



**GUIDELINES FOR THE
DIAGNOSIS, ASSESSMENT
AND MANAGEMENT OF
LYMPHOEDEMA**

February 2008

These guidelines have been published by the Clinical Resource Efficiency Support Team (CREST), which is a small team of health care professionals established under the auspices of the Central Medical Advisory Committee in 1988. The aims of CREST are to promote clinical efficiency in the Health Service in Northern Ireland, while ensuring the highest possible standard of clinical practice is maintained.

These guidelines have been produced by a multidisciplinary sub-group of health care professionals chaired by Dr Angela Garvey.

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or the Guidelines and Audit Implementation Network (GAIN) website at:
<http://www.dhsspsni.gov.uk/index/hss/gain.htm>

The guidelines along with additional learning materials and information for staff and patients may also be accessed from the Northern Ireland Lymphoedema Network web site.

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Contents

	Page No
Foreword	1
1. Guideline Development Process	3
2. Background & Introduction	5
3. Summary of Recommendations	9
4. Anatomy and Physiology of the Lymphatic System	15
5. Incidence and Prevalence of Lymphoedema	19
6. Diagnosis, Assessment and Classification of Patients with Lymphoedema	31
7. Management of Lymphoedema	51
8. Reducing the Risk of Lymphoedema	69
9. Lymphoedema Services in Northern Ireland	75
10. Educational Competencies	77
11. Audit and Research	79
12. Information Resources for Patients	81
Appendices	83
Glossary	114

Foreword

CREST was established almost 20 years ago to promote and standardise high quality services across the region while recognising the need to take account of economic constraints. CREST is currently amalgamating with the Regional Multi-professional Audit Group (RMAG) and the Northern Ireland Regional Audit Advisory Committee (NIRACC) to become GAIN (Guidelines and Audit Implementation Network). The work in promoting clinical excellence will continue through this new process.




These guidelines have been developed following the Regional Review of Services for Lymphoedema (2004) which highlighted a lack of evidence based guidelines.

Under the auspices of CREST, an expert panel was convened to review the evidence for diagnosing, assessing and treating lymphoedema. The panel included patients and health care professionals from varied backgrounds and was chaired by Dr Angela Garvey.

Lymphoedema is a swelling of body tissue due to failure in the lymphatic system and can affect people of all ages. It can be congenital or can develop as a result of cancer or its treatment or due to trauma or chronic infection. It is thought that somewhere in the region of at least 2.5 to 3 thousand people in Northern Ireland suffer from this chronic and incurable condition.

CREST would like to thank Dr Garvey, the members of the sub-group and all those who contributed in any way to the production of these guidelines. The patients who have been integral to this process have been invaluable in providing their expertise and experience for which we would like to extend special thanks.



DAVID GT STEWART
Chair of CREST



1. Guideline Development Process

1.1 Statement of Intent

Clinical guidelines are systematically developed statements which assist health care professionals in making appropriate decisions for specific clinical conditions. Guidelines aim to improve diagnosis and treatment of a particular condition and to reduce variations in clinical practice thereby improving the quality of patient care.

Guidelines are not intended to replace clinical judgement. Decisions on the most appropriate course of care for individual patients must take into consideration patient choice as well as evidence and a complete clinical picture.

1.2 The Guideline Development Process

As far as possible these guidelines for the diagnosis, assessment and management of lymphodema have been developed in accordance with recognised development tools such as those outlined by the Scottish Intercollegiate Guidelines Network (SIGN) and the National Institute for Health and Clinical Excellence (NICE).

Stakeholder Involvement

These guidelines have been developed by a CREST multidisciplinary group with strong user/patient membership. A list of the guideline membership group may be found in Appendix 1. Patients with lymphoedema have been involved at all stages of the guideline development.

Key Questions

The clinical questions to be covered by the Guidelines were developed to consensus by the multidisciplinary group taking into account patients' views, preferences and experiences. Whilst the guidelines also provide direction on service standards, education and patient advice, the main aim of the work is to provide recommendations for clinical practice based on available evidence. The main areas for which clinical evidence was sought includes the diagnosis and assessment of lymphoedema as well as its management and prevention. A full list of the questions may be found in Appendix 2. The literature search strategy is outlined in Appendix 3.

Review and Updating

This guideline was published in February 2008 and should be considered for updating in 2011.

2. Background & Introduction

The Regional Review of Lymphoedema Services DHSSPS 2004 recommended that guidelines for the management of lymphoedema should be developed to assist healthcare practitioners in the development of appropriate referral and treatment pathways for patients to ensure early identification and treatment of their lymphoedema.

Lymphoedema is a swelling of body tissue due to failure in the lymphatic system. It is chronic and incurable and requires life long management. Patients with lymphoedema also need psychosocial support.

Lymphoedema can affect people of all ages. It can be congenital or can develop as a result of cancer or its treatment or due to trauma or chronic infection. It can occur in a limb or limbs or in the head and neck, trunk or genital area. It may not become apparent for sometime after trauma (e.g. cancer surgery) and patients remain at risk of developing clinical lymphoedema at a later stage.

These guidelines will help patients at risk of lymphoedema or with primary or secondary lymphoedema have a diagnosis and treatment which is timely, effective, efficient, patient focused and with measurable outcomes. The guidelines aim to improve treatment and care for patients who suffer from:

- (1) Primary lymphoedema;
- (2) Secondary lymphoedema related to:
 - cancer of the upper limb or limbs;
 - cancer of the lower limb or limbs;
 - cancer of the head and neck, breast, trunk or genital area;
 - advanced malignancy;
 - people at risk of developing lymphoedema e.g. breast cancer related lymphoedema;
- (3) Secondary lymphoedema related to recurrent cellulitis, chronic inflammation or trauma;
- (4) Lymphoedema as a result of complex syndromes e.g. Turner's Syndrome.

2.1 The Impact of Lymphoedema

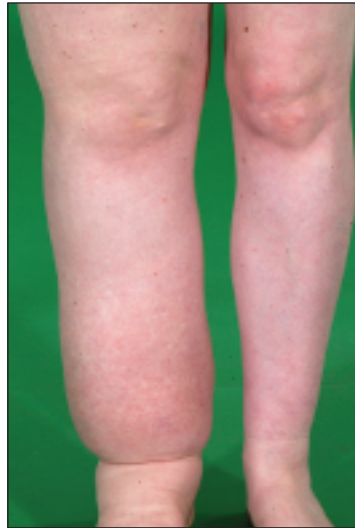
Lymphoedema is well documented as being an extremely debilitating condition:

The Physical Impact

- Patients reported difficulty in performing every day tasks.¹ Greater impairment in physical functioning was found between a breast cancer group with lymphoedema and a control group².
- In a study by Moffatt et al,³ 50% of the patients experienced pain and discomfort as a result of the disease.



Stage I unilateral upper limb lymphoedema



Stage II unilateral lower limb lymphoedema



Stage III bilateral lower limb lymphoedema

- Women with lower limb lymphoedema (LLL) said that it affected both their mobility and physical appearance.⁴
- Patients with upper or lower limb lymphoedema scored significantly lower in terms of physical health on a quality of life measure compared to a control group.⁵
- Physical wellbeing scales were reduced in patients with breast cancer related lymphoedema.⁶

The Psychological Impact

- Many patients experienced depression and anxiety, particularly the younger women.⁷
- Velanovich and Szymanski⁸ found that breast cancer related lymphoedema had a significant impact on mental health.
- Women reported that alterations to their physical appearance influenced their self-image.⁴
- Some patients reported feeling helpless and fearful about the future living with LLL.⁹
- Lymphedema patients scored significantly lower in terms of mental health on a quality of life measure, than a control group.⁵

The Psychosocial Impact

- Williams et al.⁷ found that patients experienced difficulties in opening up to their family and friends.
- Williams et al.⁷ also found that patients experienced problems in social settings due to the stigma associated with their appearance.
- In a cross-cultural study¹⁰ it was found that whilst there were no significant differences in terms of the impact of psychosocial factors between the countries, depressive coping and problematic social support exerted a negative effect on quality of life.
- Oedema caused 80% of patients within a catchment area to be off work, 9% to change their employment status, 2% to switch jobs and 8% to give up work entirely.³

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3. Summary of Recommendations


3.1 Lymphoedema: the Evidence

To date there has been limited robust research to inform evidence based guidelines on the diagnosis, assessment and treatment of lymphoedema. Despite an extensive review and appraisal of relevant literature, answers to many of the questions outlined by the group have proved inconclusive. It should also be noted that for some areas, such as diagnosis of lymphoedema, there is a moderate amount of good research studies which support the recommendations developed by the clinical experts in this area. A number of the guideline recommendations are therefore based on sound knowledge of physiological principles, along with the considered opinion and experience of clinical experts and patients suffering from the condition.

The recommendations in the following summary have not been graded; however evidence tables developed as a result of the review, outlining and grading the literature, are available on line through the Guidelines and Audit Implementation Network (GAIN) web site (www.dhsspsni.gov.uk/index/hss/gain.html).

This process of guideline development underlines the importance of developing and supporting relevant research to further the knowledge of the impact of lymphoedema on the population of Northern Ireland and how to best address it. Priorities for research are listed in section 11. It should also be noted that an absence of evidence for aspects of the treatment of any condition does not discharge the responsibility of providing a quality, equitable service for those affected.

The guideline development group are confident that the implementation of the guidance as outlined in this publication, will improve the services for and quality of life of those suffering from lymphoedema here in Northern Ireland.

 The following summarises recommendations made throughout the document. The flag symbol denotes the recommendation in the body of the text.

Summary of Recommendations

Diagnosis and Assessment of Lymphoedema

- Patients at known risk of developing lymphoedema as a result of cancer treatment should be informed of this risk and should be referred to lymphoedema treatment services for assessment if they complain of swelling, tightness or difficulty getting clothes or jewellery to fit in the affected limb.
- A complete physical and psychological assessment of all patients diagnosed with lymphoedema should be undertaken using appropriate visual analogue scales for pain and patient's impression of severity of lymphoedema.

- The SF36v2™ tool should be used to assess quality of life and psychological impact. This tool should only be used by practitioners who have been trained in its use and who are aware of trigger points for referral to psychological services.
- On the basis of a complete assessment, patients should be referred for further psychology, vascular or dietetic assessment, or to social services or occupational therapy services as necessary.
- Patients who have a family history of lymphoedema should be referred for genetic counselling.
- Patients who have a history of previous trauma to the lymphatic system e.g. previous cancer surgery, radiotherapy or recurrent cellulitis, should be referred to lymphoedema treatment services for assessment when:
 - Visible swelling is evident;

OR

 - Where there is a 5% increase in circumference at any site in uni-lateral lymphoedema compared to the other limb or compared to a pre-treatment measurement;

OR

 - Where the patient complains of symptoms, which would suggest lymphoedema such as tightness of clothing or jewellery or heaviness in the limb.
- Limb volume measurements should be calculated from circumferential measurements of the limb from fixed anatomical points using 4 cm repeated measurements. These measurements should be used to calculate individual limb volumes.
- All practitioners assessing patients or treating patients with lymphoedema should have access to a limb volume computer programme or calculator.
- Multiple frequency bioimpedance measurement has advantages over measurement of limb circumference in that it is applicable to bilateral limb lymphoedema (as the limb can be used as its own control).
- Limb volume measurements should be made as a baseline prior to treatment (e.g. surgery or radiotherapy), which is likely to cause lymphoedema.
- The diagnosis of lymphoedema is often confirmed on the basis of history and clinical examination. Radiological investigations such as CT, MRI, or ultrasound should be carried out where malignant lymphoedema is suspected. They may also have a role in differentiating lipoedema from lymphoedema in complex cases.

