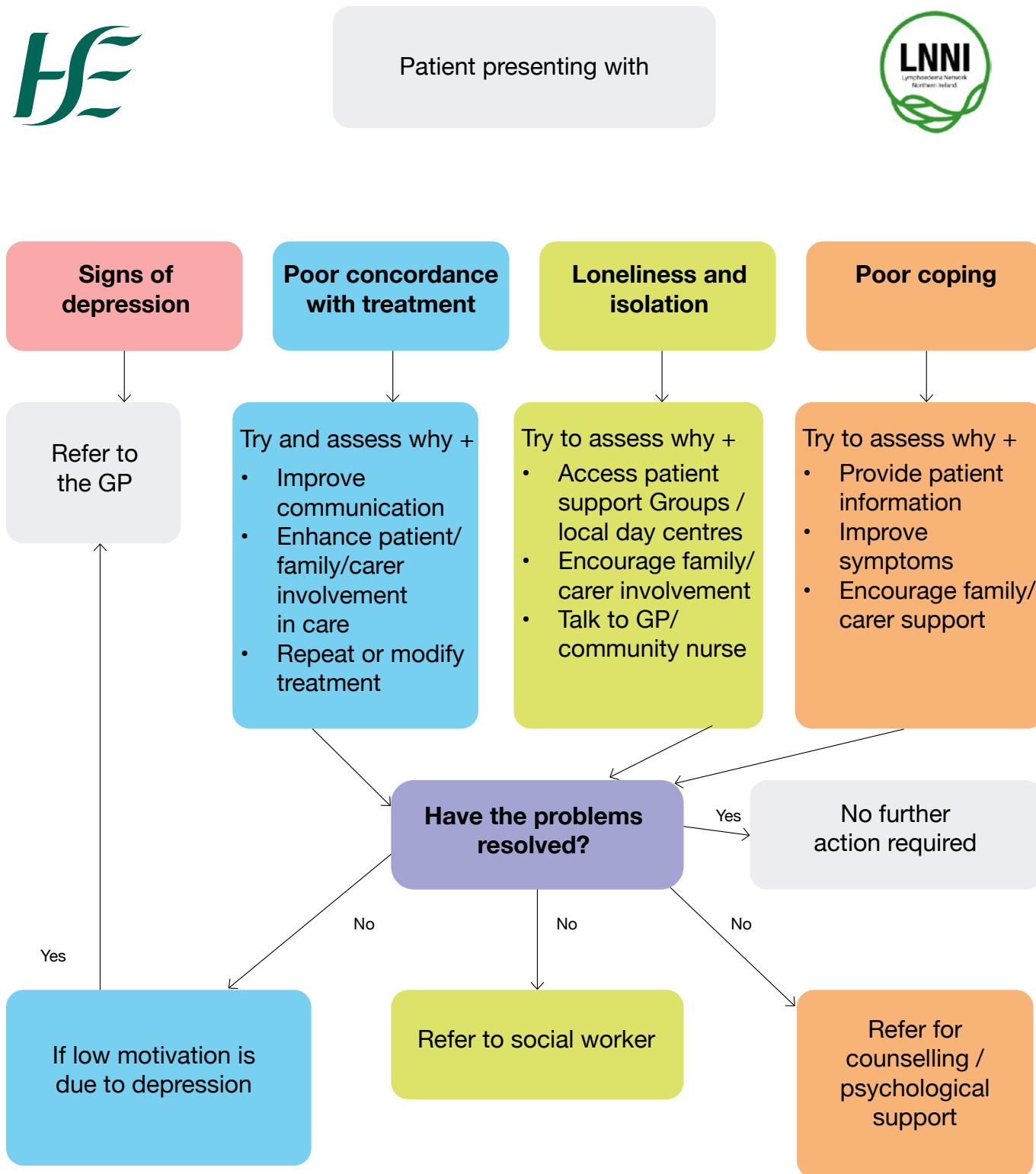
I.IV.II All- Ireland Subclinical Lymphoedema Pathway – Lower Limb





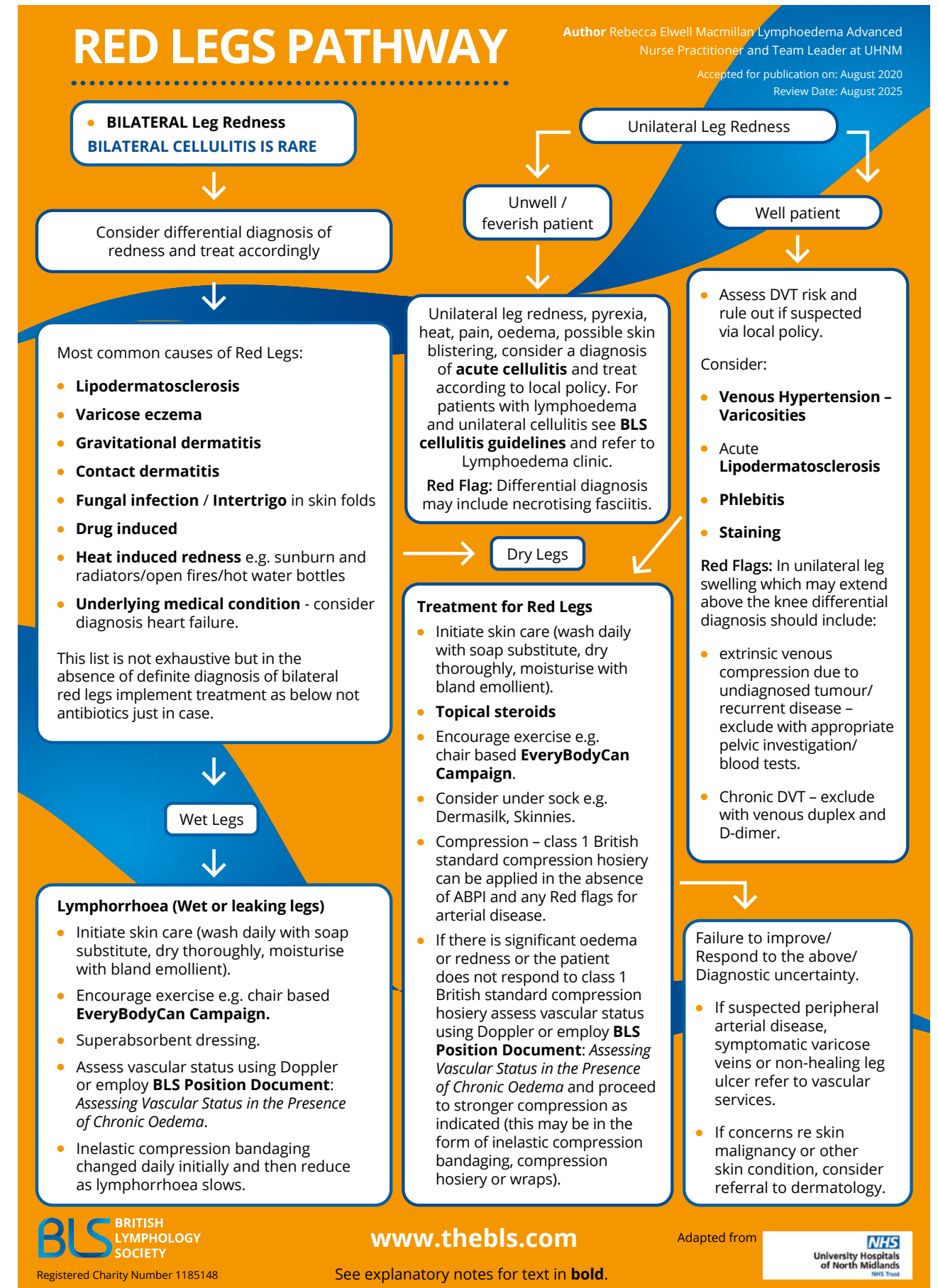
Note. Reprinted from "The Chronic Oedema 'Wet Leg' Lymphorrhoea pathway," by Lymphoedema Network Wales, 2019, Copyright (2022) by, NHS Wales.

I.VI All-Ireland Psychological Support Pathway for patients with lymphoedema



Note. Adapted from the International Lymphoedema Framework 2006 International consensus: Best practice for the management of lymphoedema. Internal Lymphoedema Framework, 2006.

I.VII BLS pathway for patients presenting with red legs



Note. Reprinted with thanks and permission from the British Lymphology Society, 2020.

5 important things you can do to help your Lymphoedema



1

Wear your sleeve, stocking or wrap every day.

- ✓ Put on first thing in the morning and take off at bedtime. Do this unless your therapist has said not to.
- ✓ Check that your new stocking looks the same as your old stocking.



2

Take good care of your skin.

- ✓ Wash and dry your skin every day.
- ✓ Rub skin cream onto your skin every night. This will help your skin and help stop infection.
- ✓ Use the skin cream your therapist tells you to.



3

Be active!

- ✓ Do not sit or stand in one position for too long.
- ✓ Help move fluid in your body.
 - Paddle your feet up and down.
 - Get up and walk around the room.
 - Go up and down one step a few times.



4

Think about your weight

- ✓ Try to stay a healthy weight.
- ✓ Try to lose weight if you are heavy.
- ✓ Eat healthy food.
- ✓ Exercise often.



5

Keep your arm or leg lifted

- ✓ Lift your arm or leg up for a short amount of time. This will help with swelling.
- ✓ Sleep in a bed.
- ✓ Do not sleep in a chair or recliner.



If you are worried about your lymphoedema or you have any questions contact your lymphoedema therapist as soon as possible.

II.1 Circumferential Limb Volume and Outcome Measurement for Lymphoedema

Date and Time:									
Circumference Metatarsal Phalangeal (MTP) or Metacarpal Phalangeal (MCP)		Right	Left	Right	Left	Right	Left	Right	Left
Mid foot (.....cm from nail bed of middle toe, ankle 0°dorsi flexion (DF)) or webspace									
1 =cm from sole of foot (ankle 0° DF) or 1 =cm from nail bed of middle finger, wrist in 0° flexion (flex)	1								
	2								
	3								
	4								
	5								
	6								
	7								
	8								
	9								
	10								
		mls	%	mls	%	mls	%	mls	%
Affected Side: Right Left Bilateral Dominant Side: Right Left Heightm	1								
	2								
	3								
	4								
	5								
	6								
	7								
	8								
	9								
	10								
Proximal Volume (mls)									
Proximal Volume Difference		mls	%	mls	%	mls	%	mls	%
Total Volume (mls)									
Total Volume Difference		mls	%	mls	%	mls	%	mls	%
Episodes of cellulitis since last review									
Weight (kg)	BMI								
Health Today Score (VAS 1-10)									
Lymphoedema Life Impact Score (LLIS)									
HbA1c (pre-diabetes 43-47 mmol/mol)									
Date and Time:									

II.II Adult Lymphoedema Assessment and Review Template

History of Oedema	Consent for assessment / ECR <input type="checkbox"/>
Location of lymphoedema: Include: progression, cellulitis, previous management of oedema/cellulitis, aggravating/relieving factors)	Date of lymphoedema onset:
Patient's Perception:	
Family History of Oedema Yes/No _____	
Ethnicity:	

Current Symptoms (✓ = present; x = absent)

	Functional restriction		Skin changes (tight / shiny)
	Heaviness		Swelling (difficulty with clothing / rings)
	Reduced ROM (objective table - see page 3)		Tingling, pins and needles, paraesthesia
	Pain (related to lymphoedema) Site: _____		
	Description: _____		
	0 (no pain)		10 (worst pain imaginable)

Cancer-Related Lymphoedema

Cancer diagnosis:		
Date(s) of surgery:		
Regional lymph node clearance <input type="checkbox"/> (Level 1/2/3)	Sentinel node biopsy <input type="checkbox"/>	Nodes +ve /removed (e.g. 2/20):
Post-operative complications:	Seroma	Details:
	Cording	
	Infection	
	Delayed Wound Healing	
Hormonal Therapy (regime, date started)		
Chemotherapy (regime, no. of cycles, date completed)		
Radiotherapy (site, date completed, length of treatment)		

Non-Cancer Related Surgery

Type of Surgery	Date	Details
CABG		
Orthopaedic		
Plastic		
Varicose vein		
Other		

Past Medical History (✓ = present; add additional conditions)

Allergies	e.g. penicillin, latex, elastoplast		
Diabetes			
Hypertension			
Sleep apnoea			
Further Details of PMH:			
General Precautions (Contraindications)			
Heart failure	<input type="checkbox"/> uncontrolled	<input type="checkbox"/> controlled	
Deep vein thrombosis	<input type="checkbox"/> acute	<input type="checkbox"/> chronic	
Phlebitis/cellulitis	<input type="checkbox"/> acute	<input type="checkbox"/> history of	No. of episodes in past year: Prophylaxis: Y/N Hospital admissions
Renal failure Stage 3+	<input type="checkbox"/> acute	<input type="checkbox"/> chronic	Stage:
Neck MLD Precautions (Contraindications)			
Thyroid	<input type="checkbox"/> hypo	<input type="checkbox"/> hyper	
Cardiac arrhythmia			
Hypersensitivity of carotid sinus			
Deep abdominal MLD Precautions (Contraindications)			
Abdominal aortic aneurysm			
Abdominal pain (unexplained)			
Abdominal radiotherapy			
Abdominal surgery (recent)			
Diverticulitis / Bowel disease			
Pregnancy / Menses			
MLLB Precautions (Contraindications)			
Peripheral arterial disease	<input type="checkbox"/> ABPI < 0.5	<input type="checkbox"/> ABPI 0.6-0.8	<input type="checkbox"/> ABPI > 1.3

Medication (especially for medications linked to oedema)

Investigations (Tick box and space for results CT/Lymphoscintigraphy/MRI/ICG/duplex scan)
--

Social History

Occupation	
Hobbies	Smoker Yes/No
Accommodation (including type, access, stairs, bathroom/toilet etc)	
Sleeps in: <input type="checkbox"/> bed <input type="checkbox"/> chair	
Services / Carer Support	
Functional limitations	

Functional Assessment Measure

<input type="checkbox"/>	Bed bound
<input type="checkbox"/>	Wheelchair user
<input type="checkbox"/>	Mobile with assistance
<input type="checkbox"/>	Mobile independent with aid
<input type="checkbox"/>	Mobile independent without aid

Height (m)	Weight (kg)	BMI (kg/m ²)