- Lymphoscintigraphy has a role in establishing the diagnosis of lymphoedema where clinical history and clinical examination are inconclusive. It is important to be certain of the diagnosis of lymphoedema before embarking on a life long programme of treatment. A uniform protocol for performing and reporting lymphoscintigraphy in lymphoedema should be developed for Northern Ireland.
- Multidisciplinary meetings should be held on a regular basis to discuss complex patients and to review issues such as skills training, audit, recent research and service development.

Management of Lymphoedema

- All patients with a diagnosis of lymphoedema should be referred to the trust's lymphoedema team. Lymphoedema should be treated as early as possible in order to prevent complications, such as infections, fibrosis and increased swelling.
- All patients with a diagnosis of lymphoedema should be referred to a therapist who has been fully trained, with a formal qualification from a recognised school of lymphoedema management.
- A thorough assessment including physical and psychosocial needs should precede any treatment.
- Following assessment, the patient should be referred to appropriate members of the multidisciplinary team as deemed necessary.
- Complex decongestive therapy (CDT) is recognised as the treatment of first choice for lymphoedema. The precisely co-ordinated interaction of manual lymphatic drainage (MLD), multi-layer lymphoedema bandaging, skin care and exercise is effectively combined as (CDT) for patients with lymphoedema.
- CDT may need to be modified in the presence of complex co-morbidities or due to patient choice. Clinical judgement may also recommend modified CDT in some circumstances. The reasons for modifying the treatment should be clearly stated on the patient's treatment plan, and be subject to audit.
- Treatment options should be discussed with the patient (and potentially carers) and an individualised patient-centred protocol agreed. If accepted by the patient and deemed appropriate by the therapist, the patient will undergo a period of CDT.
- The second phase of treatment (maintenance) encourages the transfer of control to the patient. This may include teaching the principles of nocturnal self bandaging. Initially six-monthly reviews should be therapist led. However, telephone reviews should be considered if the patient's condition is considered to be stable and the patient competent in self-monitoring.

- Patients with lymphoedema require lifelong specialist compression garments. All patients should have a minimum of two effective and appropriate compression garments at a given time (no longer than six months apart).
- Surgery is not currently recommended for the management of this condition in Northern Ireland.
- Patients with lymphoedema carry a small risk of a malignant sarcoma developing in chronic lymphoedema. Health care professionals should be vigilant in detecting angiosarcoma. This tumour is treated surgically.
- Systematic reviews of research have indicated that there is not enough evidence for the use of benzopyrones in the management of lymphoedema.
- In the presence of related cellulitis refer to the “CREST Guidelines for the Management of Cellulitis in adults”. (Appendix 7)
- All patients should be provided with accurate contact details of local lymphoedema services if advice is required.
- Patient education should include adequate information on lymphoedema and its management. Whilst given verbally, this must be supported in written format.

Reducing the Risk of Lymphoedema

- Improving knowledge about prevention is key to reducing the impact and burden of lymphoedema. More long term research studies are required to investigate risk factors for developing lymphoedema in patients undergoing surgical or radiotherapy treatment.
- As infection is intrinsically linked to lymphoedema, prompt assessment and treatment is vital to prevent recurrence and hospital admission. (refer to CREST Cellulitis Guidelines).
- Awareness of the risk of lymphoedema should be raised with all patients prior to undergoing any form of treatment which is likely to cause lymphoedema and should be included in the consent process.
- Awareness of the signs and symptoms of lymphoedema and appropriate referral pathways should be cascaded to all relevant health care professionals, particularly within primary care and the specialties of oncology, palliative care, vascular surgery, genetics and dermatology.
- Patients with an increased body mass index (BMI), particularly a BMI over 30, may be at a higher risk of developing lymphoedema and as such, should be referred to dietetic services.

- Adequate in-depth education regarding lymphological disorders and correct treatment should be taught at undergraduate level to all relevant health care professionals.

Lymphoedema Services

- Each trust should establish a dedicated lymphoedema service to identify and address the needs of patients with, or at risk of developing, lymphoedema. A suggested profile of how the teams should be configured is outlined in Appendix 9.
- All trusts should ensure that the lymphoedema guidelines are implemented in their area.
- All patients with a diagnosis of lymphoedema should be referred to a trust lymphoedema team. A proposed service referral form is provided in Appendix 10. Patients with lymphoedema should be referred to the lymphoedema service as per the referral pathway outlined in Appendix 11.
- All patients with a medically complex presentation should have access to appropriate consultant input via regional complex lymphoedema clinics (see Appendix 12 for referral criteria).
- Ongoing information should be collated on patients to support service evaluation and future planning. A review form is provided in Appendix 13.
- An education and awareness programme about lymphoedema should be established for all relevant health care professionals.
- A regional clinical network should be formed to co-ordinate service development, research, education, patient support and other governance activities.
- The lymphoedema network should promote and support the implementation of the CREST Guidelines for Lymphoedema across Northern Ireland.
- A database of patient information should be established to support service evaluation, research and future planning.

Education and Training

- All health care professionals who may come into contact with patients with lymphoedema or at risk of developing the condition should be aware of the signs and symptoms along with the relevant diagnostic and treatment pathways.

Audit and Research

- Audits should be undertaken using standardised proformas developed by the Lymphoedema Network where available.
- Partnerships between relevant institutions such as the Lymphoedema Network, Universities and the Research & Development Unit, should initiate research activity to address identified research priorities.

4. Anatomy and Physiology of the Lymphatic System

This simplistic explanation aims to provide a brief overview of what the lymphatic system is and its purpose.

The lymphatic system interacts with three other main systems in the body. It helps maintain fluid balance within the cardiovascular system, aids fat absorption in the digestive system and plays a major role in the body's immune or defence system.

How does the lymphatic system play a part in fluid balance?

The cardiovascular system, through its network of blood vessels, delivers oxygen and nutrients to the body's cells. As the blood passes through the capillary vessels, water and nutrients pass continuously into the space between the cells, known as interstitial spaces, to form interstitial fluid. The interstitial space acts as a reservoir from which the cells can absorb what they need from the surrounding fluid. Tissue cells also excrete waste matter and excess fluid back into the interstitial space. There may also be antigens or pathogens present in the interstitial fluid which may cause an immune response.

Some of the fluid and waste products pass back into the bloodstream through the capillary walls and return to the heart via the veins. However, much of the fluid and protein which cannot pass back into the blood stream is picked up by the lymphatic system. The lymphatic system is a network of vessels that is present in almost all of the body's vascularised tissue. Once fluid enters the lymphatic system, it is called lymph.

The lymphatic circulation is made up of two parts. The superficial system drains the tissues of the skin and a deep system is found in the skeletal muscle. Both circulations are linked and carry lymph toward the heart.

Small superficial lymph vessels called lymph capillaries lie very close to the blood capillaries in the interstitial space. These blind end capillaries are just one cell thick (endothelial cells). Large gaps in the walls allow excess interstitial fluid, proteins and particulate matter such as bacteria to enter the lymphatic capillaries. Elastic fibers known as anchoring filaments connect the lymph capillary endothelial cells to the surrounding connective tissue fibers. The anchoring filaments help open and close the gaps in the lymph capillary wall.

These lymphatic capillaries join with others and flow into larger vessels called pre-collectors and collectors. These vessels have smooth muscle in their walls. They also have one way valves that prevent the back flow of lymph.

During lymphatic circulation lymph flows through filtration and collection points called lymph nodes. The body has on average 600 of these bean-shaped nodules which have a fibrous outer capsule and an internal collection of immunologically active



cells. The nodes filter and destroy foreign substances such as bacteria and toxins that may be present.

The lymphatic system has no single pump like the heart. It depends on the contraction of smooth muscle in the walls of the larger lymph vessels.

The superficial lymphatic system is also divided into regions determined by the area of skin drained by a single network of lymphatic vessels (or lymphatic plexus). The exact anatomy of lymphatic regions is still being explored. It is thought that between these regions are areas which have few lymphatic vessels. These areas are described as the lymphatic watershed areas. Connections exist between watersheds and if drainage is poor in one watershed, lymph flow can be diverted to adjacent ones.

The lymph vessels ultimately open into large ducts known as the thoracic duct and right lymphatic duct which in turn open into the neck veins. The right lymphatic duct drains lymph from the right side of the head, neck and chest, and right upper limb. The lymph from the rest of the body drains into the thoracic duct. This way, the excess fluid that would otherwise collect in the interstitial space is taken back into the circulation.

Oedema is an indication that normal capillary-lymph exchange has been interrupted. This may happen if there is an obstruction of the lymphatic system. Parasites, bacterial infection, cancer or fibrotic tissue growth caused by therapeutic radiation can block the flow of lymph through the system. Lymph drainage can also be impaired if lymph nodes are removed during surgery.

What role does the lymphatic system play in the immune system?

The lymphatic system plays an essential role in the immune functions of the body. It is often the first line of defence against invading microorganisms such as bacteria or viruses. Thus when skin is damaged, microorganisms or their toxic products gain access to the tissue spaces. From there they enter the terminal lymphatics and are carried with the lymph to the first lymph node in the chain. Their foreign nature (antigenicity) is recognized by cells called macrophages which then "present" the antigen to lymphocytes which are found in large numbers in the lymph node. This results in the lymphocytes being activated. The activated lymphocytes divide repeatedly to produce many daughter cells which then attack the invading organism either by the secretion of antibodies (humoral immunity, mediated by B-lymphocytes) or by directly binding to the foreign cell (cell mediated immunity, mediated by T-lymphocytes). The lymphatic system is a circulation and as such there is constant movement of fluid and cells (mainly lymphocytes) from the blood stream through the tissue spaces through the lymph vessels and nodes and back to the blood stream. This is the basis of the immunosurveillance function of the lymphatic system. That is, the body is constantly being monitored for the presence of any cell or substance that is recognised as foreign. These could be microorganisms or toxins, as mentioned above, or body cells that have mutated and become cancerous. It is evident therefore that



when the lymphatic system is impaired, as in lymphoedema, a person is much more susceptible to infection.

How does the lymphatic system help with digestion?

The lymphatic system absorbs fats and fat soluble vitamins from the digestive system and transports them to the venous circulation.

The mucosa that lines the small intestine is covered with fingerlike projections called villi. There are blood capillaries and special lymph capillaries, called lacteals, in the centre of each villus. The blood capillaries absorb most nutrients, but the fats and fat-soluble vitamins are absorbed by the lacteals. The lymph in the lacteals has a milky appearance due to its high fat content and is called chyle.

5. Incidence and Prevalence of Lymphoedema

5.1 Defining Incidence and Prevalence

Epidemiology is a term used to describe the study of the causes, distribution and control of disease in populations. In order to better understand the burden of any particular disease or condition on a specific population, it is vital to have accurate information on how many people are currently suffering from the condition (prevalence) and how many new cases develop over a defined timeframe, usually a year (incidence). This is important as it allows us to compare different populations over time and helps greatly in planning health services.

One of the main difficulties in using this approach to study lymphoedema is that there is currently no agreed definition of lymphoedema and therefore we are not sure exactly how many people suffer from this condition here in Northern Ireland. This is further compounded by the fact that lymphoedema is not always recognised, or on some occasions is misdiagnosed due to similarities with other conditions. In evaluating the epidemiology of a condition, it is essential to describe exactly how the disease is defined. An exact and valid definition allows for comparisons between populations.

This difficulty is evident when international research regarding the incidence and prevalence of lymphoedema is taken into account. Only a relatively small number of studies have considered the level of lymphoedema in the population and all highlight the limitations of their literature reviews. Of those studies which have been undertaken, different definitions of lymphoedema and indeed different measurement tools have been used. As a result, there is no strong body of evidence which relates to population incidence or prevalence of lymphoedema, therefore making it very difficult to draw any comparisons or robust conclusions.

5.2 Population Incidence – an Overview

A systematic search of the literature revealed a limited number of papers relating to population incidence and prevalence. Whilst a few have reviewed population burden,¹⁻⁴ most identify incidence within a particular sub group e.g. those who have developed the condition as a result of cancer treatment.

Only one study reported the incidence of lymphoedema in a local population (SW London). This study found a population prevalence of 1.33 per 1000 for all ages, rising to 5.4 per 1000 in the over 65 age group.⁵ The authors also highlighted that the study was likely to underestimate the true burden of the condition. Using these rates, this would equate to 2274 people in Northern Ireland suffering from lymphoedema, with 1258 people over 65 having the condition.*

*NI Population estimate (NISRA) 2005.

Foot note: In developing the guidelines it was recognised that there are many causes of lymphoedema and incidence will vary accordingly around the world depending on the cause. This review did not include literature relating to tropical countries where lymphoedema is endemic in some areas.

In analysing a subsection of those who were clinically identified as having lymphoedema, 25% had developed the condition as a result of cancer treatment. It was interesting to note that 47% developed symptoms within one year with only 3% developing symptoms 10 years after cancer therapy.

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5.3 Incidence and Prevalence of Lymphoedema by Aetiology

This section reviews evidence in relation to how individual causes of lymphoedema contribute to the overall burden of the condition. Whilst the limitations of the research are discussed, a constant theme throughout the literature is evident in that lymphoedema is a very much underestimated and under diagnosed condition.

In considering the evidence, as would be expected, there is great variation in the type of research undertaken and indeed in the quality of research methodology used. In addition a number of limitations in undertaking this review are listed as follows:

- As there is no internationally agreed case definition of lymphoedema, nor indeed within individual countries, comparisons cannot be made between studies;
- A lack of research volume contributes to the difficulty in making judgements on whether or not methodologies have been subject to bias;
- As methods for diagnosing, measuring and assessing lymphoedema vary, the opportunity for comparing and cohorting studies is limited;
- The potentially lengthy lead time to the development of lymphoedema limits the accuracy of studies with short follow up periods;
- There are few well designed studies of sufficient size and follow up to provide accurate incidence and prevalence data;

- Much of the research was undertaken more than a decade ago, when there was very limited knowledge on the condition and as such misdiagnosis is a concern. Misdiagnosis is still a current issue;
- The passage of time also impacts on the potential to compare rates as treatment options vary and surgical procedures evolve;
- Many studies have used convenience sampling in the identification of patients;
- Symptoms may be present before clinical diagnosis is possible.

5.3.1 Primary Lymphoedema

Lymphoedema is generally classified as being either primary or secondary.

Three types of primary lymphoedema are recognised – congenital (see section 6.3), praecox and tarda. Secondary lymphoedema develops as a consequence of disruption or obstruction of the lymphatic pathways.

The population prevalence of primary lymphoedema is not known. A number of papers provide estimations which vary widely. Incidence of between 1 in 10,000 and 1 in 33,000 are reported^{1,2} and 1.5 per 100,000 in the population under 20³ as well as a suggestion that up to half of all cases of lymphoedema are primary in origin.⁴ Of the few epidemiological studies which have been undertaken, the potential extrapolation of the conclusions is constrained by methodological restrictions such as convenience sampling, diagnosis and classification issues. One such study cited by Williams⁵ identified 2743 patients from surgical units with lymphoedema, 36.8% of whom had primary lymphoedema.

Within the UK, local audits and service reviews also provide more information on the potential burden of lymphoedema. Estimations of between 8% and 28% of patients referred to lymphoedema services are identified as having a form of primary lymphoedema.⁵⁻⁸

5.3.2 Secondary Lymphoedema

The main causes of secondary lymphoedema are listed in Table 5.1. Post-infectious lymphoedemas are mainly caused by filariasis in tropical areas, and by cellulitis in occidental (western) areas. Filariasis is the most prevalent worldwide cause of lymphoedema, and is particularly common in south-east Asia. For the purpose of this review, papers relating to filariasis have been excluded as this is not an issue in Northern Ireland.

The most common causes of secondary lymphoedema in western countries are trauma or tissue damage (iatrogenic), malignant disease or venous disease.

Table 5.1: 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
Malignant Disease	<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 • Filiariasis • Tuberculosis (rare)
Inflammation	<ul style="list-style-type: none"> • Rheumatoid arthritis • Psoriatic arthritis • Dermatitis/eczema • Sarcoidosis and oro-facial granulomatosis • Podoconosis (non-filarial, non-infective, usually crystalline blockage of the limb lymphatics) • Pretibial myxoedema (rare)
Immobility and Dependency	<ul style="list-style-type: none"> • Dependency oedema • Obesity • Paralysis • Sleep Apnoea
Artificial Lymphoedema	<ul style="list-style-type: none"> • Self harm

Adapted from: Lymphoedema Framework. Best practice for the management of lymphoedema, International consensus. London MEP Ltd, 2006.

Trauma and Non-Cancer Tissue Damage

The majority of papers relating to incidence of lymphoedema after trauma are related to areas such as the surgical and/or radiotherapeutic treatment of melanomas, breast or genito-urinary cancers.

Significant rates (40-55%) of lymphoedema have been reported in patients undergoing groin dissection.^{9,10} Scars¹¹ and burns¹² are also linked to lymphoedema as causal factors, however, information on burden is limited to one study which reported a 1% incidence¹³ from a retrospective study in a burns unit. Research in this area is continuing.¹⁴

Malignant Disease

Breast Cancer

Research on the links between lymphoedema and breast cancer is perhaps the most complete and the topic of a number of reviews, many of which highlight a lack of robust evidence on incidence.¹⁵⁻²⁶

The development of lymphoedema as a consequence of breast cancer management or disease recurrence has long since been recognised. There is, however, wide variation in the reporting of incidence from 0.04% in a questionnaire survey of 1242 patients to 42.4% in patients who had been treated with a combination of surgery and radiation for breast cancer. The majority of opinion puts the incidence somewhere in the region of 12 %– 25%.²⁷⁻³⁹ A small number of papers have investigated incidence over longer time periods.⁴⁰⁻⁴²

Although a number of papers specifically investigate evolving surgical techniques such as sentinel lymph node biopsy^{43,44} and the use of radiotherapy,⁴⁵⁻⁵¹ there is little information to substantiate a population reduction in lymphoedema as a result.

Melanoma

The link between treatment of malignant melanoma and lymphoedema is well established. Earlier studies put the rates of incidence between 23% and 80%.^{5, 52-56} However, with advances in treatment options such as the introduction of sentinel lymph node (SLN) biopsy without groin dissection, the literature indicates a reduction in incidence,⁵⁷⁻⁵⁹ although there are no randomised controlled trials (RCTs) to validate these claims.

Gynaecological Cancers

The paucity of information on the incidence of lymphoedema within the gynaecological cancer survivor population is highlighted in recent reviews.^{5, 60} One study published in 2007 of 802 survivors⁶¹ identified 10% with a diagnosis of lymphoedema and a further 15% with symptomatic swelling. A similar Australian study of 468 patients found an 18% incidence.⁶²

In studies where comparisons were made,^{60,62} lymphoedema was found to be more prevalent in vulva cancer. The significant rates (up to 48%) of lymphoedema associated with treatment of vulva cancer is echoed in other papers.⁶³⁻⁶⁶

Information on cervical cancer is the most prevalent. Incidence of lymphoedema in patients undergoing hysterectomy varies from 11-50%,⁶⁷⁻⁷² although a recent retrospective American review of 1289 patients, who had a hysterectomy as part of their treatment, found lymphoedema developing only in 2.4% of patients who had lymph nodes removed.⁷³

Genitourinary Cancers

The incidence is largely unknown and varies according to type and location of tumours.⁷⁴

Literature in relation to the treatment of penile cancers indicates a wide variation in incidence from 5% to 100% (most common around 15-30%).⁷⁴⁻⁸⁰

Venous Disease

Impaired lymphatic function is identified as a common symptom of venous disease including venous ulceration.^{81, 82} A recent study on the prevalence of leg ulceration in a London population of 252,000 identified a prevalence rate of 0.45/1000 (113 patients).⁸³ 42% of these patients also had lymphoedema. Interestingly the study also highlighted that the prevalence of chronic leg ulceration is only one third of that predicted by previous studies that used similar methodologies in the 1980's.

A study of 689 leg ulcers in 555 patients identified 17 patients for whom lymphoedema was the cause of their ulcer and an additional 11 patients for which the cause was a mix of lymphoedema and venous reflux (approx 4% related to lymphoedema).⁸⁴ This study also identified a leg ulceration population prevalence of 1.5 per 1000 over 65 years.

Evidence on the incidence of other causes of venous disease such as intravenous drug use was not found, although a few papers were identified which considered this issue.^{85,86}

Infection

Chronic lymphoedema is both a risk factor for and a consequence of erysipelas/cellulitis.⁸⁷⁻⁸⁹ In western populations cellulitis is the primary cause of infection-related lymphoedema. While there is scant evidence on the incidence of cellulitis, a recent Cochrane review of the literature cited a study which identified a rate of 4 to 25 cases per 10,000 person years in the over 65 age group.⁹⁰

In a study of 176 patients admitted to hospital with cellulitis, lymphoedema was found to be a major risk factor and was present in 18% of cases.⁹¹ This finding was echoed in a retrospective study of 574 patients hospitalised over a three year period for recurrent episodes of cellulitis, with lymphoedema being highlighted as a major

risk factor.⁹² Another study suggests that cellulitis may be present in up to 50% of patients suffering from severe forms of lymphoedema as compared to 1/1000 in the general population.⁹³

Reviews of the relationship between cellulitis and lymphoedema highlight a lack of robust epidemiological data.⁹³⁻⁹⁵ One paper identifies cellulitis as a complication in 20-30% of lymphoedemas.⁹⁴ This finding is not evidenced.