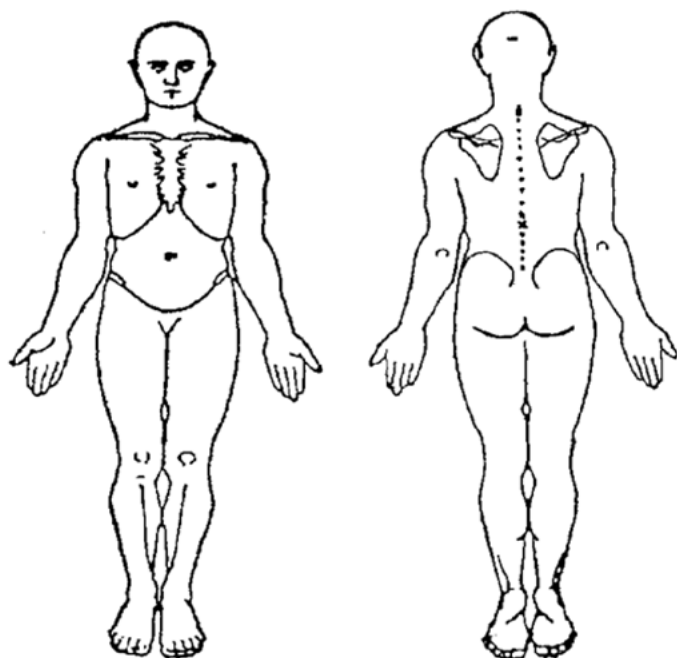
ROM

Upper Limb	ROM	Lower Limb	ROM
Neck		L/Spine	
Shoulder		Hip	
Elbow		Knee	
Wrist/Hand		Ankle/Foot	

Observations (✓ = present; x = absent; include key for body chart as necessary)

Key	√ / X		
		Broken skin (site)	
		Cancerous skin changes	
		Cellulitis	
		Discolouration	
		Dry	
		Fatty	
		Fibrotic	
		Fragile/Taut/Shiny	
		Fungal infections	
		Hyperkeratosis	
		Misshapen Limb	
		Lymphangiectasia	(lymph blisters)
		Lymphorrhoea	
		Non-pitting oedema	
		Papillomatosis	
		Pitting oedema	
		Scarring	
		Skin folds	
		Sensation	Intact/altered
		Stemmer's sign	RIGHT: Negative/positive LEFT: Negative/positive
		Temperature	RIGHT: Normal/cold/warm LEFT: Normal/cold/warm
		Genital oedema	<input type="checkbox"/> Y <input type="checkbox"/> N <input type="checkbox"/> N/A

Other:



Vascular Check List (√ = present; x = absent)

Arterial		Venous	
	Atrophic nail changes		Ankle flare (medial malleoli)
	Cyanosis		Atrophie blanche (white plaques)
	Diabetes		Dilated / varicose veins
	Distal ulceration - toes		Haemosiderin staining (purple/red/brown)
	Great toe pain		Lipodermatosclerosis (inverted bottle)
	65 years and older		Non-tender permanent redness
	Neuropathy		Soft pitting oedema
	Pain on exercise/cramps (intermittent claudication)		Ulceration or history of ulceration
	Red/blue discoloration when limb dependent		Varicose eczema
	Resting pain (on elevation) / night pain		Other:
	Slow capillary refill (i.e. takes more than 3 seconds)		
	Whiteness on elevation		

Doppler required Yes No

Date of Doppler _____

Doppler	Right	P	M	B	T	Left	P	M	B	T
Dorsalis Pedis or Toe										
Posterior Tibial										
Brachial Systolic										
T/ABPI (toe or highest ankle systolic / brachial)										

(P=Palpable pulse, M=Monophasic, B=Biphasic, T=Triphasic)

Risk Assessment	Low <input type="checkbox"/>	Medium <input type="checkbox"/>	High <input type="checkbox"/>
Mobility	Independently Mobile	Reduced Mobility (requires aid or assistance)	Immobile (assist of 2 or hoist)
BMI (kg/m2)	< 30	30-40	> 40
Patient Comprehension	Full Comprehension	Reduced Comprehension	Unable to comprehend
Vascular Status	ABPI 0.8-1.3 or ABPI not indicated from vascular checklist	ABPI 0.5-0.8 or pulses present and unable to obtain ABPI result	ABPI < 0.5 or inaudible pulses
Skin Integrity	Fully intact and good condition	Fragile and/or irritated skin	Ulceration/broken skin
Social Isolation	Independent or good support system	Has carers/family attending regularly	Socially isolated

Refer to the following risk assessments to manage any identified risks: domiciliary treatment, compression bandaging, open wounds, latex, vascular, handling heavy limbs, bandaging, mobilising with compression and local trust risk assessments.

Primary	Syndromes	
		Intestinal lymphangiectasia
Onset	Klinefelters syndrome	Noonan syndrome
Congenital	Klippel-Trenauney	Trisomy 21
Praecox	Lymphoedema distichiasis	Turner's syndrome
Tarda	Meige syndrome	Other:

Diagnosis (If there is more than 1 cause, rank in order (1 = most significant cause))

Secondary		
	Artificial Lymphoedema	Low albumin
		Self-harm
	Immobility and dependency	Dependency
		Obesity
		Paralysis
		Sleep apnoea
	Infection	Cellulitis/erysipelas
		Filariasis
		Lymphadenitis
		Tuberculosis
	Inflammation	Dermatitis/eczema
		Podoconiosis
		Pretibial myxoedema
		Psoriatic arthritis
		Rheumatoid arthritis
		Sarcoidosis and orofacial granulomatosis
	Malignant Disease	Infiltrative carcinoma
		Lymph node metastases
		Lymphoma
		Pressure from large tumours
	Trauma and tissue damage	Large/circumferential wounds
		Radiotherapy
		Scarring
		Varicose vein harvesting/surgery
		Burns
		Lymph node excision
	Venous disease	Chronic venous insufficiency
		Intravenous drug use
		Post-thrombotic syndrome
		Venous ulceration
	Medication Induced/related	
	Other	

ISL Lymphoedema Staging	0 - Latent/subclinical lymphoedema where impaired lymph transport and subtle changes are present, but swelling is not evident	II - Limb elevation alone rarely reduces swelling; pitting is manifest but may not be apparent later in Stage II due to the development of subcutaneous fat and fibrosis
	I - Accumulation of protein rich fluid that subsides with limb elevation and may cause pitting	III - Lymphostatic elephantiasis – pitting absent due to progressive development of fat and fibrosis, trophic skin changes and warty overgrowths develop
Lipoedema Staging	1 – Smooth skin, small nodules	3 – Induration and lobular fat deposits
	2 – Irregular texture larger nodules	4 – Obesity Related Lipoedema (lipolymphoedema)

Classification (√ Relevant Category)

Cancer related lymphoedema		Non cancer related lymphoedema	
CB	Breast	NCO	Obesity
CG	gynaecology	NCP	Primary
CHN	head and neck	NCV	Venous
CMel	melanoma	CELL	Cellulitis
COTH	Cancer (other)	NCDEP	Non cancer dependency/immobility
CU	Urology	NCLIP	Lipoedema

ICD 10 Codes

Code 189-0	Lymphoedema, not elsewhere specified
Code LO3	Acute lymphangitis
Code Q82.0	Hereditary lymphoedema
Code B74.9	Filariasis, unspecified
Code I97.2	Postmastectomy lymphoedema syndrome

Problem List and Goals of Treatment - discussed and agreed with patient Y/N

Problem List	Goals of Treatment	Goals of Treatment
Poor knowledge of lymphoedema	Increase knowledge of lymphoedema	Tissue softening
Increased limb volume	Reduce limb volume	Pain reduction
Altered limb shape	Restore normal limb shape	Improve AROM UL/LL
Tissue fibrosis	Improve skin integrity	Improve strength UL/LL
Poor skin condition	Patient able to carry out skincare regime	Able to carry out SLD
Reduced activity/exercise	Independent with exercise programme	Able to carry out MLLB
Pain	Patient able to don/doff garments	
Decreased range of movement UL/LL	Maintain stable lymphoedema	
Decreased strength UL/LL		

Treatment Plan: Specify review period _____, 3 months, 6 months, 1 year

Verbal Education	Garment Provision	Teach self-bandaging/wrapping
Written education provided:	Manual lymphatic drainage Frequency: Duration:	Arm MLLB leaflet
Exercise		Leg MLLB leaflet
Non-cancer advice	Multi-layer lymphoedema bandaging Frequency: Duration:	Physiotouch
Arm oncology		Intermittent pneumatic compression
Leg oncology	Bandaging cautions leaflet	Deep oscillation
Skin and nail care	Teach simple lymphatic drainage	Kinesio taping
Four key messages	Arm SLD leaflet	
Compression garment	Leg SLD leaflet	
	Head and neck SLD leaflet	

Reason for Modification (complete this section after intensive treatment period – most relevant reason)

Clinical decision	
Comorbidities	
Lack of resources	
Patient choice	

Onward Referral (✓ relevant category/categories)

Activity resources		GP	Physiotherapy (elsewhere)	Surgery
Complex treatment clinic		Obesity clinic	Plastic surgery	Treatment room nurse
Dermatology		Occupational therapy	Podiatry	Vascular
Dietetics		Oncology	Psychology	Wound care / tissue viability
District nurse		Palliative care	Sleep clinic	
Genetics		Practice nurse	Social work	

Compression Garment Information

Review Template for Adults living with Lymphoedema

Changes to History of Presenting Complaint & medical history:		Consent to assessment/treatment <input type="checkbox"/>	
Cellulitis episodes since last appointment?: Yes No			
Details/medication:			
Observations of oedematous limb/area (colour, temp, texture):			
Big toe pain		Neuropathy	
Leg pain/cramps on walking		Diabetes	
Cyanosis		Slow capillary refill (> 3 sec)	
Atrophic nail changes		Whiteness on elevation	
Distal ulceration (toes)		Pain on elevation / night pain	
Discolouration when limb dependent			
Weight (kg) increased/static/decreased		Height (m)	BMI (kg/m2)
QoL Outcome measures – EQ5D or LLS or VAS			

Circumferential measurement (see measurement chart)	
Pain (site) 0 (no pain) 10 (worst pain imaginable)	
Review of current maintenance programme	Management plan
Skin care:	New garments ordered/provided:
Exercise:	
SLD:	Treatment/advice provided:
Self MLLB:	
Garment(s): Worn? Every day <input type="checkbox"/> Most days <input type="checkbox"/> Occasionally <input type="checkbox"/> Never <input type="checkbox"/>	
Comfortable?	Ongoing 6 monthly review:
Swelling controlled?	
Other	Other:

II.III Children and Young People Lymphoedema Assessment and Review Template

All Ireland Children & Young People with Lymphoedema Assessment Form

Synopsis Page	
Informed consent obtained: No Yes	
Written consent obtained (for photographs) No Yes	
Lymphoedema Diagnosis	
Primary Type: Congenital: < 1 year Late onset: > 1 year Syndromic	
Systemic/visceral involvement (pre or post-natal)	
Disturbed growth / cutaneous manifestations / vascular anomalies	
Other:	
Secondary	
Cause of Secondary: Cancer Cellulitis Surgery Obesity Venous Disease Dependency/	
Skin Conditions Other:	
Lipoedema	
Site of Oedema	
No oedema identified	
Side: Right Left	
Arm: Upper Forearm Hand Fingers	
Leg: Thigh Below knee Foot Toes	
Midline: Breast Trunk Genital Head/Neck	
Patient Information Given:	
Leaflets given video/links given	
Patient/Carer wants correspondence in:	
English Irish	
Therapist Signature: _____ Date: _____	
Print Name: _____	

Assessment Form
Why have you come to see me today?
Current complaint as reported by patient/parent/carer*
How does it affect you day to day? (school/work/college/home/hobbies/exercise/wellbeing) *
What are the main parent/carer concerns/issues today?
Describe your limb / swollen area: :
History of Lymphoedema
Date swelling commenced
Onset: Gradual Sudden
Site: R arm L arm R leg L leg Genital Breast/Truncal Head & Neck
Trigger:
Getting worse Getting better Staying the same Fluctuates
Dominant side: Right Left Not evident
Eases oedema:
Worsens oedema:
Investigations: No Yes
Type:
Family history of lymphoedema: No Yes
If yes, 3 generations of Lymphoedema: No Yes N/A
If yes, genetics referral made No Yes N/A (refer to genetics service, UHW)
Comments:

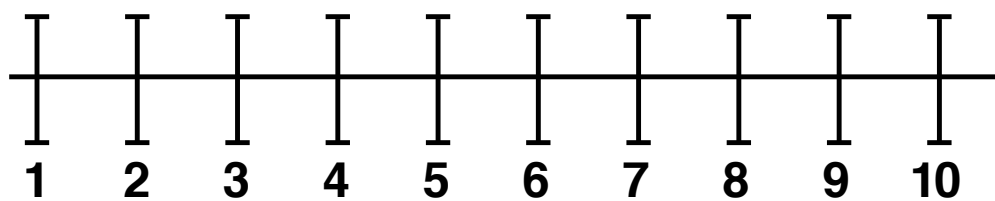
Pain: (related to lymphoedema site)	
Stated as: Unable to determine None Mild Moderate Severe	
Site: Constant Intermittent	
Description: Stabbing burning shooting ache	
0 (nothing)	10 (excruciating)
Heaviness: Unable to determine 0 (nothing) 10 (excruciating)	
Sensation: Unable to determine Normal Altered	
Medical History	
Medical Conditions:	
Cancer History: No Yes	
Lymph Node Surgery	
Clearance Sampling Sentinel Lymph Node Biopsy No. Positive:	
Radiotherapy No Yes N/A	
Date:	
Site:	
Chemotherapy No Yes N/A	
Type of chemo:	
Cellulitis	
Number of cellulitis infections: year:	No. of episodes within the last year:
Has the cellulitis required hospital admission? No Yes N/A, if yes number of nights	
Main antibiotic given	Duration:
Numbers of days off school/college/work due to cellulitis infections:	
On prophylactic antibiotics? No Yes Details:	Review date:
Has the patient had 2 episodes of cellulitis within the last year? No Yes	
Have you requested prophylactic antibiotics? No Yes	

Current Medication:	
Allergies:	
Penicillin: No Yes	Latex: No Yes Elastoplast: No Yes
Other:	
Social History:	
School/college/university:	Class name:
School nurse / special needs assistant / community nurse name:	
Occupation:	
Alcohol: No Yes	
Smoker: No Yes	
if yes how many:	
if ex-smoker, state when given up:	
Recreational drugs: No Yes Comment:	
Hobbies/activities:	
Home environment:	
Lives alone	Lives with
Sleeps in: Bed Chair Comment:	
Functional / ADL difficulties	
Additional sensory difficulties:	
Social support:	
Communication Function Classification System (CFCS) (please tick)	
CFCS Level I – a person independently and effectively alternates between being a sender and receiver of information with most people in most environments	
CFCS Level II – a person independently alternates between being a sender and receiver with most people in most environments, but the conversation may be slower	
CFCS Level III – a person usually communicates effectively with familiar communication partners, but not with unfamiliar partners, in most environments	
CFCS Level IV – the person is not consistent at communicating with familiar communication partners	
CFCS Level V – a person is seldom able to communicate effectively even with familiar communication partners	

Gross Motor Function Score				
* All the below questions relate to your Lymphoedema / vascular anomaly	None	A little	A lot	N/A
1. Are you worried about your lymphoedema?*				
2. Does your lymphoedema* cause you pain?				
3. Do you have any problems moving your body?				
4. How much do your scars bother you?				
5. How much does the heaviness of your limb bother you?				
6. Are you worried about getting cellulitis (infection)?				
7. How much does the appearance of your lymphoedema* worry you				
8. Does your lymphoedema* affect the clothes you wear?				
9. Does your lymphoedema* affect the shoes you wear?				
10. Does your lymphoedema* affect your hobbies?				
11. Does your lymphoedema* affect you attending school/work?				
12. Does your lymphoedema* affect relationships/friendships?				
13. Are you worried about wearing compression garments?				
14. Are you worried about your weight?				
15. Does your lymphoedema stop you doing any exercise?				