In one study lymphoscintigraphy was used to demonstrate significant abnormalities in lymphatic vessels after erysipelas (77% of 30 patients with at least 2 episodes of infection). However, some of these patients may have had lymphatic abnormalities before the infection, making it difficult to separate cause and effect.⁹⁶

Inflammation

There is no evidence detailing how prevalent lymphoedema is among patients suffering from inflammatory conditions. A number of case reports and series identify lymphoedema as a rare complication of dermatitis,^{97,98} and arthritis in particular.⁹⁹⁻¹⁰⁴

Immobility and Obesity

There is no evidence to support the level of risk associated with immobility and developing lymphoedema. Obesity in particular is well documented as playing a causal role in the development of lymphoedema and as a poor prognostic factor in response to lymphoedema treatment.¹⁰⁵⁻¹¹³

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6. Diagnosis, Assessment and Classification of Lymphoedema

Lymphoedema is an incurable chronic condition, which is progressive if left untreated. Early diagnosis and treatment are therefore essential to prevent physical and psychological morbidity. It is important to remember that treatment is still effective at later stages, although a longer period of treatment may be required to reduce swelling and breakdown fibrosis.

This section outlines how lymphoedema is classified and makes recommendations on how patients should be diagnosed and assessed. Forms outlined in the appendices will also be available to download from the Northern Ireland Lymphoedema Network (LNNI) website.

6.1 Definition of Lymphoedema

Lymphoedema is an 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.¹

It is associated with feelings of discomfort and heaviness, functional limitations, disfigurement, psychological distress and an elevated risk of recurrent infections².

It is important to identify patients with lymphoedema so that treatment strategies can be started at an early stage to prevent deterioration. If a patient presents with chronic swelling clinical history and examination are the key stones to establishing a diagnosis. Radiological investigations may also be necessary to establish a diagnosis and to exclude malignancy.

6.2 Lymphoedema Staging

Lymphoedema, whether primary or secondary, develops in a number of stages, from latent to late stage severe disease. (Table 6.1)

Table 6.1 Stages of Lymphoedema


<p>Stage 0/ Latent</p>	<ul style="list-style-type: none"> • The lymphatic vessels have sustained some damage which is not yet apparent. Transport capacity * is reduced but is still sufficient for the amount of lymph being removed; • Subjective complaints from patient possible; • No clinical evidence of lymphoedema, however a slower lymph flow is detected by lymphoscintigraphy with initial dermal backflow; • Lymphoedema is not present but patient is classified as being at risk from developing lymphoedema. 	<p>* Transport capacity of the lymphatic system refers to the amount of lymph which is transported by the lymphatic system utilising its maximum amplitude and frequency.</p> <p>** Stemmer's Sign (Fig 6.1) is positive when a thickened 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. The absence of a Stemmer sign does not rule out the possibility of lymphoedema.⁴</p> <p>Figure 6.1 – Stemmer's sign</p>  <p>© Blackwell Publishing Co Oxon</p>
<p>Stage 1</p>	<ul style="list-style-type: none"> • Oedema is present; • Tissues are pitting, soft, and doughy; • Swelling reduces with elevation; • Little or no tissue fibrosis, negative Stemmer's sign**. 	
<p>Stage 11</p>	<ul style="list-style-type: none"> • Oedema is present; • No reduction of swelling on elevation; • Connective tissue proliferation/fibrosis; • Pitting becomes more difficult; • Positive Stemmer's sign. 	
<p>Stage 111 Lymphostatic Elephantiasis</p>	<ul style="list-style-type: none"> • Fibrosis and sclerosis (severe induration); • Non pitting; • Positive Stemmer's sign; • Hyperkeratosis; • Lymphangiomas; • Papillomas; • Fungal Infections. 	

Table 1. based on the German Society of Lymphology,³

6.3 Classification of Lymphoedema


Lymphoedema is classified as being either primary or secondary.

6.3.1 Primary Lymphoedema

Primary lymphoedema may develop as a sporadic condition, or as part of a complex syndrome (Table 6.2). Increasingly the genetic basis for inherited forms of lymphoedema is being identified and this may lead at some time in the future to targeted therapies for affected families.

The lymphatic system develops from eight lymph sacs from the venous side of the circulation.⁵ This process requires the expression of haemopoietic signalling proteins and vascular endothelial factors, in particular vascular endothelial growth factor C (VEGF-C).⁶⁻⁹

Abnormalities in genetic expression for the genes which modulate lymphatic vascular formation, lead to developmental defects of lymphatic vasculature, causing hereditary primary lymphoedema. Familial occurrence of lymphoedema is well recognised,^{10 11} with the most common being Milroy's Disease and Meige's syndrome (Box 6.1), which display autosomal dominant inheritance patterns and a demonstrated mutation in the kinase domain of the vascular endothelial growth factor gene (VEGFR-3). Abnormalities in the FOXC2 gene mutation are responsible for Lymphoedema Distichiasis syndrome.^{8,9}

 As the genetic forms of lymphoedema are increasingly recognised, a careful family history of lymphoedema should be established during assessment. Referral to genetic screening and counselling should be made for appropriate patients and families.

Box 6.1

Milroy's Disease and Meige's Syndrome

A congenital abnormality of lymphatic vasculature.

- Aplasia
- Hypoplasia
- Hyperplasia
- Congenital valvular incompetence

Occurrence :

87% Female; 13% Male (It may be inherited or sporadic);

- Praecox occurs around 17years of age (83%);
- Tarda occurs around 35 years (17%);
- Milroy's starts in the first few years after birth;
- Meige's starts around puberty.


Table 6.2: Syndromic Classification of Lymphoedema

Syndrome name	Age of onset	Clinical features of lymphoedema	Other clinical features	Inheritance/ gene type	How common?
Milroy's (Noone-Milroy) Disease	Birth/ Childhood	Peripheral	None	Autosomal Dom VEGFR3	1 : 33,000 live births
Lymphoedema Distichiasis	Puberty	Lower limb	Double row of eye lashes Corneal abrasions	Autosomal Dominant FOXC2	Very Rare
Meige's Syndrome	Puberty	Peripheral Lymphoedema	None	Autosomal Dominant	Commonest Inherited lymphoedema
Noonan Syndrome	Birth	Lower limbs, feet, genital, cystic hygroma Intestinal lymphangectasia	Craniofacial, Musculoskeletal, Cardiovascular, Endocrine/ sex characteristics, Neurological abnormalities	Autosomal Dominant With variable expression or sporadic	1 : 1000-2500 births
Turner's Syndrome	Birth Childhood	Dorsum of hands & feet. May resolve in adulthood	Craniofacial, Musculoskeletal, Cardiovascular, Endocrine/ sex characteristics,	Single x Sporadic	1 : 2,500 female births
Klinefelters Syndrome	Birth Childhood	Peripheral Lymphoedema	Tall stature, Sex characteristics Neuropsychiatric manifestations	XXY Sporadic	1 : 500-700 male births
Klippel- Trenaunay	Childhood Puberty	Lower limb lymphoedema	Port wine stain Limb hypertrophy, varicose veins	Sporadic	1:20,000-40,000 births
Trisomy 21	Birth Childhood	Peripheral lymphoedema Nucal facial	Down's Syndrome	Trisomy 21	1:650-1000 births
Intestinal Lymphangectasia	Childhood Young adult	Usually unilateral Lower limb	Malabsorption of fat and protein		Very rare

6.3.2 Secondary Lymphoedema

Secondary lymphoedema is caused by damage to the lymph nodes and vessels. The potential causes are listed in Section 5, Table 5.1

It should be noted that cancer related secondary lymphoedema can occur immediately after surgery or some years later.

 Patients at known risk of developing lymphoedema as a result of cancer treatment should be informed of this risk and should be referred to lymphoedema treatment services for assessment if they complain of swelling, tightness or difficulty getting clothes or jewellery to fit in the affected limb.

Pathophysiology

Oedema, whatever the underlying cause, is due to an imbalance between capillary filtration and lymph drainage. Most examples of limb oedema are caused by an increase in capillary filtration which overwhelms lymph drainage capacity e.g. heart failure, nephrotic syndrome. The swelling from lymphoedema results from a failure of lymph drainage in circumstances when capillary filtration is not increased.¹²



Figure 6.2 Dependency Oedema
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6.4 Clinical Features of Lymphoedema

- Recurrent cellulitis
- Pitting or non-pitting oedema (refer to staging - section 6.2)
- Skin changes
- Fibrosis/hyperkeratosis
- Thickened skin folds
- Misshapen limb

Symptoms (self reporting /on assessment)

- Swelling - inability to wear rings and watches , difficulty fitting into clothes/shoes
- Tingling, pins and needles (paraesthesia)
- Skin changes – skin tight and shiny, feels taut
- Reduced range of movement- in affected limb joints
- Functional restriction
- Heaviness
- Pain caused by:
 - Skin tightness;
 - Reduced function;
 - Muscle strain, weight pulling on soft tissue structures, e.g. tendons, ligaments;
 - Nerve compression;
 - Inflammation or infection.

Skin Changes

- Hyperkeratosis (thickened brown pigmentation).
- Lymphangiomata (Lymph blisters).
- Pappilomatosis (wartlike growths on the skin due to fibrosis over dilated lymphatics) (Fig 6.3).
- Fungal infections.
- Lymphorrhoea (leakage of lymph fluid through the skin surface) (Fig 6.4).



Figure 6.3 Papillomatosis
© Blackwell Publishing Co.Oxon




Figure 6.4 Lymphorrhoea


6.5 Complications of lymphoedema which may present at initial diagnosis and which may require prompt treatment/management

1. **Cellulitis** - this is a common infection in lymphoedema and untreated lymphoedema may masquerade as cellulitis.¹³
2. **Vein Thrombosis** - detected by venogram, or ultrasound. May be associated with sudden swelling or an increase in swelling, and may be painful. It can occur in upper or lower limb.
3. **Fungating Tumour** - e.g. chest wall post breast cancer. May co-exist with lymphoedema; referral to tissue viability specialists and oncology is recommended. Treatment of the lymphoedema is palliative in this context.

6.6 Clinical Diagnosis and Assessment of Lymphoedema

In order to treat patients with lymphoedema, a holistic approach needs to be adopted. It is suggested that any physical, emotional or psychosocial problems a patient is experiencing should be recorded, as these factors may precipitate or perpetuate quality of life difficulties associated with lymphoedema. Such factors need to be addressed routinely with recording of results in patient notes to identify changes in emotional well-being over time.

 On the basis of a complete assessment, patients should be referred for further psychology, dietetic or vascular assessment, or to social services or occupational therapy services as necessary.

 A complete physical and psychological assessment of all patients diagnosed with lymphoedema should be undertaken using appropriate visual analogue scales for pain and patient's impression of severity of lymphoedema. A detailed assessment form is outlined in Appendix 6.

An accurate diagnosis of lymphoedema is essential for appropriate therapy. It is important that patients referred to lymphoedema treatment services should have had other causes of oedema e.g. heart failure or malignancy excluded prior to referral.

6.6.1 Assessment Requirements:

a) Medical History

This should exclude a history of cardiac failure and sleep apnoea and any co-morbidity which would modify or be a contraindication of treatment (refer to section 7.3). Patients with lower limb lymphoedema should be screened for risk of arterial disease, especially those who have a history of:

- Cerebral vascular accident;
- Transient Ischaemic attack (TIA);
- Myocardial infarction;
- Angina;
- Diabetes;
- Rheumatoid arthritis;
- Intermittent claudication;
- Night pain which penetrates big toe;
- Nerve damage – peripheral neuropathy;
- Operations – especially surgical procedures to vascular system or abdominal area;
- Smoker.

Patients with lymphoedema at risk of arterial vascular disease should be referred to vascular surgeons for assessment if there are concerns about applying compression. Patients with lymphoedema who are referred from vascular clinics should have had an assessment of their arterial vascular system prior to referral. This may be assessed using ankle brachial pressure index (ABPI) using a range of different sized sphygmomanometer cuffs and Doppler probes. It is recognised that ABPI readings may be difficult to ascertain in a grossly swollen limb. In some situations where it is not possible to perform ABPI due to severe oedema, toe pressures or oxygen saturation using toe pulse oximetry, may give some indication of arterial vasculature. Toe brachial pressure index (TBPI) should only be performed when ankle brachial pressure index (ABPI) is not possible. It is calculated for each leg: (TBPI = highest toe systolic reading for that leg/highest brachial systolic reading). There were no studies found on the use of ABPI, TBPI or oximetry in the literature review for assessment of the arterial system in patients with lymphoedema.

Physical Examination for Evaluating a Swollen Limb:¹⁴

- Detectable enlargement of the limb or trunk;
- Number of skin folds at the axilla, along the limb, digits;
- Skin colour (e.g. erythema; brownish pigmentation);
- Skin temperature;
- Skin texture (soft, hardened, shiny, taut, fibrosis,ulceration);
- Asymmetric increase in subcutaneous adipose tissue;
- Presence of lower limb hair;
- Stage of oedema - 0- III;
- Pulses may be difficult to palpate in a grossly swollen limb;
- Range of movement;
- Neurological deficits;
- Venous collaterals and/or congestion;
- Measurement of limb volume.

b) Family & Social History


 Those patients who have a family history of lymphoedema should be referred to a genetics service.

c) Psychological Assessment


As well as assessing patients' physical problems, it is also essential to address any emotional or psychosocial problems, which they may experience as a result of their condition. Those who demonstrate emotional difficulties should be referred to specialist psychological services. To measure how emotional difficulties exert an impact on daily life a number of quality of life (QOL) questionnaires have been developed, the most common of which are outlined in Appendix 4.

Overall conceptual and methodological rigour is lacking when looking at assessment tools in the literature. There is a need for an encompassing, unequivocal tool to assess quality of life / psychological issues in individuals with lymphoedema for all areas of the body.

There are no specific quality of life measures for patients with lymphoedema. The two most commonly used questionnaires on this target population are SF-36 and FACT-B. SF-36™ is a general health measure which numerous studies have found useful in assessing QOL in patients with cancer. FACT-B focuses specifically on QOL in breast cancer patients, but is not primarily aimed at those who suffer from lymphoedema. Both questionnaires are short, yet have been found to be sensitive and reliable.

 The SF36v2™ should be used to assess QOL in the absence of an unequivocal lymphoedema quality of life tool to assess the psychological impact. This tool should only be used by practitioners who have been trained in its use and who are aware of trigger points for referral to psychological services. A copy of the SF36v2™ is provided in Appendix 5.


Links with Other Multidisciplinary Groups

 Multidisciplinary meetings should be held on a regular basis to discuss complex patients and to review issues such as skills training, audit, recent research and service development. Please refer to section 7.7 for further information on the roles of multidisciplinary team members.


6.6.2 Establishing a Diagnosis of Lymphoedema

In many instances the diagnosis of lymphoedema can be made on taking the medical history and by clinical examination, including measurement of limb volume. This is mainly relevant to lymphoedema affecting the limbs where measurement can be attempted. It is not applicable to oedema of head, neck, trunk or genital area. Photography, with patient consent, may be of use in monitoring response to treatment. Measurement of lymphoedema can be carried out using the following methods:

- (1) **Water displacement method.** This method is considered the gold standard for calculating limb volume, particularly for hands and feet. It is based on the principle that an object displaces its own volume of water, however issues of hygiene and the practicalities of water emersion limit its use.^{15 16}
- (2) **Bioimpedance.** (see Glossary) Bioimpedance measures tissue resistance to an electrical current, which estimates extra cellular fluid volume. Studies have shown that this is a sensitive method for diagnosing lymphoedema and in monitoring change when a patient is being treated.¹⁷⁻²²
- (3) **Perometry.** This uses an infrared optoelectronic system to measure limb volume. It can measure of limb volume quickly and accurately and compares well with Water Displacement Methods²³.
- (4) **Circumferential volume measurement.** Calculation of volume from circumferential measurements is the most widely used method of calculating limb volume. It has been shown to correlate with water displacement¹⁵

 Measurements are calculated from fixed anatomical points along the limb with repeated 4 cm measurements producing more accurate volume calculations than 8 cm repeat measurements.¹⁶

Circumferential measurements have been compared to other methods,^{15, 24, 25} and though it is apparent that while measurements are not Interchangeable, there is a good correlation between the different methods used.

 Circumferential measurements of limbs are put into a specialist calculator or computer programme for calculation of limb volume, which can be used to aid diagnosis and monitor response to treatment.

6.6.3 Which method should be used to calculate limb volume?

Ideally the method used to calculate limb volume should be easy to use, accessible, non-invasive and inexpensive. It should have a high sensitivity and specificity. (Table 6.3.) Circumferential limb volume measurement is the most widely used method of assessing limb volume. It is easily accessible, though training is required to ensure reliability.


 Multiple frequency bioimpedance measurement has advantages over measurement of limb circumference in that it is applicable to bilateral limb lymphoedema. Although, it is not generally available in Northern Ireland it is looking to be a promising, effective and efficient way of assessing and monitoring lymphoedema and should be considered for use in the longer term (over the next 5 years).

Table 6.3: Comparisons of Methods to Calculate Limb Volume


Method	Applicable	Non-Invasive	Cost	Readily available In N.I.	New equipment required	New training required	Specificity	Sensitivity
Water Displacement (unilateral limb)	<ul style="list-style-type: none"> • Unilateral Limb • Hands and Feet 	Yes	Minimal	No	Yes	Yes	Mod.	High
Bioimpedance	<ul style="list-style-type: none"> • Unilateral Limb • Bilateral Limb 	Yes	£2K - upper limb £10K - upper & lower limb	No	Yes	Yes	High	High
Circumferential Measurement	<ul style="list-style-type: none"> • Unilateral Limb: base line • Bilateral: response to Rx 	Yes	Minimal	Yes	No	Ongoing training of new practitioners	Mod.	35 – 91%
Perometry	<ul style="list-style-type: none"> • Unilateral Limb • Bilateral Limb 	Yes	Not known	No	Yes	Yes	High	Mod

6.6.4 How much difference in volume between limbs is lymphoedema?


Limb volume measurement is used to determine if lymphoedema is present in a limb. Calculation of limb volume measurement has limitations. This may be due to a pre-existing difference in volume related to the dominant limb or to increased muscle bulk or fat rather than to fluid volume.^{17,28} Debate exist as to how much of an increase in limb volume constitutes a diagnosis of lymphoedema.

A study comparing different definitions of lymphoedema in post breast cancer patients used 4 diagnostic criteria.²⁶ Limb volumes were measured and different measurement criteria for the diagnosis of lymphoedema (200 ml limb volume increase detected by perometry, 10% arm volume increase on perometry, 2 cm circumferential increase or a report of heaviness or swelling either now or in the past year) were compared to the clinical detection of lymphoedema. In this study the 10% limb volume change corresponded to a conservative definition, whereas the 2 cm difference between limbs or from a known pre-treatment baseline constituted a more liberal definition. In a further study,²⁵ a 10% volume increase calculated using circumferential limb measurement was shown to have a sensitivity rate of 49%, whereas a 5% increase in circumference was shown to have a 91% sensitivity rate. Other studies have shown that using a circumferential difference of greater than 5 cm between limbs or from a known pre-treatment baseline give a low pick up rate of lymphoedema²⁷.

It was the consensus of the CREST Lymphoedema group that a 5% increase in circumferential measurement should trigger a reference to lymphoedema treatment services.

 It is recommended that limbs are measured prior to surgery/radiotherapy, both of which may increase risk of developing lymphoedema. Both limbs should be measured and changes from the baseline or between limbs may be used as a measure of lymphoedema.

6.7 Referral criteria for people who are known to be at risk of developing lymphoedema

 It was the consensus of the CREST lymphoedema group that patients who have a history of previous trauma to the lymphatic system e.g. previous cancer surgery, radiotherapy or recurrent cellulitis, should be referred to lymphoedema treatment services for assessment when:

- Visible swelling is evident;

OR

- Where there is a 5% increase in circumference at any site in uni-lateral lymphoedema compared to the other limb or compared to a pre-treatment measurement;²⁵

OR

- Where the patient complains of symptoms, which would suggest lymphoedema such as tightness of clothing or jewellery or heaviness in the limb.^{29 30 27 31}

6.8 Differential Diagnosis of Lymphoedema (Table 6.4)

Table 6.4 lists a comparison of symptoms of oedema which could be used as an aid for differential diagnosis e.g. chronic venous insufficiency (CVI) and lipoedema.

Oedema secondary to medical problems e.g. cardiac failure is usually bilateral, symmetrical and markedly pitting.³²

Lipoedema can be described as a bilateral, symmetrical, flabby swelling that arises from the deposition of adipose tissue. The cause of Lipoedema is not fully understood.

Lipolymphoedema is a combination of lipoedema, obesity and lymphoedema. Lipolymphoedema can also develop in combination with dependency oedema, chronic venous insufficiency, and other vascular diseases.

Chronic venous insufficiency (CVI) is a condition in which the veins of the legs do not efficiently return blood to the heart, commonly caused by damage to the veins. CVI diminishes the capacity of the venous system, thereby increasing the workload for the lymphatic system in the affected area.

Table 6.4: Differential Diagnosis of Lymphoedema


Lymphoedema	CVI	Lipoedema	Lipolymphoedema
Male and Female.	Male and female.	Common in women.	Common in women.
Age of onset is any age.	Age of onset – older person. May occur in combination.	Age of onset – 60-70 % around puberty.	Can be a complication of lipoedema.
Family History 20%.	Family history.	Family history 16-50%.	
Stemmer's generally positive.	Stemmer's sign negative.	Stemmer's sign negative.	Stemmer's sign positive when lymphoedema complicates lipoedema.
Patients complain of tingling or tightness more than pain.	Pain, particularly after walking or standing is a major characteristic of CVI.	Patients complain of pain, especially present along the shin, on palpation. It is rarely severe but can cause severe tenderness (50%).	Pain is often present.
The swelling of lymphoedema can occur in the entire limb, trunk, head & neck, genitalia. It often occurs in one limb and is not symmetrical. It usually affects the entire limb including the hands and feet. Pitting occurs in early stages.	The swelling of CVI usually occurs in the ankles and lower legs.	The swelling of lipoedema tends to occur in a bilateral, symmetrical pattern. In the upper limb from shoulder to wrist, in the leg from hips to ankles causing a flap of skin over feet. Hands and feet not involved. Pitting oedema of the legs occurs especially during the second half of the day and during hot weather.	The swelling seems to follow the pattern of lymphoedema. It is asymmetric in presentation. Swelling noticeable in dorsum of feet. Pitting oedema occurs in early stages, i.e. Stage I as with lymphoedema.