This is not applicable for this patient (please state reason) _____

Any other issues causing distress (in relation to lymphoedema)? _____



Using the scale shown with 0 = none and 10 = extremely

How much knowledge do you have about lymphoedema / vascular anomaly? _____

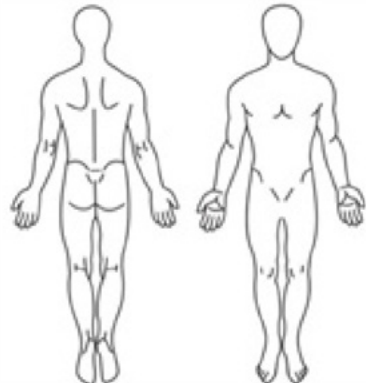
How confident are to help your child in managing their lymphoedema / vascular anomaly? _____

General Examination			
Dysmorphia (bodily abnormality, non-lymphoedema related)	No	Yes	
Vascular malformations	No	Yes	
Yellow nails	No	Yes	
Overgrowth of limb	No	Yes	
Cleft palate	No	Yes	
Ptosis(drooping of eyelid)	No	Yes	
Neck webbing	No	Yes	
Distichiasis (double eyelashes)	No	Yes	
Wide spaced nipples	No	Yes	
Ascites (fluid in abdomen)	No	Yes	
Venous disease	No	Yes	
Hydrocele/genital lymphoedema	No	Yes	
Incontinence	No	Yes	
Skin Assessment (please identify on body charts)			
Active cellulitis today?			
No	Yes		
Antibiotics requested? No Yes N/A			
Prescribed: No Yes N/A, if yes, prescribed by:			
Affected Body Part Description			
Discolouration: No Yes Detail:			
Temperature: Normal Cold Hot			
Shape: NormalDistorted Detail:			
Blisters	No	Yes	Hyperkeratosis No Yes
Taut	No	Yes	Shiny No Yes
Ulcer/wound	No	Yes	Lymphorrhoea No Yes
Skin folds	No	Yes	Fibrosis No Yes
Fatty	No	Yes	Fungal infection No Yes
Eczema	No	Yes	Rash No Yes
Seroma	No	Yes	Cording No Yes
Papillomatosis	No	Yes	Warts No Yes
Haemosiderin staining	No	Yes	

Tests			
Pitting Test: Location 1:	Positive Negative	Location 2:	Positive Negative
Stemmer's sign: Location 1:	Positive Negative	Location 2:	
Pain at night or at rest:	No	Yes	N/A
Intermittent claudication:	No	Yes	N/A
Capillary refill: (Normal is < 3 sec)			
Blanching on elevation:	No	Yes	N/A
Vascular assessment required:	No	Yes	N/A

Range of Movement (ROM)											
		RIGHT					LEFT				
		100%	75%	50%	25%	0%	100%	75%	50%	25%	0%
Shoulder	Hip										
Elbow	Knee										
Wrist	Ankle										
Fingers	Toes										

Functional Impact:

Body Chart	
Photo taken: No Yes	
Were circumferential measurements captured & recorded? Yes No - reason why not	
Was weight captured & recorded? Yes No - reason why not	
Was height captured & recorded? Yes No - reason why not	
Was BMI captured & recorded? Yes No - reason why not	
Consider percentile growth pattern (if appropriate)	

Education, Advice and Lymphoedema Care Plan
Assessment/findings:
What are the therapist's main concerns/issues today?
What are the patient's goals/hopes/aspirations?
What are the parent/carer goals/hopes/aspirations? (if applicable)
<i>The patient/parent/carer has agreed to take the following actions:</i>
Skin care:
Movement/exercise:
Compression garment required: No Yes; Details of garment:
Was garment: provided or ordered
Compression advice:
Weight/Lifestyle:
Simple Lymphatic Drainage:
Intensive Treatment: Please circle if the following is needed
Decongestive Lymphatic Therapy Multi-Layer Lymphoedema Bandaging LymphAssist Manual Lymphatic Drainage
Manual Therapy / Scar Management Electrotherapy (Physiotouch/Oscillator/Laser)
Detail:
Referral to other service required?: No Yes to
Completed? No Yes
Letter to patient/parent/carer copying in GP: <input type="checkbox"/> No <input type="checkbox"/> Yes, if No reason

Outcome of Lymphoedema Assessment					
Patient discharged. No Yes, Discharge code					
Factors Affecting the Outcome of Treatment					
Vascular complications	No	Yes	Mobility problems	No	Yes
Lack of support	No	Yes	Recurrent cellulitis	No	Yes
Pain	No	Yes	Progressive co-morbidities	No	Yes
Excess body weight	No	Yes	Cognitive impairment	No	Yes
Ability to don/doff garments.	No	Yes	Sedentary lifestyle	No	Yes
Functional problems	No	Yes	Chronic skin condition	No	Yes
Psychosocial	No	es	Sleeping in chair	No	Yes
LNW Outcome					
1 At Risk	2 Mild	3 Moderate	4 Severe	5 Complex	5W Complex with wound
ISL Staging					
Stage 0 (Latency)	Stage 1 (Mild/pitting)	Stage 2	Stage 3 (Moderate/non-pitting)	(Complex/fibrosis/skin changes)	
BLS Grouping					
Group 0 – Latent (at high risk)		Group 3.1 – Complex LO: one limb			
Group 1 – Early Lymphoedema		Group 3.2 – Complex LO: multiple limbs			
Group 2 – Uncomplicated (established) LO		Group 3.3 – Complex midline LO			
		Group 4 - Palliative			
Therapist Signature:					
Print Name:					
Date:					
Time:					
TIU		TIU total:		Date of next appointment:	

Modified and published with thanks to Lymphoedema Network Wales (V4.5 2020)

II.IV Genital Lymphoedema Assessment Templates

II.IV.I Lymphoedema Network Wales Genital Oedema Assessment Form

*(N.B. this is supplementary to main assessment/review documentation and to the patient self-report of GO document, LLGLQ).

Past Medical History influencing Genital Oedema			
Details including any other HCP involved			
Obesity	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Diabetes	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Cardiac oedema	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Gynaecology	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Urology	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Vascular/DVT	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Pelvic/skin cancer	<input type="checkbox"/> No	<input type="checkbox"/> Yes	(see also main assessment form) _____
Neurological disorder	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Liver/Renal disorder	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Perinatal complications	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Fertility treatments (< 3 months)	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Urinary problems/incontinence	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Bowel issues/faec. Incontinence	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Crohn's disease	<input type="checkbox"/> No	<input type="checkbox"/> Yes	_____
Recent/long-standing Sexually Transmitted Infection <input type="checkbox"/> No <input type="checkbox"/> Yes _____			
Removal of lymph nodes, other surgery or trauma likely to cause scarring in pelvis: <input type="checkbox"/> No <input type="checkbox"/> Yes (Describe possible link with genital oedema) _____			
Radiotherapy to pelvis or chemotherapy <input type="checkbox"/> No <input type="checkbox"/> Yes (if yes see main assessment document)			
Recent/current symptoms (follow up on responses in self-report form) Summarise identified problems with ADL's, change to sexual function or urinary/bowel function, symptoms/pain (type and onset?), daily fluctuations of oedema, emotional impact and how symptoms have changed over time.			
Additional questions on current symptoms that may need further investigation (red flags)			
	No	Yes	Comments and action
Sudden onset of oedema or sudden change in distribution?			
Recent onset of severe pain?			
Unexplained weight loss?			
Saddle anaesthesia? (numbness in buttocks/groin)			
Haematuria?			
Untreated UTI? (as opposed to recurring or persistent UTI)			
Any recent un-investigated urinary or bowel changes?			
(Female) Any unusual bleeding or discharge p/v?			
Rapid weight gain (days) after IVF?			
Infections – fungal infections, recent/recurrent/persistent urine infections, uterine or penile infections (give recent history and any treatments/health teams involved)			

Cellulitis of the genital area (if different to main assessment sheet)
 Number of past cellulitis infections: _____ No. of episodes within the last year: _____
 Has the cellulitis caused hospital admission? No Yes N/A, if yes, number of nights _____
 Main antibiotic given _____ Duration: _____
 Numbers of days off work due to cellulitis infections: _____
 On prophylactic antibiotics? No Yes Details: _____
 Has the patient had 2 episodes of cellulitis within the last year? No Yes
 Have you requested prophylactic antibiotics? No Yes

Current medication, allergies/sensitivities (review list on main assessment for hormonal/vaginal treatments, medications which increase thrush, other creams/gels or possible irritants applied to this area, also ask about soaps and self-medication/application of herbal remedies).

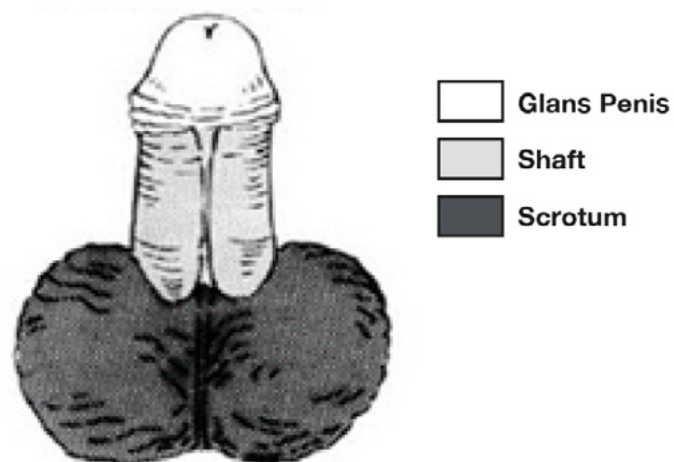
Social History (review main assessment for factors affecting genital oedema and its treatment, and consider challenges to hygiene (self-care/carers/shower/bath/frequency)).

Physical examination

Observation and palpation

Identify areas of swelling and consistency with lymphoedema? (Report groin mass or palpable bladder).
 Wounds, ulcerated lesions, fibrosis, nodules, warts, weeping, discharge, pitting, strong odour or other changes.

Male	Description of changes	Female



Patient discussion regarding GO (wishes/aims, fears/anxiety, agreed approach)

Summary of GO assessment.

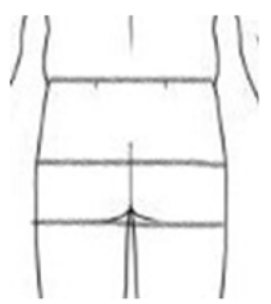
Plan in relation to GO (including referrals to other services whether direct or via GP)

Return to Main Assessment pages and incorporate GO assessment findings and plan.

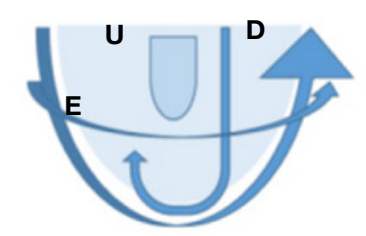
Description of changes to	pubic bone area / bikini line	
	sacrum	
	outer hips (iliac crests)	

*diagram adapted from Sobel et al 2005.

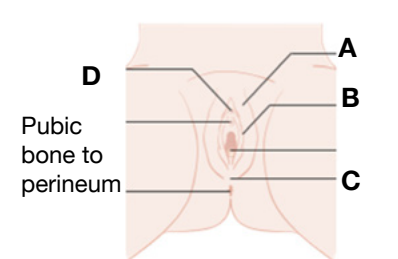
Measurements (as appropriate)



1 Waist/hip circumferences



2 Male scrotal measurements



3 Female measurements

Circumferences (cm)		Lengths and widths (cm)	
waist T		pubic bone to perineum (front to back) D	
widest part of hips H		outer labia A to C (left)	
under gluteal fold K		outer labia A to C (right)	
		width outer labia at midpoint (left) B	
Scrotum: anterior rim of anus to base of penis		width outer labia at midpoint (right) B	
penis circ. shaft midpoint (if visible)		penis length shaft and glans (if visible)	
horizontal circumference of scrotum E		underneath scrotum groin to groin U	

Note: Lymphoedema Network Wales devised these self-assessment tools for genital lymphoedema in both female and male patients based on a tool originally developed at the University of Glasgow

**Reproduced with thanks from UK Generic GLQ V1.1; 20 February 2018.
Original design Noble-Jones, University of Glasgow. Part funded by BLS 24.01.2014**

II.IV.II Lower Limb and Genital Lymphoedema Questionnaire for Women (LLGLQw)


Self-completion questionnaire for women who have lower limb oedema and may have genital area oedema / lymphoedema

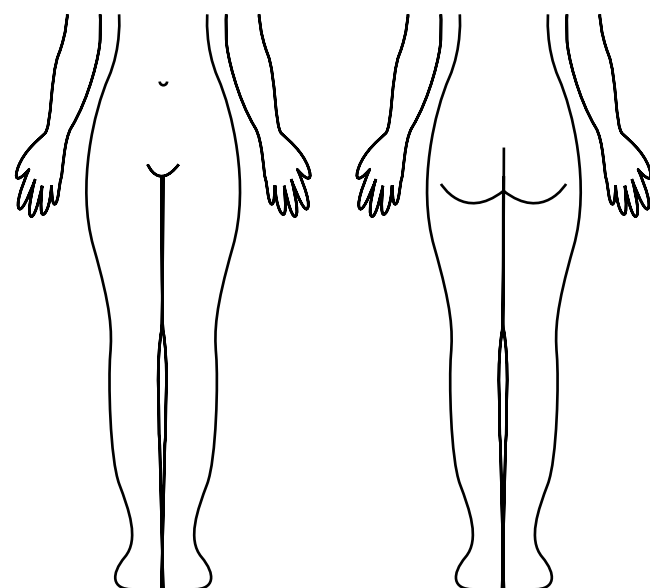
Name, contact details and DOB or hospital number:





Swelling in the legs/genitals can be quite normal for a few weeks after some treatments or with some chronic conditions. Sometimes these can be difficult to describe but this questionnaire may help. Please complete the questions below to help us give you the appropriate advice and care.

Personal Impact		Not at all (or not relevant) 0	A little bit 1	Quite a bit 1	Very much 1
Over the last month how the swelling affected your daily activities					
(for example)			✓		
I have swelling:	in my leg(s)				
	in my genitals				
If you feel you have no swelling at all you do not need to complete the rest of this questionnaire					
The swelling is worse by the end of the day					
The swelling is affecting:	which clothes/shoes I can wear				
	my sitting				
	getting in/out of bed				
	my walking				
	passing urine				
The skin around the swollen area:	my sexual function				
	feels tight				
	has changed colour				
	feels different				
The swelling gives me discomfort:	feels wet/cold				
	in my leg(s)				
	in my genitals				
I need to take painkillers for the discomfort					

Please continue overleaf/next page.