Table 6.4: Differential Diagnosis of Lymphoedema (continued)

Lymphoedema	CVI	Lipoedema	Lipolymphoedema
The swelling of lymphoedema is present at all times, does not reduce at night. In early stages may reduce with elevation.	The swelling of CVI increases during the day and is reduced at night during sleep. As the CVI becomes more severe the swelling continues to increase and does not resolve at night.	The swelling of lipoedema develops in the legs during the second half of the day and decreases during sleep.	The swelling follows the pattern of lymphoedema.
Protein rich fluid causes the swelling of lymphoedema.	Protein-poor fluid causes the swelling of CVI. This fluid does not increase the risk of infection within the tissues.	Early stage - nodules develop Late stage - overhanging fat folds develop.	
Skin changes occur in the later stages of untreated lymphoedema and include fibro-papillomatosis, hyperkeratosis and cracks in the deeper folds of skin; however colour of the skin does not change.	Skin changes occur as the CVI progresses and the tissues in the affected areas become thin and shiny. In the early stages the skin colour changes to blue-purple (like new bruising). It may be associated with eczema.	Lipoedema is prone to bruising and subcutaneous bleeding and often appears after mild trauma. Normal skin consistency.	Often have deepened skin folds.
Infections related to lymphoedema often have sudden onset and include cellulitis. Increasingly serious infections, including open wounds and skin cracks may develop in untreated stage 3 lymphoedema.	Chronic leg ulcers develop as CVI progresses. These are difficult to heal and may become infected.	Prone to fungal infections due to presence of large fold of overhanging skin.	Possible risk of infections including cellulitis.

6.9 Radiological Investigations

The role of radiological investigations in confirming the diagnosis of lymphoedema has yet to be agreed, with many experts stating that lymphoedema can confidently be diagnosed from the clinical history and clinical findings.

 Lymphoscintigraphy, MR, CT and ultrasound can be used in diagnosing lymphoedema or to differentiate it from lipoedema, DVT or to detect recurrence of malignancy.

 Lymphoscintigraphy is beneficial in limb swelling where the diagnosis is unclear. However there is no standard protocol with international differences in colloid used, injection site (dermal or subcutaneous) and exercise protocol. Debate exists as to whether lymphoscintigraphy should be quantitative or qualitative and if epifascial as well as subfascial lymphatics should be imaged.³³⁻⁴⁰

Many studies of lymphoscintigraphy are in the area of breast cancer related lymphoedema (BCRL) where it can detect early lymphoedema with 73% sensitivity and 100% specificity however in some patients with lymphoedema lymphoscintigraphy is negative.³⁴

CT scans may be used in the diagnosis of lymphoedema to rule out underlying malignancy, and to differentiate from lipoedema.^{41,42} It has been found to have 93% sensitivity and 100% specificity for lymphoedema.⁴¹ A uniform protocol for performing and reporting lymphoscintigraphy in lymphoedema should be developed.

MRI scans can show thickening of the dermis in lymphoedema. The subcutaneous tissue may show a honeycomb pattern or a reticular pattern if oedema is marked.⁴³⁻⁴⁵ MRI scans of lower limb lymphoedema have been carried out using Gadodiamide as a contrast agent given intradermally to facilitate visualisation of the lymphatic pathways.⁴⁶

Ultrasound scans, particularly high resolution doppler, can help to differentiate lymphoedema from lipoedema and may also be helpful in the detection of lymphoedema of the head and neck.^{44,47}

Key recommendations: Diagnosis and Assessment

- Patients at known risk of developing lymphoedema as a result of cancer treatment should be informed of this risk and should be referred to lymphoedema treatment services for assessment if they complain of swelling, tightness or difficulty getting shoes/clothes or jewellery to fit in the affected limb.
- A complete physical and psychological assessment of all patients diagnosed with lymphoedema should be undertaken using appropriate visual analogue scales for pain and patient's impression of severity of lymphoedema.
- The SF36v2™ tool should be used to assess quality of life and psychological impact. This tool should only be used by practitioners who have been trained in its use and who are aware of trigger points for referral to psychological services.
- On the basis of a complete assessment, patients should be referred for further psychology, dietetic or vascular assessment, or to social services or occupational therapy services as necessary.
- Patients who have a family history of lymphoedema should be referred for genetic counselling.
- Patients who have a history of previous trauma to the lymphatic system e.g. previous cancer surgery, radiotherapy or recurrent cellulitis, should be referred to lymphoedema treatment services for assessment when:
 - Visible swelling is evident;
 - OR**
 - Where there is a 5% increase in circumference at any site in uni-lateral lymphoedema compared to the other limb or compared to a pre-treatment measurement;
 - OR**
 - Where the patient complains of symptoms, which would suggest lymphoedema such as tightness of clothing or jewellery or heaviness in the limb.
- Limb volume measurements should be calculated from circumferential measurements of the limb from fixed anatomical points using 4 cm repeated measurements. These measurements should be used to calculate individual limb volumes.
- All practitioners assessing patients or treating patients with lymphoedema should have access to a limb volume computer programme or calculator.
- Multiple frequency bioimpedance measurement has advantages over measurement of limb circumference in that it is applicable to bilateral limb lymphoedema (as the limb can be used as its own control).

- Limb volume measurements should be made as a baseline prior to treatment (e.g. surgery or radiotherapy), which is likely to cause lymphoedema.
- The diagnosis of lymphoedema is often confirmed on the basis of history and clinical examination. Radiological investigations such as CT, MRI, or ultrasound should be carried out where malignant lymphoedema is suspected. They may also have a role in differentiating lipoedema from lymphoedema in complex cases.
- Lymphoscintigraphy has a role in establishing the diagnosis of lymphoedema where clinical history and clinical examination are inconclusive. It is important to be certain of the diagnosis of lymphoedema before embarking on a life long programme of treatment. A uniform protocol for performing and reporting lymphoscintigraphy in lymphoedema should be developed for Northern Ireland.
- Multidisciplinary meetings should be held on a regular basis to discuss complex patients and to review issues such as skills training, audit, recent research and service development.


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7 Management of Lymphoedema


 All patients with a diagnosis of lymphoedema should have access to effective and efficient management of the condition, with the aims of management being to:

- (a) restore maximum functional independence;
- (b) reduce risk of infection;
- (c) provide long-term control of limb volume;
- (d) improve limb shape;
- (e) maximise lymph drainage in affected areas and minimise fibrosis;
- (f) restore maximum musculoskeletal function and correct postural imbalance;
- (g) provide psychological support;
- (h) educate patients in understanding their condition and rationale for treatment;
- (i) promote self-management.

 It should be noted that a thorough assessment of every patients' physical and psychological needs should precede any treatment. Patients should be referred to appropriate members of the wider multidisciplinary team as necessary.

7.1 Complex Decongestive Therapy


 Complex Decongestive Therapy (CDT)* is the recognised conservative management of lymphoedema. It consists of two phases categorised as intensive (decongestive) and maintenance (conservation). The initial phase is an intensive therapist led period of daily treatment (five days per week) for up to six weeks. The second phase of treatment, maintenance, encourages the transfer of care from professional to patient/carers and continues for the rest of the patient's life including regular check ups or further intensive treatment (and issuing of new containment garments). Initially six-monthly reviews should be therapist led. However, telephone reviews should be considered if the patient's condition is considered to be stable and the patient competent in self-monitoring.


 As a professional requirement to treat lymphoedema, training must be undertaken at a recognised school of lymphoedema management.


CDT is a set of four therapeutic measures consisting of two phases of management:

Phase 1 - Intensive	Phase 2 - Maintenance
<ul style="list-style-type: none"> • Manual lymphatic drainage (MLD) • Multilayer lymphoedema bandaging (MLLB) • Skin care • Remedial exercises 	<ul style="list-style-type: none"> • Compression garments • Skin care • Regular exercise • MLD (as appropriate) • Simple lymph drainage (as appropriate) • Nocturnal bandaging (as appropriate)

*Complex decongestive therapy (CDT) is also known as Decongestive Lymphatic Therapy (DLT) and Complex Physical Therapy (CPT)

 In order to achieve a significant therapeutic result, the components must be used in combination rather than in isolation. If performed in an adequate manner, CDT is an effective treatment for lymphoedema.

 CDT may need to be modified in the presence of complex co-morbidities or due to patient choice. Advanced localised cancer, for example, can result in additional damage to the superficial lymphatics further reducing interstitial drainage. As a result, palliative treatments may require modification, and outcomes may be reduced or difficult to maintain. Clinical judgement may also recommend modified CDT in other circumstances, such as for elderly patients living alone and not able to manage daily visits or intensive treatment. The reasons for modifying the treatment should be clearly stated on the patient's treatment plan.

 It is essential that treatment options are discussed with the patient (and potentially carers) and an individualised patient-centred protocol agreed. If accepted by the patient and deemed appropriate by the therapist, the patient should undergo a period of CDT. It may however not be clinically appropriate or acceptable to the patient to participate in the full programme.

The literature is conflicting in some areas regarding the effectiveness of the component parts of CDT, however many trials confirm the superior benefit of CDT¹⁻³ compared to other suggested forms of management such as surgery or pneumatic pumps.

Components of CDT

Manual Lymphatic Drainage (MLD)

MLD is the use of specific massage techniques (based on the knowledge of lymphatic anatomy and physiology) which mobilises the skin and stimulates the lymphatic system. It facilitates lymphatic flow into the venous circulation utilizing lymph vessels and existing lymphovenous anastomoses. However, it is found to be most effective when combined with compression bandaging, skin care and exercise.

There is very little trial based supporting evidence for MLD. Some studies⁴⁻⁶ report a small change in limb volume which is attributed to MLD. However, the actual study methodology of some of the other trials may have influenced the outcomes. International expert opinion has agreed a consensus that MLD is an integral component of CDT as it is the only technique to move fluid out of a limb and away from the areas of congestion.



Multilayer Lymphoedema Bandaging (MLLB)

In the intensive phase of treatment, multilayer bandaging (using specific short-stretch bandages which exhibit low resting pressures and high working pressures) is used to provide support for underlying tissues and to act as a counterforce to voluntary muscle activity, hence preventing re-accumulation of fluid. It can also reduce areas of fibrosis and reshape the limb.^{4,5,7}

This layered approach to bandaging includes the use of:

- A skin protection layer (non-compression);
- A padding layer (may be foam or layered wool);
- Short stretch compression bandages.




In the maintenance phase, patients may need to be taught nocturnal self-bandaging to supplement compression garments. Incorrect bandaging may cause more damage.