Therapist to calculate after completion	Score for Personal Impact Section (score above / 48) x 100= % limited	
If you have swelling of your legs or genitals please show in this picture where you feel it is, by shading like this: 		



Please tick any relevant		✓
	I feel swollen inside	
	The inside is sticking out	
	The outside is swollen	
	The inside and outside feels swollen	

On average this week how severe has the swelling been?				
	0 = No swelling	1 = A little bit	2 = Quite a bit	3 = Very swollen
Legs				
Genitals				

Is there anything else you would like to tell us about how this is affecting you physically or emotionally?

	Yes	No
During the last year, have you needed antibiotics for infections (cellulitis) in your leg(s) or genitals?		
Have you been offered any advice or treatment for the things you have identified here?		
Would you like to discuss this with us?		
Nurse/therapist to complete:		
Name of nurse/therapist _____ has discussed this form with the patient		
Signed: _____		
Date: _____		

II.IV.III Lower Limb and Genital Lymphoedema Questionnaire for Men (LLGLQm)

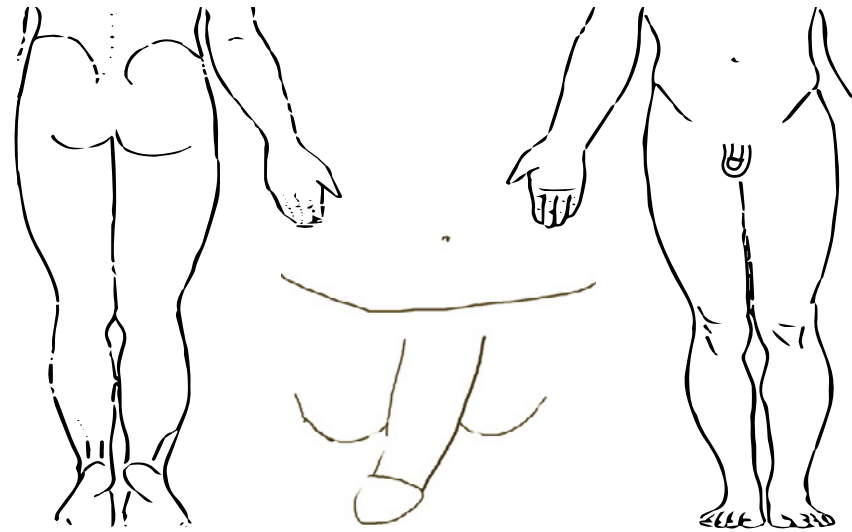
Self-completion questionnaire for men who have lower limb oedema and/or genital area oedema / lymphoedema

Swelling in the legs/genitals can be quite normal for a few weeks after some treatments or with some chronic conditions. Sometimes these can be difficult to describe but this questionnaire may help. Please complete the questions below to help us give you the appropriate advice and care.

Personal Impact	Not at all (or not relevant) 0	A little bit 1	Quite a bit 1	Very much 1
Over the last month how has the swelling affected your daily activities:				
Please give one tick per row (for example)				
I have swelling:				
in my leg(s)				
in my genitals				
If you feel you have no swelling at all you do not need to complete the rest of this questionnaire				
The swelling is worse by the end of the day				
The swelling is affecting:				
which clothes/shoes I can wear				
my sitting				
getting in/out of bed				
my walking				
passing urine				
my sexual function				
The skin around the swollen area:				
feels tight				
has changed colour				
feels different				
feels wet/cold				
The swelling gives me discomfort:				
in my leg(s)				
in my genitals				
I need to take painkillers for the discomfort				
Therapist to calculate after completion	Score for Personal Impact Section (score above / 48) x 100= % limited			

On average this week how severe has the swelling been?				
	0 = No swelling	1 = A little bit	2 = Quite a bit	3 = Very swollen
Legs				
Genitals				

If you have swelling of your legs or genitals please show in this picture where you feel it is, by shading like this:



Is there anything else you would like to tell us about how this is affecting you physically or emotionally?

	Yes	No
During the last year, have you needed antibiotics for infections (cellulitis) in your leg(s) or genitals?		
Have you been offered any advice or treatment for the things you have identified here?		
Would you like to discuss this with us?		
<i>Nurse/therapist to complete:</i>		
Name of nurse/therapist _____ has discussed this form with the patient		
Signed: _____		
Date: _____		

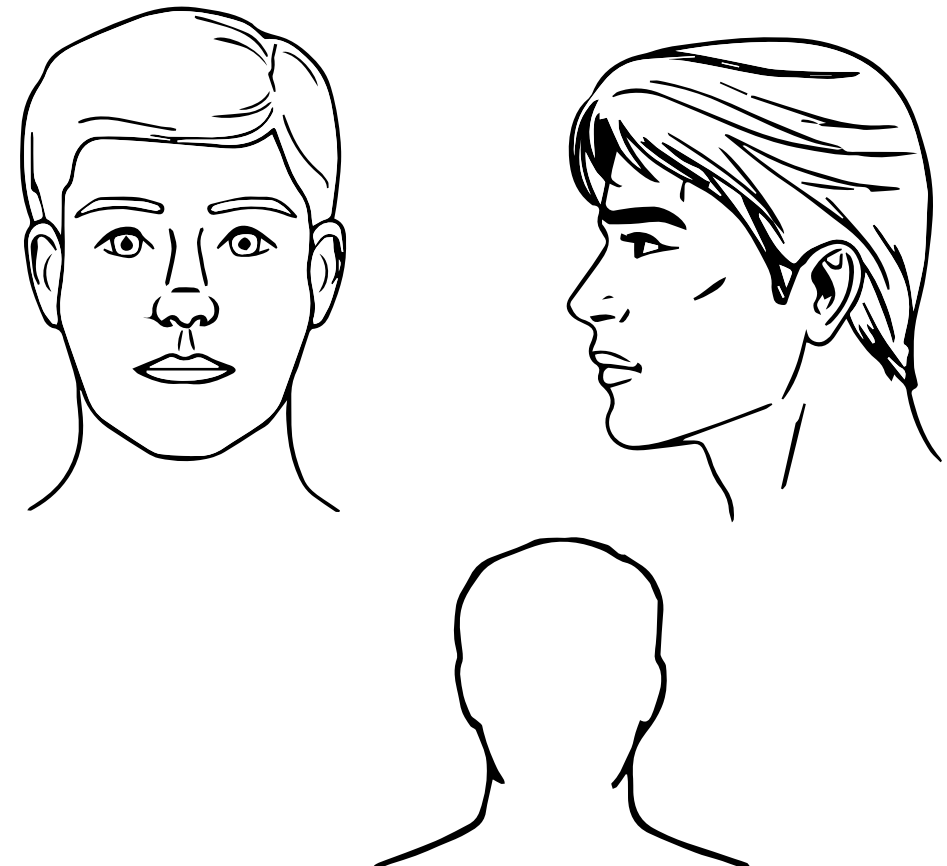
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II.V Head and Neck Lymphoedema Assessment Template

Head and Neck Lymphoedema Assessment

Pt Name	Date of Assessment	H&C No
Subjective (time of onset, increasing/decreasing factors)		
Diagnosis including staging		
Follow up treatment e.g. surgery, rtx, nodal involvement, no of nodes removed, no of affected nodes		

Visual examination (facial asymmetry, wounds, irradiation, location of oedema, skin changes)(face, neck, oral cavity, shoulders) & **Palpation** (tissue changes, heaviness, pitting, 'lumps and bumps')



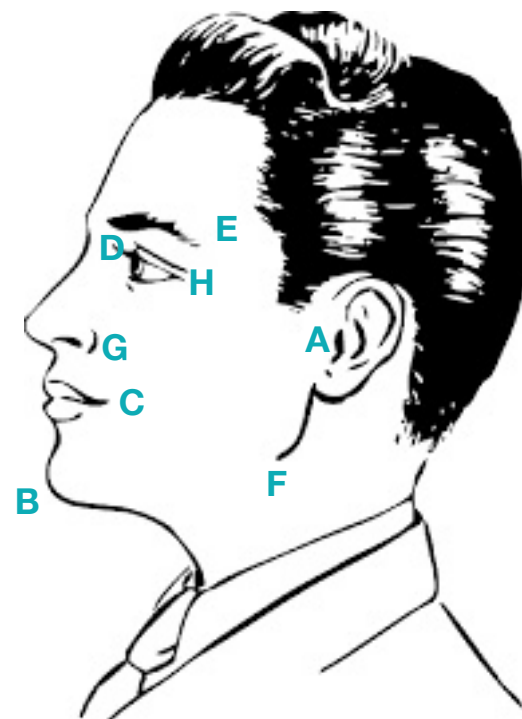
Intra oral (ask and observe- Palate, gums, cheeks, lips and tongue)

Cervical ROM

Flexion	
Extension	
Right Side Flexion	
Left Side Flexion	
Right Rotation	
Left Rotation	

Facial Measures

Measure		Right	Left
Tragus to mental protuberance	A-B		
Tragus to mouth angle	A-C		
Mandibular Angle to mental protuberance	B-F		
Mandibular angle to nasal wing	F-G		
Mandibular angle to internal eye corner	F-D		
Mandibular angle to external eye corner	F-H		
Mental protuberance to inner eye corner	B-D (vertical)		
TOTAL FACIAL COMPOSITE			



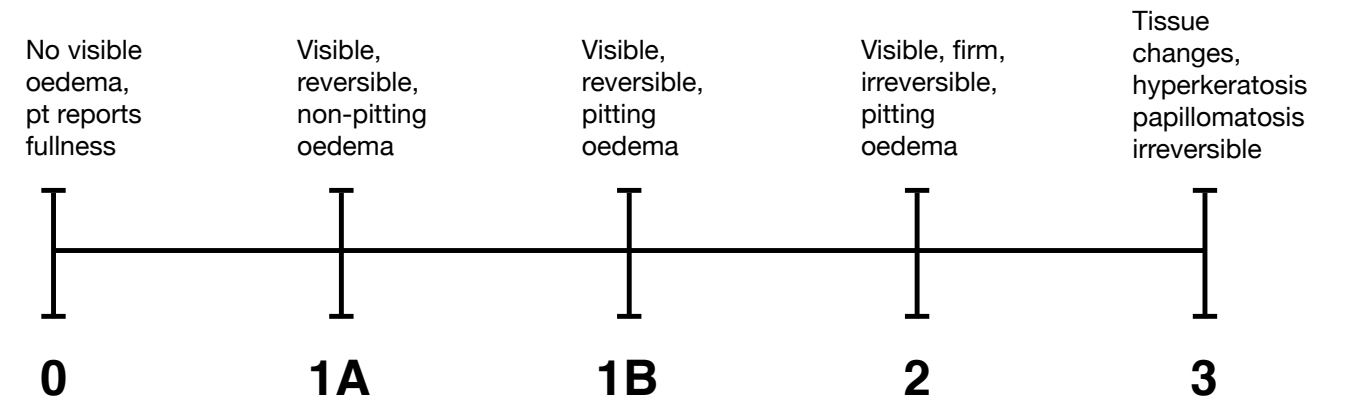
Neck Circumferential Measures

- Superior** (just below mandible) _____ cm
- Middle** (midway b/t top and bottom) _____ cm
- Inferior** (lowest circumferential location) _____ cm

Speech

Speech	(quality, Consider referral to Speech and Language Therapist)	
Swallow	(impairment, Consider referral to Speech and Language Therapist)	
Breathing	(? laryngectomy, tracheostomy)	

MDACC HNL Scale



Appendix III. Treatment Guidance Documents

III.I Standard Skin Care Protocol

<p>Patients should be encouraged to carry out skin care twice daily as follows:</p> <ul style="list-style-type: none"> • Conduct a thorough examination of the limb looking for signs of changes in the skin condition for example increased dryness, infection, injury, changes in shape because of the change in the distribution of oedema and signs of any of the conditions discussed in this document. • Use warm water to cleanse area comfortably and thoroughly. • Great care must be taken when washing and drying between the digits to reduce the risk of fungal infections. Consider using a spray cleanser if soap and water not available. • Moisturisation – patients should apply a moisturiser (note that the main aim of a moisturiser is to stop the evaporation of water from the skin) at least, once a day, preferably twice. This can depend on whether patients are able to apply their garments following moisturisation. For such patients, it is recommended to wait 30 minutes after applying cream before attempting to put on a garment because this will allow time for the preparation to be absorbed and make application easier. Moisturisation can also be completed by the patient (or carers) at night when their compression garments have already been removed. • Skin damage – reducing the risk of damage to the integrity of the skin, will reduce the chance of infection. Historically patients were advised to not let a medical practitioner take blood or give an injection into the affected limb. There is little robust evidence to support this advice, but any break in the surface of the skin creates an opportunity to trigger infection. It is therefore recommended to take all reasonable precautions to avoid any puncture wounds. • Great care should be taken when working in the garden to try to prevent cuts and insect bites. The use of protective gloves, for example, will reduce the risk of cuts. All cuts should be treated promptly with an antiseptic. • The removal of unwanted hair: it is recommended to remove hair above the hair follicle, i.e. not extract the hair from the follicle, and leave it exposed and therefore create a portal for infection. Careful consideration must be taken regarding plucking, waxing, and shaving over the swollen area (or area at risk of swelling). The use of an electric razor or depilatory cream is recommended, however individualised options should be discussed between the patient and HCP. 	
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Modified from An Introduction to Skin Care for those Managing Lymphoedema (Hobday, 2021).

III.II Compression Guidance: Indication for use of compression garments and associated pressure range

	Category 1a 14-17 mmHg	Category 1.b 18-21 mmHg	Category 2 22-30 mmHg	Category 3 31-40 mmHg	Category 4 41-50 mmHg	Category 4 super > 51 mmHg
Varices	Superficial or early varices Varices during pregnancy (Drug Tariff; BNF)	Varices of medium severity, ulcer treatment and prevention of recurrence, mild oedema, varices during pregnancy	Varices of medium severity, Severe varicose veins, Varices during pregnancy	Gross varices,		
DVT prophylaxis while travelling	Prevention of deep vein thrombosis for travellers	Prevention of deep vein thrombosis for travellers	Post thrombotic venous insufficiency, Prevention of deep vein thrombosis for travellers	Post thrombotic venous insufficiency		
Stage of treatment	Subclinical lymphoedema	Subclinical lymphoedema	Maintenance	Intensive management		
Stage of lymphoedema	Early/mild lymphoedema, ISL stages 0-II, No or minimal shape distortion	Early/mild lymphoedema, ISL stages 0-II, No or minimal shape distortion	Mild oedema, Gross oedema, Moderate/severe lymphoedema, ISL late stage II-III, Some shape distortion	Gross oedema, Severe lymphoedema, ISL stage III, Shape distortion Gross forefoot oedema, Retromalleolar swelling	Severe complex lymphoedema, ISL stage III, Shape distortion, 'Pressure resistant' (i.e., medium, or high pressure garments do not contain swelling)	Severe complex lymphoedema, ISL stage III, Shape distortion, 'Pressure resistant' (i.e., medium, or high pressure garments do not contain swelling)
Lipoedema	Lipoedema mild to moderate no deep skin folds	Lipoedema-mild to moderate no deep skin folds	Lipoedema moderate to severe	Lipoedema moderate to severe		
Cardiac disease	Controlled cardiac oedema	Controlled cardiac oedema				

Dependency	Dependency oedema	Dependency oedema				
Neurological	Neurological deficit	Neurological deficit				
Associated skin conditions	Primary prevention of spider and visible superficial veins; tired aching heavy legs, ankle flare; mild hyperkeratosis; hyperpigmentation; venous dermatitis:	Ongoing maintenance and early/medium intervention – varicose eczema/contact dermatitis, atrophie blanche, Severe varicose veins, moderate hyperkeratosis, healed ulcer, Recurring ulcer, cellulitis, chronic oedema (toes, feet, leg)	Ongoing maintenance and early/medium intervention – varicose eczema/contact dermatitis, moderate hyperkeratosis, atrophie blanche	Acute or chronic lipodermatosclerosis, severe hyperkeratosis, skin folds, papillomatosis, lymphangiomas, lymphorrhoea (wet legs)		
Ulcers			Ulcer treatment and prophylaxis and prevention of recurrence phlebolympoedema (healed ulcer), Healed ulcer, Recurring ulcer	Ulcer treatment and prophylaxis and prevention of recurrence phlebolympoedema (active ulcer),		
Other considerations	Patient tolerance, palliation, elderly/arthritic, pressure sensitive,	Patient tolerance, palliation, elderly/arthritic, pressure sensitive,	Patient tolerance	Patient tolerance	Patient tolerance	Patient tolerance

Appendix IV. Miscellaneous Guidance Documents

IV.I Clinical Problem Solving Flowchart

Is patient's condition maintained? YES → continue current maintenance programme	
NO → consider the following options:	
1. Is patient compliant with garment regime? (wears recommended garment(s) at all times (may include overnight))	
Yes	→ does patient have the correct choice of garment? (style, class, stiffness, flat knit, MTM, layering of garments, foam inserts)
	→ does patient need to wear overnight compression? (if limb refilling overnight - consider self MLLB, wraps, Mobiderm etc.)
	→ does patient need to do SLD? (revise practice and consider teaching family member if patient not capable)
	→ does patient need to do MLLB periodically? (revise practice and consider teaching family member if patient not capable)
	→ does patient participate in regular exercise? (encourage muscle pump exercises (as per leaflet), swimming, walking, Pilates etc.)
	→ is patient compliant with recommended daily skin care regime?
	→ is patient compliant with garment care regime? (as per individual manufacturer care instructions)
	→ does patient elevate legs regularly throughout day and sleep in a bed? (feet should be at least level with hips - review sleep pattern)
	→ does patient's condition generally deteriorate during a holiday? - if yes, review choice of holiday destination e.g. long haul, hot climate
	→ have you considered adjunctive treatments such as lymphatic taping, intermittent pneumatic compression?
	→ is patient a healthy weight? (discuss impact on lymphatics and a plan to increase activity and diet changes/ referral to other support)
No	→ why is patient not compliant with garment regime - are there any issues that we can address?
	→ are there donning/doffing difficulties? - yes -> review fit of garment; use of application aids; organise family member or carer to assist
	→ is garment causing irritation/discomfort? - if yes - review fit, type, material of garment
	→ are there any psychological issues that need to be addressed?
2. Does patient have regular AIEs?	
Yes	→ is patient on a low dose long term antibiotic therapy?
	→ does patient use antimicrobial emollient & cleanser on a daily basis?
	→ does patient suffer from fungal infections? - if yes, has the fungal infection been treated adequately?
	→ does patient need to be reviewed by dermatology?
	→ are patient's garments washed daily?
	→ does patient demonstrate good general hygiene e.g. shoes & clothes?

3. Does patient have an acute or chronic skin condition (including wounds)?	
Yes	→ does patient need to be reviewed by the GP or dermatology?
	→ does patient need to be reviewed by TVN or general nursing?
	→ has patient an appropriate skin care regime? (correct choice of emollient & cleanser)
	→ does patient require anti-fungal, anti-microbial or steroid treatment?
	→ are any of the creams, dressings or garments causing a skin reaction? (is there a clear demarcation line/contact element?)
	→ does patient need to be maintained in a wrap rather than compression garments? Or nighttime Mobiderm?
4. Does patient have pain or discomfort that they associate with their lymphoedema?	
Yes	→ have other sources of pain been excluded?
	→ is patient on optimal pain relief?
	→ is there a psychological component to the pain/discomfort that needs to be addressed?
5. Does patient present with increasing fibrosis?	
Yes	→ does patient have the correct choice of garment? (style, class, stiffness, flat knit, MTM, layering of garments, foam inserts)
	→ does patient need to wear overnight compression? (consider self-MLLB, wraps, Mobiderm etc.)
	→ does patient need to do SLD? (consider teaching family member if patient not capable)
	→ does patient need to do MLLB periodically? (consider teaching family member if patient not capable)
	→ is there a role for pneumatic compression, LymphaTouch or Deep Oscillation?
5. Does patient present with increasing fibrosis?	
Yes	→ does patient have the correct choice of garment? (style, class, stiffness, flat knit, MTM, layering of garments, foam inserts)
	→ does patient need to wear overnight compression? (consider self-MLLB, wraps, Mobiderm etc.)
	→ does patient need to do SLD? (consider teaching family member if patient not capable)
	→ does patient need to do MLLB periodically? (consider teaching family member if patient not capable)
	→ is there a role for pneumatic compression, LymphaTouch or Deep Oscillation?
6. Does patient present with an increased/increasing BMI?	
Yes	→ encourage participation in weight loss management programme - refer to dietitian, psychology, GP, bariatric services as appropriate
	→ encourage any increase in activity and ideally participation in exercise programmes (see LNNI website for local trust programmes)
	→ consider weight loss surgery if previous weight loss management programmes have failed
	→ if BMI > 40 kg/m2 refer to LNNI bariatric policy

IV.ii Minimum Data Set

MINIMUM DATA SET FOR LYMPHOEDEMA SERVICES

Date:

--	--	--	--	--	--	--	--

Day

Month

Year

Clinic -
Patient Number

Demographics			
1. Gender: <input type="checkbox"/> Male <input type="checkbox"/> Female <input type="checkbox"/> Other <input type="checkbox"/> Prefer not to say			Age: <input type="text"/>
2. Level of Obesity (please record BMI if available)			
BMI <input type="text"/>	<input type="checkbox"/> Obesity class III (BMI > 40)	<input type="checkbox"/> Normal (BMI 18.5-24.9)	
Or/Or	<input type="checkbox"/> Obesity class II (BMI 35-39.9)	<input type="checkbox"/> Underweight (BMI < 18.5)	
<input type="checkbox"/> Tick if level of obesity has been estimated only	<input type="checkbox"/> Obesity class I (BMI 30-34.9)	<input type="checkbox"/> Blank	
	<input type="checkbox"/> Overweight (BMI 25-29.9)	(WHO classification)	
3. Mobility			
<input type="checkbox"/> Bed bound	<input type="checkbox"/> Mobile independent with aid		
<input type="checkbox"/> Wheelchair User	<input type="checkbox"/> Mobile independent without aid		
<input type="checkbox"/> Mobile with assistance			
4. Classification of most likely cause. (Most likely cause at first assessment – please tick one of the causes)			

Wheelchair User		Yes		No	
Breast cancer				Non cancer - Primary	
Gynae cancer				Non cancer – Venous origin	
Urology cancer				Non cancer - Secondary to infection	
Head and neck cancer				Non cancer – Secondary to immobility	
Melanoma or other skin cancer				Non cancer – Secondary to Obesity	
Other cancer				Non cancer - Lipoedema	
				Non cancer – other	

Other secondary contributing factors:

No: Obesity Dependency Primary lymphoedema

Venous disease Infection Lipoedema

5. Is the care currently provided for the person considered palliative? (ie advanced progressive life limiting illness)

Yes No

6. Severity of the Swelling - ISL Severity Staging (please tick one)

<input type="checkbox"/> ISL stage 0	Subclinical state. Swelling not evident despite impaired lymph transport.
<input type="checkbox"/> ISL stage I	This represents early onset of the condition where there is accumulation of tissue that subsides with limb elevation. The oedema may be pitting at this stage.
<input type="checkbox"/> ISL stage II	Limb elevation alone rarely reduces swelling. There may or may not be pitting as tissue fibrosis is more evident.
<input type="checkbox"/> ISL stage III	The tissue is hard (fibrotic) and pitting is absent. Skin changes such as thickening, hyperpigmentation, increased skin folds, fat deposits and warty overgrowths develop.

7. Lymphoedema History (please tick one)

Length of time with symptoms prior to presentation for assessment

< 3 months; 4-6 months; 7 months-1 year; > 1-2 years;

> 2-5 years; > 5-10 years; > 10 years

8. Cellulitis

Has the patient ever had cellulitis?

In the past year, has the patient had cellulitis in the affected areas due to the swelling?

Yes No If yes, How many times:

In the past year, has the patient been admitted to hospital as a result of cellulitis?

Yes No If yes, How many times:

How many episodes of cellulitis in lifetime?

How many admissions to hospital due to cellulitis in lifetime?

9. Site of Oedema (check all that apply)

Comments

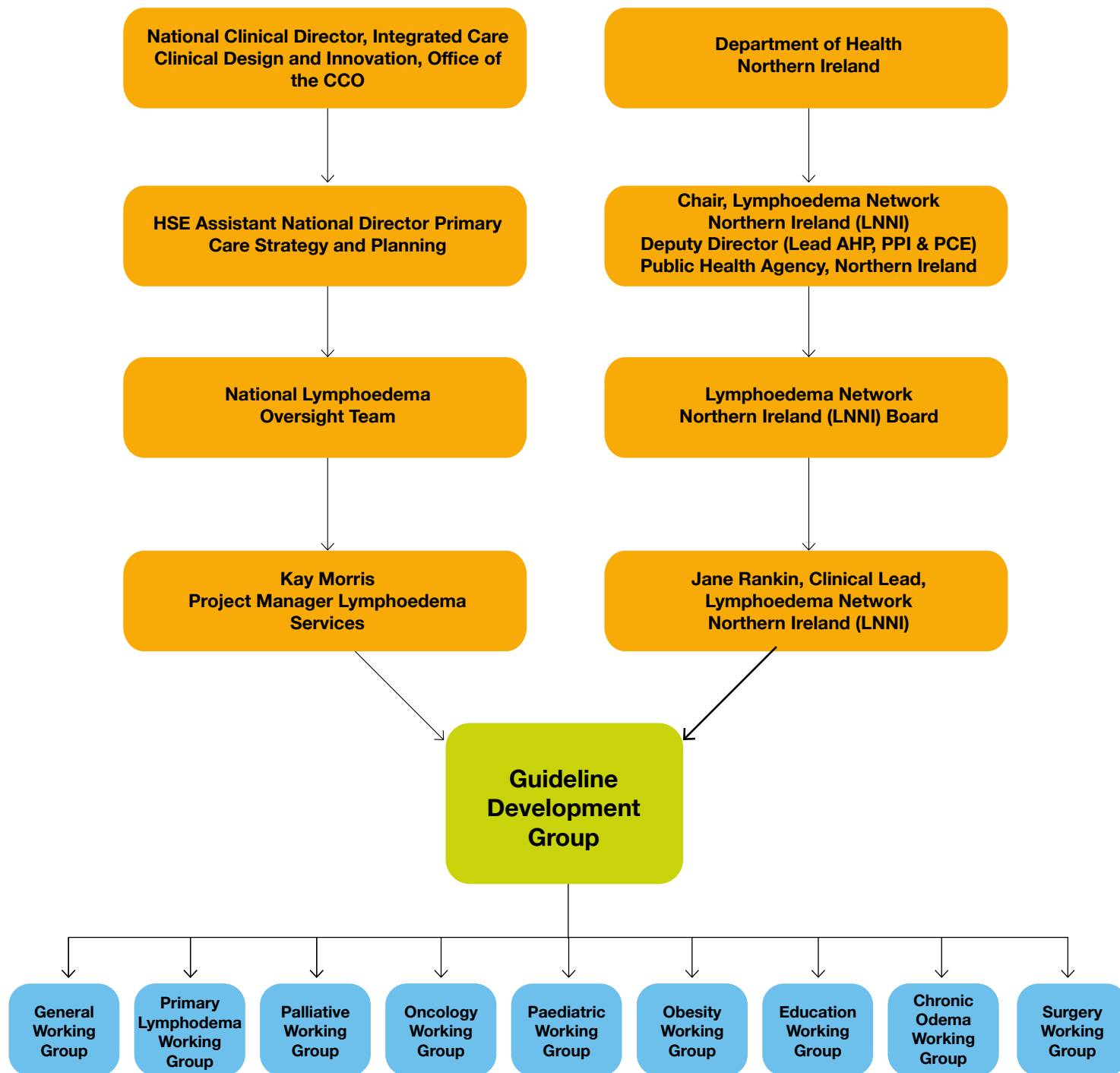
Upper Limb	<input type="checkbox"/> Yes <input type="checkbox"/> No	
Right arm Only	<input type="checkbox"/> Yes	
Left arm Only	<input type="checkbox"/> Yes	
Bilateral arm	<input type="checkbox"/> Yes	
Hand	<input type="checkbox"/> Yes	
Distal Only	<input type="checkbox"/> Yes	
Proximal (+/- root of limb)	<input type="checkbox"/> Yes	
Breast / chest wall Oedema	<input type="checkbox"/> Yes <input type="checkbox"/> No	
Right Only	<input type="checkbox"/> Yes	
Left Only	<input type="checkbox"/> Yes	
Bilateral	<input type="checkbox"/> Yes	
Lower Limb	<input type="checkbox"/> Yes <input type="checkbox"/> No	
Right leg Only	<input type="checkbox"/> Yes	
Left leg Only	<input type="checkbox"/> Yes	
Bilateral leg	<input type="checkbox"/> Yes	
Foot	<input type="checkbox"/> Yes	
Below knee Only	<input type="checkbox"/> Yes	
Above knee (+/- root of limb)	<input type="checkbox"/> Yes	
Truncal Oedema associated with leg oedema	<input type="checkbox"/> Yes <input type="checkbox"/> No	
	<input type="checkbox"/> Right <input type="checkbox"/> Left	