Remedial Exercise

Remedial exercises are carried out in the intensive phase of treatment in conjunction with multilayer bandages, and in the maintenance phase with a compression garment. The aim is to enhance the efficiency of the muscle pump, hence increasing lymph circulation. Each patient is given an individually tailored exercise programme suited to their particular requirements and abilities.

The use of exercise in the management of patients with breast cancer related lymphoedema is supported.⁸ Although there is a lack of trial evidence regarding other underlying causes, the basic principles of exercise associated with lymphoedema pathology can also be related to other recognised aetiologies.

Skin Care

Good skin care is essential in the management of lymphoedema in order to maintain skin integrity and reduce risk of infection. The components of good skin care are meticulous hygiene, use of emollients, monitoring for cuts and abrasions and prompt action at the first sign of infection (please refer to Appendix 7 for the CREST Cellulitis Guidelines).  For more complex skin conditions e.g. psoriasis or eczema the patient should be referred to a local dermatology department to optimise the outcome of their






lymphoedema treatment. Similarly, patients with complex wounds/ulcers should be assessed and receive shared care from a local tissue viability service.

The use of skin care in the management of patients with breast cancer related lymphoedema is supported in the literature.⁸ There is a lack of trial evidence regarding other underlying causes; however the basic principles of skin care associated with lymphoedema pathology can also be related to other recognised aetiologies.


7.2 Compression Garments

 Towards the end of CDT each patient should be measured for an appropriate compression garment.⁹⁻¹¹ It is essential that the correct garment is provided for each individual patient. Ongoing reassessment is of paramount importance to ensure continued suitability of the garment. Incorrectly fitting garments can worsen the swelling and cause further permanent damage. The clinician must take into account:


- Limb shape and size/distribution of swelling;
- Presence of skin folds;
- Lymphoedema status (stage I-III)-texture of skin;
- Skin sensitivity;
- Overall status of underlying disease e.g. cancer, arterial disease, diabetes;
- Patient's functional ability (for donning and doffing);
- Patient's choice regarding material, colour, texture, fabric etc;
- Patient's compliance.

The garments are available with a variety of attachments including shoulder attachments, separate hand pieces and waist attachments. Specialist compression garments are also available for swelling of the torso or genitalia. Donning and doffing

aids are available for easier application of compression garments. A number of garments are now available on prescription. However, care must be taken when ordering prescription garments that they are of suitable compression for the management of lymphoedema rather than other conditions e.g. for venous ulcers. The cost of garments varies greatly depending on patient requirements. See Appendix 8. for suppliers.

 In the maintenance phase of treatment, containment of the affected limb is achieved through the daily wearing of these compression garments. Their effectiveness relies on the clinical reasoning and skills exhibited by the health care practitioner to provide the most appropriate garment in a timely manner, and the concordance agreement with each individual patient regarding long-term compliance.

Garments should only be assessed for and issued by a trained clinician.

 All patients should be provided with accurate contact details of local lymphoedema services should they need to access further advice.

7.3 Treatment Contraindications

7.3.1 Contraindications for Manual Lymphatic Drainage

	Absolute Contraindication Contraindication	Relative Contraindication
General	<ul style="list-style-type: none"> • Uncontrolled heart failure – risk of increased cardiac oedema; • Acute deep vein thrombosis (DVT) – risk of dislodging the clot; • Acute infective episode (cellulitis, erysipelas); • Superior vena cava obstruction (SVCO) – risk of increase obstruction of SVC which could potentially be fatal; • Acute renal failure. 	<ul style="list-style-type: none"> • Malignancy – risk of spread of active cancer (see section 7.6)
Neck treatment	<ul style="list-style-type: none"> • All absolute general contraindications; • Thyroid dysfunction – risk of introducing increased thyroid hormones into the circulation; • Hypersensitivity of carotid sinus – risk of acute drop in arterial blood pressure and acute slowing of heart rhythm; • Cardiac arrhythmia – risk of cardiac arrest; • Patients over the age of 60 – increased possibility of arteriosclerosis of carotid artery – risk of embolism. 	<ul style="list-style-type: none"> • Patients over the age of 60 – increased possibility of arteriosclerosis of carotid artery – risk of embolism.

	Absolute Contraindication Contraindication	Relative Contraindication
Deep abdominal treatment	<ul style="list-style-type: none"> • All general contraindications; • Pregnancy; • Menstruation; • Recent abdominal surgery; • Radiation fibrosis/colitis/cystitis; • History of DVT in pelvic veins; • Inflammatory bowel disease; • Diverticulitis; • Cirrhosis of liver; • Abdominal Aortic aneurysm; • Unexplained pain; • Ileus. 	
Face treatment	<ul style="list-style-type: none"> • All general contraindications; • Inflammatory conditions of the face. 	
Oral Cavity treatment	<ul style="list-style-type: none"> • All general contraindications; • Inflammatory conditions of the oral mucosa; (Remove dental prosthesis prior to treatment). 	

7.3.2 Contraindications for Multilayer Bandaging


	Absolute Contraindication Contraindication	Relative Contraindication
Multilayer Bandaging	<ul style="list-style-type: none"> • Cardiac odema; • Peripheral arterial disease (ABPI <0.5); • Acute infection. 	<ul style="list-style-type: none"> • Arterial high blood pressure; • Cardia arrhythmia; • Scleroderma ; • Chronic polyarthritis; • Sudeck’s atrophy; • Malignant lymphoedema; • ABPI 0.6-0.8 – specialist consideration.

7.4 Surgical Management of Lymphoedema

In addition to conservative management of lymphoedema, there are surgical procedures which have been utilised in the treatment of this condition. These are not currently available in Northern Ireland. Surgical techniques include debulking operations and amputation, lymphovenous microsurgical reconstructive procedures and liposuction. There is no conclusive evidence to suggest that any of the above procedures are significantly effective, either in the short or long term management of lymphoedema, and are therefore not recommended.¹²⁻²²

 There is no indication for surgery in the treatment of lymphoedema.

7.5 Drug Management of Lymphoedema

 There is no conclusive evidence to recommend the use of benzopyrones in the treatment of lymphoedema.²³

7.6 Limitations of Literature Review of the Management of Lymphoedema

The following section (7.7) provides an overview of lymphoedema treatments as referred to in currently available literature, the limitations of which are considered as follows:

- There is a lack of high quality research in this specialist field. Whilst a Cochrane review has taken place and some recommendations made, the three reviewed studies all have limitations and have yet to be replicated. As such their results must be read with caution. A few other studies have also reported positive outcomes and are summarised in the following table. However, the overall lack of good research volume limits the available evidence to ensure correct review judgements.
- The vast majority of cited evidence is based on international expert opinion and consensus rather than RCTs.
- The effectiveness of CDT is attributed to the combination of its component parts. It is therefore difficult to assess various single aspects (e.g. MLLB) of this form of management in a research setting. This has ethical implications for future study design.
- The available studies commonly recruited breast cancer related lymphoedema patients into their trials. As such the areas of non-breast cancer, advanced disease and primary lymphoedema are still very much under investigated.

7.7 Comparison of Methods to Treat Lymphoedema


Method	Applicable	Non-invasive	In-expensive	Readily available in N.I	New equipment required	Effectiveness & benefits of treatment	Long term control	Ref no
CDT	Whole body.	Yes.	Yes.	No – very few funded posts regionally. Limited service (more readily available for oncology patients).	Yes – garment & bandage supplies (recurrent lifelong resource requirement).	Effective - - increased quality of life; - decreased limb volume; - decreased pain; - decreased lymph capillary pressure; - decreased infection rates.	Good if patient is compliant and has regular follow up (6 monthly).	2 24 25 26 1
MLLB	Upper and lower limbs, and male genitalia.	Yes.	Yes	No – very few funded posts regionally.	Yes – bandaging supplies (appropriate bandages are available on drug tariff).	Effective - - increased quality of life; - decreased limb volume especially in early stage of intensive phase (first 2 weeks).	Good if followed by compression garments. Nocturnal bandaging may also be included in phase 2 of management.	7 5 4
MLD	Whole body.	Yes.	Yes.	No – very few funded posts regionally.	No.	Effective - In combination with MLLB, skin care and exercise, especially in presence of fibrosis.	Good if followed by compression hosiery.	11 5 6 4 27
Skin care	Whole body.	Yes.	Yes.	Yes.	No, but access to pharmaceuticals required e.g. - emollients; - antibiotics; - antifungal. and at times specialist dermatological assessment.	Effective - - improved skin condition; - decreased risk of infection (cellulitis) or resolution of infections.	Good if patient complies with life long programme of care.	8

Method	Applicable	Non-invasive	In-expensive	Readily available in N.I	New equipment required	Effectiveness & benefits of treatment	Long term control	Ref no
Exercise (including deep breathing exercises).	Whole body.	Yes.	Yes.	Yes, if advised by a trained health care professional.	No.	Yes, in combination with MLD, MLLB/garments and skin care. Increased well being, confidence, control, motivation and no increase in lymphoedema.	Good if patient is compliant.	8
Garments (off the shelf and made to measure).	Whole body.	Yes.	No, especially the made to measure format. New garments required every 6 months. Patient should have at least 2 effective garments at all times.	Garments must be prescribed by a trained healthcare professional (some are now available on drug tariff). No current designated regional budget.	Yes. Garment supplies (stock and specialist orders).	Yes if fitted correctly and patient monitored regarding suitability, compliance and general health. Increased effectiveness if used following MLLB. Need to be worn daily.	Good if patient is compliant and condition stable.	10 9
Pneumatic Compression Pump.	Upper and lower limbs only.	Yes.	No.	Not currently used in Northern Ireland.	Recurrent compression garments cost – dedicated budget required. Yes.	Yes, but must be in combination with MLD (not to be used in isolation). Requires specific compartmentalised sequential pressure at precise settings. To be used under the guidance of a therapist.	Correct equipment must be available for long term use.	3

7.8 The Lymphatic System and Cancer – Management Considerations.

It is known that cancer spreads to lymph nodes and to distant organs via lymphatics and blood vessels.²⁸ The presence or absence of tumour in lymph nodes is important in staging of disease and in prognosis.^{29,30} Removal of regional lymph nodes is part of the surgical control of many cancers and lymphoedema may occur as a consequence in cancer survivors. The role of sentinel lymph node biopsy, especially in breast cancer is well established, allowing accurate identification of axillary lymph node status at the time of breast cancer surgery and consequent sparing of the axillary nodes where the sentinel node does not demonstrate metastatic cancer. This is beneficial to patients as it demonstrates reported lower incidence rates of lymphoedema and improved quality of life where it has been possible to conserve the axillary lymph nodes.³¹

The process of metastatic spread of cancer is complex involving an interaction of many factors including the new growth of intratumoural lymphatic vessels under the influence of vascular endothelial growth factors VEGFC and VEGFD and the carriage of tumour cells in lymphatic vessels.^{29,30} The discovery of a direct relationship between experimental tumour associated lymphangiogenesis and enhanced lymph node metastasis has raised the question as to whether any new therapeutic agents developed to treat lymphoedema by enhancing lymphangiogenesis could also increase the risk of tumour metastasis.²⁹ Similar concerns have been raised about the use of manual lymphatic drainage (MLD) which can enhance regional lymphatic drainage. There has been very little research in this area, however, a retrospective study of 191 patients with head and neck cancer and lymphoedema concluded that lymphatic drainage therapy for patients presenting with lymphoedema after cancer therapy does not increase the rate of local recurrence.³²

 Patients with lymphoedema carry a small risk of a malignant sarcoma developing in chronic lymphoedema.^{33,34} Health care professionals should be vigilant in detecting angiosarcoma. This tumour is treated surgically.