Appendix VI. Membership of the PPPG Development Group

Project Leads	
Ms. Kay Morris	Project Manager, Lymphoedema Services, PC Strategy and Planning, HSE
Ms. Jane Rankin MBE	Clinical Lead for Lymphoedema Network Northern Ireland (LNNI)
Guideline Development Group Members	
Dr. Mary Costello	RANP Tissue Viability and Lymphoedema Care, Mountmellick PCC, Laois
Dr. Jean O'Connell	Consultant Endocrinologist, St Columcille's and St Vincent's University Hospitals.
Ms. Judith McGlynn	Senior Physiotherapist, Community Physiotherapy Department
Ms. Shirley Real	Group Lead Allied Health, UL Hospitals Group
Ms. Catherine Quinn	Senior Physiotherapist in Oncology, Beaumont Hospital.
Ms. Deirdre Cullivan	Senior Physiotherapist, Mater Hospital
Ms. Claire Griffin	Nurse Tutor / Specialist Co-ordinator, Regional Centre of Nursing & Midwifery Education, Midland Regional Hospital, Tullamore.
Ms. Pamela Webster	Senior Occupational Therapist, Children's Health Ireland (CHI) at Crumlin
Guideline Working Group Members	
Ms. Sinead Cobbe	Senior Physiotherapist, University Hospital Limerick
Ms. Pippa McCabe	Lymphoedema Lead, South Eastern Health and Social Care Trust
Ms. Katie McCarthy	Dietitian, Weight Management Service, St Columcille's Hospital
Mrs. Keira O'Regan	Senior Physiotherapist, Lymphoedema and Kerry Specialist Palliative Care Unit
Ms. Emer O'Malley	Senior Physiotherapist, St Columcille's Hospital
Ms. Lynn O'Connor	Senior Physiotherapist, Oncology, Palliative Care and Lymphoedema, Sligo University Hospital
Ms. Lynsey Robertson	Occupational Therapist, Cork University Hospital
Ms. Helen Murphy	Senior Occupational Therapist / Lymphoedema Therapist, Cork University Hospital
Ms. Elaine Durkin	Tissue Viability Clinical Nurse Specialist, HSE
Ms. Fiona Fewer	Senior Physiotherapist Oncology / Lymphoedema, University Hospital Waterford
Ms. Gillian Carnduff	Vascular Nurse Specialist, Southern Health and Care Trust
Ms. Emily Coldrick	Community Nurse, Ashbourne, Co. Meath
Ms. Elaine Stowe	Lymphoedema Clinical Lead, Northern Health and Social Care Trust

Ms. Comfort China	Nurse Tutor, Royal College of Surgeons in Ireland
Ms. Maxine Shaw	Advanced Practitioner Physiotherapist, Burns and Plastic Department, Belfast Health and Social Care Trust
Ms. Emma Christie	Advanced Practitioner Physiotherapist in Palliative Care and Lymphoedema Management, Belfast Health and Social Care Trust
Ms. Gillian McCollum	Lymphoedema Lead (Job share), Belfast Health and Social Care Trust
Ms. Jeannie Donnelly	Tissue Viability Nurse Lead, Belfast Health and Social Care Trust
Ms. Jill Hamilton	Lymphoedema Lead, Western Health and Social Care Trust
Ms. Jill Lorimer	Lymphoedema Lead (Job share), Belfast Health and Social Care Trust
Ms. Louise Purcell	Senior Physiotherapist, Our Lady of Lourdes Hospital
Ms. Lynne Whiteside	Lymphoedema Lead, Southern Health and Social Care Trust
Ms. Mary Hickey	Senior Physiotherapist, HSE.
Ms. Paula Gorman	Senior Physiotherapist, Primary Care CHO 8
Ms. Thelma Dunne	Senior Physiotherapist, Mater Hospital
Ms. Patricia Walsh	Community Nurse, Rosscarberry, Co. Cork
Dr. Mary Paula Colgan	Associate Professor of Vascular Disease, Trinity College and St. James's Hospital
Patient Representative	
Ms. Ann Mulvin	Service user representative (Republic of Ireland)
Ms. Carolyn McKeown	Service user representative (N.I.)
Research Staff	
Dr. Aoife Reilly	Research Coordinator
Dr. Amy Blair	Research Assistant
Dr. Conor Mac Dermott Casement	Research Assistant
Mrs. Jane Burns	Research Librarian

Project Organogram



Appendix VII. Members of the External Review

The following individuals/organisations were invited to review this clinical guideline:

Allied Health Profession Leads Northern Ireland
British Lymphology Society - Children's Specialist Lymphoedema Interest Group (CLSIG)
British Lymphology Society – Scientific Committee
British Lymphology Society (BLS)
Chartered Society of Physiotherapy (CSP)
Department of Public Health Nursing, HSE
Guidelines and Audit Implementation Network (GAIN), Regulation and Quality Improvement Authority (RQIA), Department of Health, Northern Ireland
Individual family/carers representationw
Individual medical/nursing representation from Paediatrics, Dermatology and Vascular surgery
Integrated Care Programme for the Prevention and Management of Chronic Disease
International Lymphoedema Framework (ILF)
Irish Society of Chartered Physiotherapists (ISCP)
Lymphoedema Ireland
Lymphoedema Support Network (LSN)
Manual Lymphatic Drainage (MLD) Ireland
National Cancer Control Programme
National Clinical Programme for Dermatology
National Clinical Programme for Obesity
National Clinical Programme for Palliative Care
National Clinical Programme for Surgery
National Lymphoedema Framework Ireland
National Lymphoedema Partnership (NLP)
Northern Ireland Cancer Network (NICaN) Lead, Macmillan Partnership Lead, and Cancer Strategy 'Living Well' Lead
Northern Ireland Specialist Palliative Care Forum
Office of the Nursing and Midwifery Services Director (ONMSD), HSE
Oncology Physiotherapy Specialists, HSCTs, N.I.
Public Health Agency (PHA)
The Association of the Occupational Therapists of Ireland (AOTI)
University of Ulster – Nursing and Physiotherapy

The following individuals reviewed this guideline on behalf of their organisation and/or collated their groups reviews:

- Dr. Leanne Morgan, Clinical Lead, The Regulation and Quality Improvement Authority, NI
- Dr. Brian Creedon, National Clinical Lead for Palliative Care and Clinical Director National Office of Clinical Audit, HSE
- Ms. Margaret Sneddon, Chairperson, British Lymphoedema Society (BLS) , and the full BLS Executive and Scientific Committees
- Ms. Jane Nicklin, Network Facilitator, National Lymphoedema Partnership (NLP) , and the full National Lymphoedema Partnership committee
- Ms. Norah Kyne, Chairperson, National Lymphoedema Framework Ireland (NLF)
- Ms. Joni Meskell, Chairperson Lymphoedema Ireland
- Ms. Marie Todd, Lymphoedema Clinical Nurse Specialist on behalf of the BLS and the BLS Children's Lymphoedema Specialist Interest Group
- Ms Karen Morgan, National Lymphoedema Education and Research Lead in Wales, Lymphoedema Network Wales, on behalf of the BLS
- Dr. Melanie Thomas, National Clinical Lead for Lymphoedema in Wales, NHS Wales, on behalf of the BLS
- Dr. Andrew Hughes, Consultant in Palliative Medicine, NHS England, on behalf of BLS
- Ms. Margaret Anne Garner, Secretary Lymphoedema Network Scotland, on behalf of the BLS
- Ms. Virginia Pye National Lead for Public Health Nursing (PHN) Services, HSE
- Dr Kristiana Gordon, on behalf of the National Lymphoedema Partnership

Appendix VIII. Conflict of Interest Declaration Form Template

Conflict of Interest Declaration Form (Template)



CONFLICT OF INTEREST DECLARATION

This must be completed by each member of the PPPG Development Group as applicable

Title of PPPG being considered:

Please circle the statement that relates to you

1. I declare that I DO NOT have any conflicts of interest.

2. I declare that I DO have a conflict of interest.

Details of conflict (Please refer to specific PPPG)

(Append additional pages to this statement if required)

Signature

Printed name

Registration number (if applicable)

Date

The information provided will be processed in accordance with data protection principles as set out in the Data Protection Act. Data will be processed on to ensure that committee members act in the best interests of the committee. The information provided will not be used for any other purpose.

A person who is covered by this PPPG is required to furnish a statement, in writing, of:

(i) The interests of the person, and

(ii) The interests, of which the person has actual knowledge, of his or her spouse or civil partner or a child of the person or of his or her spouse which could materially influence the person in, or in relation to, the performance of the person's official functions by reason of the fact that such performance could so affect those interests as to confer on, or withhold from, the person, or the spouse or civil partner or child, a substantial benefit.

Appendix IX. Approved Policies, Procedures, Protocols and Guidelines Checklist

Title: All-Ireland Lymphoedema Management Guidelines 2022

Standards for developing Clinical PPPG	Checklist
Stage 1 Initiation	X
The decision making approach relating to the type of PPPG guidance required (policy, procedure, protocol, guideline), coverage of the PPPG (national, regional, local) and applicable settings are described.	X
Synergies/co-operations are maximised across departments/organisations (Hospitals/Hospital Groups/Community Healthcare Organisations (CHO)/National Ambulance Service (NAS)), to avoid duplication and to optimise value for money and use of staff time and expertise.	X
The scope of the PPPG is clearly described, specifying what is included and what lies outside the scope of the PPPG.	X
The target users and the population/patient group to whom the PPPG is meant to apply are specifically described.	X
The views and preferences of the target population have been sought and taken into consideration (as required).	X
The overall objective(s) of the PPPGs are specifically described.	X
The potential for improved health is described (e.g. clinical effectiveness, patient safety, quality improvement, health outcomes, quality of life, quality of care).	X
Stakeholder identification and involvement: The PPPG Development Group includes individuals from all relevant stakeholders, staff and professional groups.	X
Conflict of interest statements from all members of the PPPG Development Group are documented, with a description of mitigating actions if relevant.	X
The PPPG is informed by the identified needs and priorities of service users and stakeholders.	X
There is service user/lay representation on PPPG Development Group (as required).	X
Information and support is available for staff on the development of evidence-based clinical practice guidance.	X

Stage 2 Development	Checklist
The clinical question(s) covered by the PPPG are specifically described.	X
Systematic methods used to search for evidence are documented (for PPPGs which are adapted/adopted from international guidance, their methodology is appraised and documented).	X
Critical appraisal/analysis of evidence using validated tools is documented (the strengths, limitations and methodological quality of the body of evidence are clearly described).	X
The health benefits, side effects and risks have been considered and documented in formulating the PPPG.	X
There is an explicit link between the PPPG and the supporting evidence.	X
PPPG guidance/recommendations are specific and unambiguous.	X
The potential resource implications of developing and implementing the PPPG are identified e.g. equipment, education/training, staff time and research.	X
There is collaboration across all stakeholders in the planning and implementation phases to optimise patient flow and integrated care.	X
Budget impact is documented (resources required).	Budget impact deemed not necessary.
Education and training is provided for staff on the development and implementation of evidence-based clinical practice guidance (as appropriate).	X
Three additional standards are applicable for a small number of more complex PPPGs: Cost effectiveness analysis is documented. A systematic literature review has been undertaken. Health Technology Assessment (HTA) has been undertaken.	HTA and cost effectiveness analysis deemed necessary

Stage 3 Governance and Approval	Checklist
Formal governance arrangements for PPPGs at local, regional and national level are established and documented.	X
The PPPG has been reviewed by independent experts prior to publication (as required).	X
Copyright and permissions are sought and documented.	X

Stage 4 Communication and Dissemination	Checklist
A communication plan is developed to ensure effective communication and collaboration with all stakeholders throughout all stages.	X
Plan and procedure for dissemination of the PPPG is described.	X
The PPPG is easily accessible by all users e.g. PPPG repository.	X


Stage 5 Implementation	Checklist
Written implementation plan is provided with timelines, identification of responsible persons/units and integration into service planning process.	X
Barriers and facilitators for implementation are identified, and aligned with implementation levers.	X
Education and training is provided for staff on the development and implementation of evidence-based PPPG (as required).	X
There is collaboration across all stakeholders in the planning and implementation phases to optimise patient flow and integrated care.	X

Stage 6 Monitoring, Audit, Evaluation	Checklist
Process for monitoring and continuous improvement is documented.	X
Audit criteria and audit process/plan are specified.	X
Process for evaluation of implementation and (clinical) effectiveness is specified.	X

Stage 7 Revision/Update	Checklist
Documented process for revisions/updates and review, including timeframe is provided.	X
Documented process for version control is provided.	X

I confirm that the above Standards have been met in developing the All-Ireland Lymphoedema Management Guidelines 2022.

Name of Person(s) signing off on the PPPG Checklist:

Name:	Signature <u></u>
	Date <u>7/06/2022</u>

Appendix X. Search Strategy

1. Scoping Tasks

- Creation of search strategy (identifying keywords, search strings and MESH terms)
- Undertake rigorous search execution
- Creation of EndNote document library
- Support development of inclusion/exclusion guidelines
- Recommend data extraction tools & methods

2. Methodological approach

Using subject-appropriate databases, online discovery tools and accessing of repositories (primarily for grey literature) a comprehensive search of resources were undertaken. These were structured by a search strategy scaffold to ensure as much relevant information as possible was identified to support the development and updating of these guidelines.

3. Search Strategy Development:

Identification of key topics and subject headings was undertaken from a review of the literature.

Initial scoping was undertaken using PubMed for a number of reasons. The first being access to current medical publications in this subject area. The second being the capacity to develop search terms and search strings and the resulting verification of Medical Subject Headings (MESH). The MESH feature is not standard in all databases but is essential for double checking permutations for verification and sourcing.

• Medical Subject Headings (MESH)

MESH

MeSH Categories

Diseases Category

Hemic and Lymphatic Diseases

Lymphatic Diseases

Lymphoedema

Breast Cancer Lymphoedema

Elephantiasis

Elephantiasis, Filarial

Non-Filarial Lymphoedema

A broad subject search was undertaken in this subject area. Individual surgical or medical questions were reviewed as research packets. These were supplied with workshop and relevant articles. A Comprehensive list of all articles as well as comparative guidelines were sourced, shared and added to the master EndNote file.

Criteria Application:

Data Extraction Methods:

Step 1: Resulting bibliographic information from searches was included in an EndNote library. These titles along with their abstracts were reviewed as a first pass against the inclusion/exclusion guidelines.

Step 2. The articles identified as suitable for full text review were supplied. These were read in full and evaluated against the inclusion/exclusion guidelines. This were processed using EndNote where folders for inclusion and exclusion as well as 'unsure' were created and articles in these categories were migrated to the appropriate folder.

To underpin the systematic measurement of inclusion, a Prisma Flow chart was used to monitor the deductive process of article selection, which was key for the overall article management flow.

4. Outcomes of Interest

- Lymphoedema diagnosis
- Lymphoedema treatment
- Lymphoedema complications
- Lymphoedema pathophysiology

5. Inclusion/Exclusion Criteria

Inclusion/Exclusion Criteria: These guidelines are intended to support the standardisation of care and to encourage best clinical practice. The following criteria were applied:

Inclusion Criteria	Exclusion Criteria
Research Publications (1990 onwards)	Research Publications prior to 1990
Lymphoedema management	Exclude non lymphoedema complications
Lymphoedema treatment	Pregnant patients
Lymphoedema Care [General-inclusive]	
Surgical patients	
Obese patients	
Oncology patients	
Palliative care patients [Palliative care general]	
Paediatric patients	
Chronic oedema patients	
Education	

6. Methodology for Searching

A comprehensive literature review of existing lymphoedema management guidelines was undertaken which included national and international publications. Guidelines sourced were appraised by two reviewers, using the Agree II tool (Brouwers et al., 2010). Based on Agree II scores, decisions were made on which guidelines to include in the development of this document.

To address gaps in existing lymphoedema management guidelines, specific research questions were formulated using the population, intervention, comparison and outcome (PICO) framework and a literature search was undertaken to answer the questions posed. All results were reviewed by the work streams and helped in the generation of recommendations presented in this document. Searching and screening was conducted independently by each work stream, each consisting of at least 3 reviewers, which increased confidence that all relevant and current evidence were identified for the review. Refer to Section 7.0 (Search Strategy Development) for the full search strategy including databases and online search resources used.

The above inclusion and exclusion criteria were applied when searching for literature in the following domains: bibliographic databases including PubMed, Embase, Cochrane Library, LILACS, IBECs and relevant grey literature. Types of articles included systematic reviews, original research articles and grey literature. Initial overall subject searching was conducted for the topic of lymphoedema. The next steps in searching reviewed additional inclusion, exclusion parameters.

7. Search Strategy Development

Name	Subject Coverage
PubMed	Online version of Index Medicus produced by the US National Library of Medicine (NLM). Contains over 25 million records. In addition to Medline, Pubmed contains: <ul style="list-style-type: none"> • 'In process' citations • Some older citations • Citations to non-medical journals • Citations to eBooks Subject coverage = medical, biomedical & life sciences.
Cochrane Library Cochrane Reviews Other reviews Trials	Intervention and diagnostic reviews, critically appraised and re-structured abstracts, register of clinical trials
Medline	Three different versions: PubMed, OVID Medline and EBSCO Medline
Embase	An Elsevier resource focused on European studies, and conference abstracts.
Web of Knowledge	Conference abstracts, citation searching.
SCOPUS	Largest abstract and citation database of peer-reviewed literature: scientific journals, books and conference proceedings. Delivering a comprehensive overview of the world's research output in the fields of science, technology, and medicine.
CINAHL Complete	The definitive research tool for nursing and allied health professionals
Web of Knowledge	Social Science
ERIC	General Education

Grey Literature

Grey literature was also searched in line with the HSE Library's Guide to Grey Literature (<http://www.hselibrary.ie/east>) as well as the New York Academy of Medicine guidelines on Grey Literature (Grey Literature and Online sources searched:

Name	Reference Note
Google Scholar http://scholar.google.com/	Extensive range of articles in a range of related subject areas. Many open access articles and specialist articles are available.
Open Grey (http://www.opengrey.eu)	Resource for information on grey literature in Europe
NLM (National Library of Medicine, US) (http://www.ncbi.nlm.nih.gov) NLM Databases: http://www.nlm.nih.gov/nichs/db.html NLM Library Catalogue: http://locatorplus.gov/	Databases Indexed: <ul style="list-style-type: none">• Health Services Research Projects in Progress (HSRProj)• Health Services and Sciences Research Resources
Institutional repositories: • OpenDOAR (http://www.opendoar.org/) • Bielefeld Base (http://www.basesearch.net/Search/Advanced) · Lenus (http://www.lenus.ie/hse/) • RIAN (http://rian.ie/)	Digital collections of scholarly output from: <ul style="list-style-type: none">• Academic and professional organisations• International/European Research• Irish – HSE & Academic Research
Social Science Research Network (http://ssrn.com/)	Covers specialised research networks in the social sciences. Includes abstracts database of forthcoming papers and working papers as well as Electronic Paper Collection of full text documents.
Journal Author Name Estimator (JANE) http://jane.biosemantics.org/	The Journal/Author Name Estimator (JANE) is a free online bibliographic journal selection tool. Journal selection tools, also known as journal matching or journal comparison tools, are popular resources that help authors determine the most appropriate in scope journal to publish their manuscripts.
Prospero https://www.crd.york.ac.uk/PROSPERO/	International database of prospectively registered systematic reviews in health and social care. Key features from the review protocol are recorded and maintained as a permanent record in PROSPERO. The aim is to provide a comprehensive listing of systematic reviews registered at inception, to help avoid unplanned duplication.

Search Strings

The development of search strings included MESH terms, subject headings and keywords as an essential part of the overall searching methodology. The inputting of terms was matched via various algorithms to content of databases and other online resources. The goal was to be broad enough in scope to match the largest range of articles but narrow and focussed enough to capture the most relevant results.

```

1_ Search: Lymphoedema Filters: from 1990/1/1 - onwards
("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields])
Translations
Lymphoedema: "lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields]

2_ ((("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields]) AND 2002- onwards [Date - Publication]) AND (("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]) AND ("manage"[All Fields] OR "managed"[All Fields] OR "management s"[All Fields] OR "managements"[All Fields] OR "manager"[All Fields] OR "manager s"[All Fields] OR "managers"[All Fields] OR "manages"[All Fields] OR "managing"[All Fields] OR "managment"[All Fields] OR "organization and administration"[MeSH Terms] OR ("organization"[All Fields] AND "administration"[All Fields]) OR "organization and administration"[All Fields] OR "management"[All Fields] OR "disease management"[MeSH Terms] OR ("disease"[All Fields] AND "management"[All Fields]) OR "disease management"[All Fields]))))
Translations
Lymphoedema: «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields]
Patient: «patient s»[All Fields] OR «patients»[MeSH Terms] OR «patients»[All Fields] OR «patient»[All Fields] OR «patients s»[All Fields]
Management: "manage"[All Fields] OR "managed"[All Fields] OR "management's"[All Fields] OR "managements"[All Fields] OR "manager"[All Fields] OR "manager's"[All Fields] OR "managers"[All Fields] OR "manages"[All Fields] OR "managing"[All Fields] OR "managment"[All Fields] OR "organization and administration"[MeSH Terms] OR ("organization"[All Fields] AND "administration"[All Fields]) OR "organization and administration"[All Fields] OR "management"[All Fields] OR "disease management"[MeSH Terms] OR ("disease"[All Fields] AND "management"[All Fields]) OR "disease management"[All Fields]

3_ Search: Lymphoedema AND Surgical patients Filters: from 2002/1/1 - onwards
(("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields]) AND (("surgical procedures, operative"[MeSH Terms] OR "surgical"[All Fields] AND "procedures"[All Fields] AND "operative"[All Fields]) OR "operative surgical procedures"[All Fields] OR "surgical"[All Fields] OR "surgically"[All Fields] OR "surgicals"[All Fields]) AND ("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]))
Translations
Lymphoedema: «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields]
Surgical: «surgical procedures, operative»[MeSH Terms] OR («surgical»[All Fields] AND «procedures»[All Fields] AND «operative»[All Fields]) OR «operative surgical procedures»[All Fields] OR «surgical»[All Fields] OR «surgically»[All Fields] OR «surgicals»[All Fields]
patients: "patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]

```

4. Search: **(Lymphoedema AND (2002-onwards AND (management) AND Obese Patients NOT prior condition** ((("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields]) AND 2002/01/01:2021/05/31[Date - Publication] AND ("manage"[All Fields] OR "managed"[All Fields] OR "management s"[All Fields] OR "managements"[All Fields] OR "manager"[All Fields] OR "managers s"[All Fields] OR "managers"[All Fields] OR "manages"[All Fields] OR "managing"[All Fields] OR "managment"[All Fields] OR "organization and administration"[MeSH Terms] OR ("organization"[All Fields] AND "administration"[All Fields]) OR "organization and administration"[All Fields] OR "management"[All Fields] OR "disease management"[MeSH Terms] OR ("disease"[All Fields] AND "management"[All Fields]) OR "disease management"[All Fields]) AND 2002/[Date - Publication] AND ((("obese"[All Fields] OR "obesity"[MeSH Terms] OR "obesity"[All Fields] OR "obese"[All Fields] OR "obesities"[All Fields] OR "obesity s"[All Fields]) AND ("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields])))) NOT ((("prior"[All Fields] OR "priors"[All Fields]) AND ("condition s"[All Fields] OR "conditions"[All Fields] OR "disease"[MeSH Terms] OR "disease"[All Fields] OR "condition"[All Fields]))))

Translations

Lymphoedema: «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] **management:** «manage»[All Fields] OR «managed»[All Fields] OR «managements»[All Fields] OR «managements»[All Fields] OR «manager»[All Fields] OR «managers»[All Fields] OR «managers»[All Fields] OR «manages»[All Fields] OR «managing»[All Fields] OR «managment»[All Fields] OR «organization and administration»[MeSH Terms] OR («organization»[All Fields] AND «administration»[All Fields]) OR «organization and administration»[All Fields] OR «management»[All Fields] OR «disease management»[MeSH Terms] OR («disease»[All Fields] AND «management»[All Fields]) OR «disease management»[All Fields] **Obese:** «obese»[All Fields] OR «obesity»[MeSH Terms] OR «obesity»[All Fields] OR «obese»[All Fields] OR «obesities»[All Fields] OR «obesity-s»[All Fields] **Patients:** «patient-s»[All Fields] OR «patients»[MeSH Terms] OR «patients»[All Fields] OR «patient»[All Fields] OR «patients-s»[All Fields] **prior:** «prior»[All Fields] OR «priors»[All Fields] **condition:** «condition-s»[All Fields] OR «conditions»[All Fields] OR «disease»[MeSH Terms] OR «disease»[All Fields] OR «condition»[All Fields]

5. Search: **Lymphoedema AND Oncology patients** Filters: **from 2002-onwards** ((("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields]) AND ((("neoplasms"[MeSH Terms] OR "neoplasms"[All Fields] OR "oncology"[All Fields] OR "oncology s"[All Fields]) AND ("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]))))

Translations

Lymphoedema: «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] **Oncology:** «neoplasms»[MeSH Terms] OR «neoplasms»[All Fields] OR «oncology»[All Fields] OR «oncology-s»[All Fields] **patients:** «patient-s»[All Fields] OR «patients»[MeSH Terms] OR «patients»[All Fields] OR «patient»[All Fields] OR «patients-s»[All Fields]

6. Search: **(Oncology patients AND Lymphoedema** ((("neoplasms"[MeSH Terms] OR "neoplasms"[All Fields] OR "oncology"[All Fields] OR "oncology s"[All Fields]) AND ("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]) AND 2002/01/01:2021/05/31[Date - Publication] AND ((("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields])

Translations

Oncology: «neoplasms»[MeSH Terms] OR «neoplasms»[All Fields] OR «oncology»[All Fields] OR «oncology-s»[All Fields] **patients:** «patient-s»[All Fields] OR «patients»[MeSH Terms] OR «patients»[All Fields] OR «patient»[All Fields] OR «patients-s»[All Fields] **Lymphoedema:** «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields]

7. Search: **(palliative care patients AND (2002/1/1:2021/5/31[pmat]) AND Post-operative) AND (Lymphoedema AND (2002/1/1:2021/5/31[pmat]))** Filters: **from 2002/1/1 - 2021/5/31**

((("palliative care"[MeSH Terms] OR "palliative care"[All Fields] AND "care"[All Fields]) OR "palliative care"[All Fields] AND ("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]) AND 2002/01/01:2021/05/31[Date - Publication] AND ("postoperative period"[MeSH Terms] OR ("postoperative"[All Fields] AND "period"[All Fields]) OR "postoperative period"[All Fields] OR ("post"[All Fields] AND "operative"[All Fields]) OR "post-operative"[All Fields]) AND ((("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields])

Translations

palliative care: «palliative care»[MeSH Terms] OR («palliative»[All Fields] AND «care»[All Fields]) OR «palliative care»[All Fields] **patients:** «patient-s»[All Fields] OR «patients»[MeSH Terms] OR «patients»[All Fields] OR «patient»[All Fields] OR «patients-s»[All Fields] **Post-operative:** «postoperative period»[MeSH Terms] OR («postoperative»[All Fields] AND «period»[All Fields]) OR «postoperative period»[All Fields] OR («post»[All Fields] AND «operative»[All Fields]) OR «post-operative»[All Fields] **Lymphoedema:** «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields]

8. Search: **Chronic oedema patients** Filters: **from 2002/1/1 - 2021/5/31** ((("chronic"[All Fields] OR "chronical"[All Fields] OR "chronically"[All Fields] OR "chronicities"[All Fields] OR "chronicity"[All Fields] OR "chronicization"[All Fields] OR "chronic"[All Fields]) AND ("edema"[MeSH Terms] OR "edema"[All Fields] OR "edema"[All Fields] OR "oedemas"[All Fields] OR "oedema"[All Fields]) AND ("patient s"[All Fields] OR "patients"[MeSH Terms] OR "patients"[All Fields] OR "patient"[All Fields] OR "patients s"[All Fields]))

Translations

Chronic: «chronic»[All Fields] OR «chronical»[All Fields] OR «chronically»[All Fields] OR «chronicities»[All Fields] OR «chronicity»[All Fields] OR «chronicization»[All Fields] OR «chronics»[All Fields] **oedema:** «edema»[MeSH Terms] OR «edema»[All Fields] OR «edemas»[All Fields] OR «oedemas»[All Fields] OR «oedema»[All Fields] **patients:** «patient-s»[All Fields] OR «patients»[MeSH Terms] OR «patients»[All Fields] OR «patient»[All Fields] OR «patients-s»[All Fields]

9. Search: **(Lymphoedema AND (2002/1/1:2021/5/31[pmat])) AND (Education) AND (Patient Education)** Filters: **from 2002/1/1 - 2021/5/31**

((("lymphoedema"[MeSH Terms] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields] OR "lymphoedema"[All Fields] OR "lymphoedemas"[All Fields]) AND 2002/01/01:2021/05/31[Date - Publication] AND ("educability"[All Fields] OR "educable"[All Fields] OR "educates"[All Fields] OR "education"[MeSH Subheading] OR "education"[All Fields] OR "educational status"[MeSH Terms] OR ("educational"[All Fields] AND "status"[All Fields]) OR "educational status"[All Fields] OR "education"[MeSH Terms] OR "education s"[All Fields] OR "educational"[All Fields] OR "educative"[All Fields] OR "educator"[All Fields] OR "educator s"[All Fields] OR "educators"[All Fields] OR "teaching"[MeSH Terms] OR "teaching"[All Fields] OR "educate"[All Fields] OR "educated"[All Fields] OR "educating"[All Fields] OR "educations"[All Fields]) AND ("patient education handout"[Publication Type] OR "patient education as topic"[MeSH Terms] OR "patient education"[All Fields]))

Translations

Lymphoedema: «lymphoedema»[MeSH Terms] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] OR «lymphoedema»[All Fields] OR «lymphoedemas»[All Fields] **Education:** «educability»[All Fields] OR «educable»[All Fields] OR «educates»[All Fields] OR «education»[Subheading] OR «education»[All Fields] OR «educational status»[MeSH Terms] OR («educational»[All Fields] AND «status»[All Fields]) OR «educational status»[All Fields] OR «education»[MeSH Terms] OR «education-s»[All Fields] OR «educational»[All Fields] OR «educative»[All Fields] OR «educator»[All Fields] OR «educator-s»[All Fields] OR «educators»[All Fields] OR «teaching»[MeSH Terms] OR «teaching»[All Fields] OR «educate»[All Fields] OR «educated»[All Fields] OR «educating»[All Fields] OR «educations»[All Fields] **Patient Education:** «patient education handout»[Publication Type] .or. «patient education as topic»[MeSH Terms] .or. «patient education»[All Fields]

Imposed Limits

In the initial search there were no limits on the time frame of publication, format or languages as the search was intended to be as broad and as inclusive as possible ensuring the capture of all relevant evidence. The scope for the review was international and national so no geographical limits existed. A defined time limit of post 1990 was included to ensure capture of all literature since the last Lymphoedema guidelines (guidance notes) were published.

Data synthesis

Data synthesis was undertaken and a narrative summary of the data was provided to each work stream (Moore and Cowman, 2008).

1. Scoping Tasks: The scope of this review will incorporate the following tasks, all of which were needed to complete a substantive literature review of the evidence.

2. Methodological approach: The methodological approach was applied across four significant areas of investigation. These were:

- Search Strategy development
- Criteria application (Inclusion/Exclusion)
- Data Extraction Methods
- Data Analysis

3. Search Strategy Development: This involved the development of a robust, inclusive and replicable search strategy. This concentrated on developments in lymphoedema care management and practice since 2002.

- **Criteria Application:** The development and implementation of extensive inclusion/exclusion criteria was used to facilitate the identification of appropriate and relevant information in each of the areas identified in the research call.

- **Data Extraction Methods:** For purposes of this review, a detailed coding sheet was developed to capture essential bibliographic data, as well as inclusion/exclusion criteria, identification of new methods or clinical approaches for lymphoedema management and other developments in this area. In instances of disagreement between the reviewers, the final decision rested with the principal investigator (PI).

Data Analysis

The data analysis followed these steps: All sourced literature that met the inclusion criteria for the review was evaluated through an online template. Data was extrapolated from the online template to identify consistencies and inconsistencies in reviewer evaluation of the literature. Any inconsistencies were reviewed by the two chairs of the guideline. The AGREE II (Brouwers et al., 2010) 7 point scale for Clinical Guidelines was applied. The Evidence-Based Literature Critical Appraisal tool was used for all other literature. As part of the data analysis function, results were organised into themes and classifications.

Outcomes of Interest

The outcomes of interest were the most effective methods of: lymphoedema assessment, diagnosis, treatment and prevention. Clinical questions given to the research team by each the work stream groups were answered using the PICO model.

Appendix XI. Acknowledgments

Name	Reference Note
Prof Anne-Marie Tobin	National Clinical Programme for Dermatology, HSE
Dr Siobhán Ní Bhriain	National Clinical Director, Integrated Care, Clinical Design and Innovation, Office of the CCO
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Mr Robert Mercer	The Regulation and Quality Improvement Authority (RQIA) / GAIN, Northern Ireland
Ms Joan Hardy	Secondary care, Policy and Legislation, Department of Health and Social Care Northern Ireland
Mr Eamon Farrell	Allied Health Professions Consultant, Public Health Agency, Northern Ireland
Ms Ceara Gallagher	Allied Health Professions Consultant, Public Health Agency, Northern Ireland

Appendix XII Copyright/Permissions Sought

Below are a list of organisations/authors who were contacted to seek permission to re-produce/adapt and/or include content within this guideline:

- [British Lymphology Society \(BLS\)](#)
- National Lymphoedema Partnership (NLP)
- [Lymphoedema Network Northern Ireland \(LNNI\)](#)
- [International Society of Lymphology \(ISL\)](#)
- [International lymphoedema framework \(ILF\)](#)
- [Health Service Executive \(HSE\)](#)
- [Lymphoedema Network Wales](#)
- [European Reference Network for Rare Vascular Diseases \(VASCERN\) Guidelines](#)
- The Dutch lymphedema guidelines based on the International Classification of Functioning, Disability, and Health and the chronic care model (Damstra and Halk, 2017)
- Queensland Health lymphoedema clinical practice guideline (2014): The use of compression in the management of adults with lymphoedema
- The St. George's Classification Algorithm of Primary Lymphatic Anomalies (Gordon et al., 2021)
- University of Worcester (Hobday, A. 2021) An Introduction to Skin Care for those Managing Lymphoedema

Appendix XIII Glossary

Air plethysmography: a non-invasive test which measures change in calf volume in response to various manoeuvres and measures some pathophysiological mechanisms of chronic venous insufficiency.

Ankylosing spondylitis: an inflammatory arthritis that mainly affects the spine but can affect other joints.

Ankle Brachial Pressure Index: a non-invasive method of assessing peripheral arterial perfusion in the lower limbs. It is calculated by using a ratio of the blood pressure at the ankle to the blood pressure in the upper arm.

Ascites: an abnormal accumulation of swelling due to fluid in the abdomen.

Axilla: armpit or underarm

Axillary dissection: removal of lymph nodes from the axilla.

Axillary cording / axillary web syndrome: a condition that is common after removal of lymph nodes from the axilla. It involves scarring and hardening of soft tissues in the axilla and arm.

Bariatric: relating to or specializing in the treatment of obesity.

Cachexia: a disorder characterised by extreme loss of weight and muscle bulk.

Cellulitis: an infection of the skin and subcutaneous tissues.

Circumferential tape measurement: the use of a tape to measure the circumference of a limb at selected anatomic locations at repeated 4 cm intervals along the limb.. This is a common method used to calculate limb volumes and fit compression garments.

Complete decongestive therapy (CDT): the recognised conservative two-phased approach to the management of lymphoedema - synonymous with DLT.

Compression garments:

- **Flat knit:** knitted as a flat (made to measure) piece and joined with a seam. Material is firmer and thicker than a circular knit garment.
- **Circular knit:** knitted on a cylinder with no seam. Garments are shaped by varying stitch height and yarn tension.
- **Custom Made:** Made to exact measurements of an individual's body part and specially manufactured for that patient
- **Off-the-Shelf:** Standard garments that come in various sizes to fit many different patients

Compression: the degree of pressure (usually expressed as a range of mmHg) intended to be applied to an area of swelling.

Cytokine: a category of proteins involved in cell signalling. They regulate immunity, inflammation, and haematopoiesis.

Distichiasis: a condition characterised by a double-row of eyelashes.

Decongestive Lymphatic Therapy (DLT): the recognised, conservative two-phased approach to the management of lymphoedema - synonymous with CDT.

Deep Vein Thrombosis (DVT): A clot located in a non-superficial vein of the body.

Emollient: a class of moisturising treatments used to soften and hydrate the skin.

Endothelial Cells: thin flattened cells found lining blood vessels and lymphatics.

Erysipelas: a relatively common bacterial skin infection affecting the top layers of skin.

External lymphoedema: external lymphoedema is visible swelling on the outside of the body.

Fibrosclerotic tissue changes: a process where normally soft tissue such as fat begins to harden.

Folliculitis: Inflammation of hair follicles.

Haematoma: an abnormal collection of blood outside of a blood vessel.

Hemihypertrophy: excessive asymmetric growth of one side of the body.

Hydrops fetalis: a life-threatening condition where a newborn has an abnormal accumulation of fluid around the lungs, heart, or abdomen .

Hyperkeratosis: a thickening of the outer layer of the skin.

Hypertelorism: an abnormally increased distance between body parts usually referring to the orbits.

Indocyanine green lymphography: a minimally invasive imaging technique used to view lymphatic vessels using indocyanine green dye.

Internal lymphoedema: Internal lymphoedema is swelling in areas only visible with investigations e.g. airway.

Interstitial fluid: fluid within the tissues of the body.

Interstitial spaces: spaces within the tissues that are outside of the blood vessels are known as interstitial spaces or compartments. Most of the body's fluids that are found outside of the cells are normally stored in two spaces; the blood vessels (where the fluids are called the blood volume) and the interstitial spaces (where the fluids are called the interstitial fluid).

Intraluminal pressure: pressure within the lumen (inside) of a vessel.

Intermittent pneumatic compression: the action of devices placed around limbs that inflate and deflate to move fluid.

Juvenile rheumatic disease: inflammatory disease in children which can attack joints, muscles, bones, and organs.

Kaposiform haemangioendothelioma : a tumour which grows in blood vessels.

Kinesio tape: a proprietary therapeutic tape (and technique) which can lift the skin and may aid lymphatic movement and drainage.

Limb volume bioimpedance method: a measure of tissue resistance to an electrical current which estimates extracellular fluid volume.

Limb volume perometry method: use of an infrared optoelectronic system to measure limb volume.

Liners: an adjunctive garment to line the skin usually used to protect skin integrity

Lipoedema: a bilateral symmetrical swelling arising from the deposition of adipose tissue.

Liposuction: a surgical procedure where adipose tissue is removed.

Lipoaspiration: a procedure where a substance is injected under the skin which causes fat cells to shrink which can then be aspirated out of the body.

Lipofibromatosis: a benign paediatric soft tissue tumour arising usually in the distal extremities.

Lipolymphoedema: is a combination of lipoedema obesity and lymphoedema.

Low level laser therapy: the use of thermally safe energy level photons to alter biological activity.

Lymphangitis: an infection of the lymphatic vessels.

Lymphangiectasia: a pathologic dilation of lymphatic vessels.

Lymphadenitis: infection of the lymph nodes.

Lymphangiogenesis: formation of new lymphatic vessels.

Lymphaticovenous anastomosis: the connection of a lymphatic vessel to a nearby vein. Created surgically to drain target lymphatic fluid.

Lymphorrhoea: leakage of lymphatic fluid from skin.

Lymphangiomas: lymphatic blisters.

Lymphocyte: A white blood cell that creates an immune response when activated by a foreign molecule (antigen).

Lymphoedema: is the result of accumulation of fluid containing proteins and other elements in the tissue spaces due to an imbalance between interstitial fluid production and transport (usually low output failure). It arises from congenital malformation of the lymphatic system or from damage to the lymphatic vessels and/or lymph nodes.

Lymphoscintigraphy: is a nuclear medicine imaging modality involving injection of radioactive tracers to delineate lymphatic drainage patterns.

Maintenance phase: the phase where the patient or client should complete their own self-management including skin care, exercise, use of garments and self-performed manual lymphatic drainage.

Manual lymphatic drainage (MLD): a specific form of massage to stimulate the lymphatic system.

Medical nutrition therapy: Medical nutrition therapy (MNT) is an evidence-based approach used in the nutrition care process (NCP) of treating and/or managing chronic diseases, often used in clinical and community settings, that focuses on nutrition assessment, diagnostics, therapy, and counselling. MNT is often implemented and monitored by a registered dietitian and/or in collaboration with physicians and regulated nutrition professionals. For these guidelines, MNT is used as a standard language in nutritional therapeutic approaches for obesity interventions.

Milroy disease: a congenital disease characterised by lymphoedema.

Model of care (MOC): a description of how a service should ideally be delivered.

Multilayer lymphoedema bandaging (MLLB): a specialist bandaging technique used to encourage lymph movement and reduce fibrosis.

Massive localized lymphoedema: a non-malignant condition typically in obese patients where lymphatic tissue can mimic a tumour.

Neoadjuvant chemotherapy: chemotherapy that is delivered before the primary treatment (often surgery).

Noonan syndrome: a congenital disorder that causes parts of the body to not develop properly, often resulting in unusual facial features, short stature, and cardiac abnormalities.

Nutrition interventions: a term describing evidence-based nutrition-related approaches to improving health outcomes. Intentionally distinct from the term “diet” to broaden focus from weight-only focussed models.

Onychomycosis: a fungal nail infection

Optoelectronic perometry: use of an infrared optical scanner to calculate the volume of a limb.

Papilloma: a benign epithelial tumour.

Papillomatosis: a warty appearance of the skin due to fibrosis over dilated lymphatic vessels.

Paracentesis: a procedure in which a needle or catheter is inserted into the peritoneal cavity to aspirate ascitic fluid.

Parkes Weber syndrome: a congenital vascular anomaly that results in a child having many abnormal blood vessels.

Pectus excavatum: a condition in which a person’s breastbone is sunken into his or her chest.

Phlebolymphoedema: swelling caused by both chronic venous insufficiency (CVI) and lymphatic insufficiency.

Pitting oedema: swelling that leaves an indentation in the skin when pressed.

Prehabilitation: interventions completed to improve a patient’s health prior to an anticipated stressor, often surgery.

Psoriatic arthritis: a form of arthritis seen in patients with psoriasis.

Radical mastectomy: removal of the breast, underlying muscle, and associated lymph nodes.

Reflexology: an alternative medicine practice of massage. It is based on a pseudoscientific system involving hand and foot massage of zones and reflex areas purportedly to effect physiological changes to specifically targeted areas.

Sclerotherapy: a procedure involving injection of a substance to induce collapse and closure of a vessel.

Selenium: a chemical element found in soil, water, and some foods.

Sentinel lymph node biopsy: a surgical investigative procedure to identify presence and spread of malignancy to a local primary draining lymph node.

Seroma: a collection of fluid under the surface of the skin.

Short-stretch bandages: bandages designed to extend to no more than 60% of their original length when stretched. They provide the high working-pressure required for lymphatic drainage.

Single nucleotide polymorphisms: a variation at a single position in a DNA sequence which varies among individuals.

Skin necrosis: death of skin tissue.

Stemmer’s Sign: a test for lymphoedema which is positive when a skin fold at the dorsum of the fingers or toes cannot be lifted or is difficult to lift. The presence of this sign is an early diagnostic indication of lymphoedema. It has a low sensitivity and as such a negative Stemmer’s sign does not exclude a diagnosis of lymphoedema.

Subcutaneous tissue: tissue beneath the surface of the skin.

Subclinical lymphoedema: a stage of lymphoedema where swelling may not be visible but the limb may feel heavy or extra fluid may be evident on investigations.

Systemic sclerosis: an autoimmune condition causing degenerative changes and scarring in the skin, joints, and internal organs.

Telangiectasia: a condition in which small, linear red blood vessels are visible on the skin.

Thyrotoxicosis: an excess of thyroid hormone in the body causing clinical symptoms of hyperthyroidism.

Tissue dielectric constant (TDC): an index value describing the portion of an incident electromagnetic wave that is reflected to a sensor probe on the skin. Used in research as a marker of lymphoedema due to the relationship between TDC and local tissue water levels between patients.

Tonometry: a method used to quantify tissue compressibility or stiffness of tissue.

Tissue fibrosis/induration: a process characterised by the formation and deposition of excess fibrous connective tissue, causing tissue hardening.

Tissue viability nurse (TVN): a specialist nurse with expertise in wound healing, skin care and associated therapy.

Turner syndrome: a condition where when one of the X chromosomes (sex chromosomes) is missing or partially missing, causing developmental problems.

Ultrasound imaging: a minimally invasive radiology modality involving the use of sound waves and echoes to generate a variety of image types.

Vascular malformation: an abnormality in the development of blood vessels, usually congenital.

Venepuncture: accessing a vein usually to take a blood sample or to give an intravenous fluid.

Venous thromboembolism: an umbrella term including deep vein thrombosis and subsequent pulmonary embolus.

Venous occlusion plethysmography: a type of non-invasive measurement of blood flow in a limb.

Water displacement: Is a functional application of Archimedes' principle, which states that the volume of displaced fluid is equivalent to the volume of an object fully immersed in a fluid.