7.9 The Role of the Extended Multidisciplinary Team

Breast Care Nurse

The Breast Care Nurse (BCN) offers information and support to patients with breast problems, facilitates the decision making process and education of patients with breast cancer and is trained to recognize anxiety and depression and offer counselling where appropriate. The BCN sees patients in relation to body-image, psycho-sexual issues and rehabilitation. Advice is given regarding menopausal systems, such as vaginal dryness and night sweats, which can impact on those who also have lymphoedema. Patients with upper arm lymphoedema may need to have bras and prosthesis refitted. The BCN has a major role in the education of an at risk patient and should be responsible for informing patients who are to undergo axillary node clearance or mastectomy of the risk of lymphoedema and how to reduce this risk. BCNs should also ensure that pre and post operative bilateral arm circumferential

measurements are taken and recorded. Some BCNs are also fully trained lymphoedema therapists.

Dermatology

Dermatology is the diagnosis and management of skin disease. Dermatology staff should be involved in the care of patients with lymphoedema of whatever aetiology as many of the complications of lymphoedema manifest as skin disease e.g. elephantiasis skin changes including hyperkeratosis and toe space maceration; tinea infection, cellulitis, skin malignancy e.g. lymphangiosarcoma.

Some skin disease may result in secondary lymphoedema e.g. cellulitis and rarely other skin infections, dermatitis/eczema for example varicose, psoriasis, rosacea, granulomatous conditions e.g. oro-facial granulomatosis and sarcoidosis. Also the presence of skin complications may affect the successful management of lymphoedema e.g. ulceration, recurrent cellulitis, eczema and psoriasis.

Dermatology specialist nurses with recognised lymphoedema management training can be involved in lymphoedema management.

Dietetics

A small number of studies and reports, dating back over the last 60 years, have made reference to the potential effect of diet and body weight on the aetiology and possible management of lymphoedema. Specifically, references in the literature do exist about the potentially beneficial influence of weight reduction and low fat diets on arm swelling. However, there have been no randomized controlled trials to test this hypothesis.

Diet may help in the management of lymphoedema via the following mechanisms:

- A reduction in the number of adipocytes (fat cells) that would otherwise contribute to the swelling of the affected limb;
- A reduction in the size of adipocytes may have a beneficial effect on the arm;
- A reduction of fat under the arm (in the axilla) may improve lymph drainage through this area.

Overall weight reduction has shown significant correlation with a reduction in limb volume. However, there appears to be no significant beneficial effect of a low fat diet alone in women with lymphoedema. A dietitian should be consulted for all patients with associated weight problems to support the general advice regarding healthy living provided by the lymphoedema therapist.

Patients with hypoproteinaemia should also be referred to the dietetic service for nutritional advice. Whilst this condition is not the cause of true lymphoedema it can produce a similar physical presentation which also responds to compression.



Occupational Therapy

The role of the occupational therapist in the management of lymphoedema focuses on assisting the person to be more independent and make sense of their functional limitations, improving their quality of life and increasing their feelings of self-worth and self-esteem, thus helping the individual to resume normal life roles. This is achieved by completing a holistic assessment to evaluate the impact of the disease and its symptoms on the individual's physical, functional, psychological, social and spiritual status, and involves assessing and analysing loss of function, ability to carry out daily living activities (e.g. self-care, dressing, toileting, meal preparation, shopping, laundry), fatigue, pain, pressure relief problems, leisure and work roles, body image problems, relationships and sexuality in conjunction with the multi-disciplinary team.

Intervention may include advice and education on personal care including clothing, alternative coping strategies, prescription of equipment (including bariatric) to aid independence, and provision of housing adaptations. Risk management and advice on safety issues are also addressed e.g. limb care and the effect of excessive reaching, lifting and carrying. Pressure care is essential due to the reduced skin integrity.

Palliative Care Team

Complex palliative care problems for patients who also experience lymphoedema are often addressed. 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.

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.”

Patients who present with advanced disease may not always tolerate full decongestive therapy, and as a consequence, 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.

Care and responsibility for the patient with lymphoedema may be provided by the multi-professional team in various settings, for example, hospice, community (out-patient and/or domiciliary) or hospital as suits the patient and carers. However, the overall management of the patient with lymphoedema should be led and monitored by the lymphoedema therapist.

Physiotherapy

Physiotherapy is concerned with human function and movement, and maximises potential using physical approaches to promote, maintain and restore physical, psychological and social well-being. Knowledge of functional anatomy and ergonomics facilitate management particularly in complex multi-pathology cases.

Patients with lymphoedema may have predisposing problems directly related to the cause of the lymphoedema e.g. post mastectomy painful/stiff shoulder or breast scarring. Lymphoedema can itself cause other soft tissue problems such as postural changes resulting in a painful neck or back, and mobility issues due to limb size, loss of joint movement and general de-conditioning. As such, the physiotherapist can mobilize scar tissue, advise on postural alignment, prescribe specific exercises and stretches and if necessary, supply additional equipment to promote mobility, manage symptoms and improve quality of life. Other associated symptoms and conditions, such as cancer related fatigue, anxiety, general de-conditioning and pain can also be addressed by the physiotherapist.

Podiatry

Treatment of foot conditions by an experienced podiatrist is essential in the management of patients with lower limb lymphoedema as this condition can strain the skin's mechanical properties, hence leaving it at risk of breakage, infection and inflammation. Foot wounds and ulceration are noted in some patients with lymphoedema, particular where vascular changes exist. Fungal skin infections may also occur due to the close proximity of swollen digits. Good foot hygiene and close monitoring are therefore essential to prevent any secondary foot infections which may lead to hospital admission. Increased swelling in tissues surrounding the nails can also lead to in-grown toenails potentially requiring nail surgery.

Lymphoedema may restrict general mobility, and patients may not be able to physically reach and look after their feet. Advice on footwear, to accommodate the swelling, and provision of bespoke footwear may be required to enhance comfort and maintain mobility.

Tissue Viability (TVN)/Wound Management Nurse

The wound management nurse/TVN plays a key role in the management of people with lymphoedema. This includes the holistic assessment of the patient with particular emphasis on skin care/hygiene to prevent damage and minimise infections using appropriate emollients/topical treatments for hyperkeratosis and antifungal agents for the treatment of Athletes' foot. Wound or ulcer management is also essential for chronic ulceration, the majority of which only improve when adequate oedema management is achieved; dressings and treatments are designed specifically to suit the characteristics of the wound and exudates. Lower limb lymphoedema patients may also require lymphorrhoea (lymph leakage) management. The TVN also provides educational advice regarding the recognition and timely treatment of early signs of infection and cellulitis management; and general advice on health promotion

issues such as diet, exercise and smoking cessation. This educational role also includes wound management care for the other members of the multidisciplinary team.

The TVN, in conjunction with the overall lymphoedema plan and lymphoedema therapist, may also be involved in the fitting and re-measurement of compression garments in the maintenance phase of management. TVNs who are fully trained lymphoedema therapists may treat patients with lymphoedema and must be linked to the trust multidisciplinary treatment team.

Key Recommendations: Management of Lymphoedema

- All patients with a diagnosis of lymphoedema should be referred to the Trust's Lymphoedema Team. Lymphoedema should be treated as early as possible in order to prevent complications such as infections, fibrosis and increased swelling.
- All patients with a diagnosis of lymphoedema should be referred to a therapist with a formal qualification from a recognised school of lymphoedema management.
- A thorough assessment including physical and psychosocial needs should precede any treatment.
- Following assessment, the patient should be referred to appropriate members of the multidisciplinary team as deemed necessary.
- Complex decongestive therapy (CDT) is recognised as the treatment of first choice for Lymphoedema. The precisely co-ordinated interaction of manual lymphatic drainage (MLD), multi-layer lymphoedema bandaging, skin care and exercise is effectively combined as (CDT) for patients with lymphoedema.
- CDT may need to be modified in the presence of complex co-morbidities or due to patient choice. Clinical judgement may also recommend modified CDT in some circumstances. The reasons for modifying the treatment should be clearly stated on the patient's treatment plan, and be subject to regular audit.
- Treatment options should be discussed with the patient (and potentially carers) and an individualised patient-centred protocol agreed. If accepted by the patient and deemed appropriate by the therapist, the patient will undergo a period of intensive treatment Complex Decongestive Therapy (CDT).
- The second phase of treatment (maintenance) encourages the transfer of control to the patient. This may include teaching the principles of nocturnal self bandaging. Initially six-monthly reviews should be therapist led. However, telephone reviews should be considered if the patient's condition is considered to be stable and the patient competent in self-monitoring.

- Patients with lymphoedema require lifelong specialist compression garments. All patients should have a minimum of two effective and appropriate compression garments at a given time (no longer than six months apart).
- Surgery is not currently recommended for the management of this condition in Northern Ireland.
- Patients with lymphoedema carry a small risk of malignant sarcoma developing in chronic lymphoedema. Health care professionals should be vigilant in detecting angiosarcoma. This tumour is treated surgically.
- Systematic reviews of research have indicated that there is not enough evidence for the use of benzopyrones in the management of lymphoedema.
- In the presence of related cellulitis refer to the “CREST Guidelines for the Management of Cellulitis in Adults”. (Appendix 7)
- All patients should be provided with accurate contact details of the local lymphoedema services if advice is required.
- Patient education should include adequate information on lymphoedema and its management. Whilst given verbally, this must be supported in written format.

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8 Reducing the Risk of Lymphoedema - the Evidence

Over and above avoidance of the aetiological factor where possible e.g. Sentinel node biopsy (SNB) instead of routine axillary clearance in breast cancer treatment, evidence in support of reducing the risk of lymphoedema is limited. A number of studies have been undertaken to explore this area, which are summarised below. However, there is no strong body of evidence in support of any particular action to prevent lymphoedema. Most research limitations relate to insufficient study follow up, as lymphoedema may occur several years after a causative event. As a result, advice for patients has largely been developed based on a combination of knowledge of physiological principles and experience, both from patients and professionals.



 More long term research studies are required to investigate risk factors for developing lymphoedema.

Radiation¹⁻⁸ and the extent of surgical excision^{2,4 7,9-11} as a cause of lymphoedema is well documented. In terms of cancer treatment, avoidance of these causative factors is generally not possible. However a number of studies focus on actions that may be taken to reduce the chance of developing secondary lymphoedema in patients known to be at risk.

Surgical Technique

- Variations in cancer surgical techniques are discussed in a variety of papers to reduce risk of lymphoedema whilst not impacting on cancer outcome.¹²⁻¹⁸
- A small number of papers report advances in lymphatic microsurgery (mainly related to breast cancer) as a way of preventing lymphoedema, however there is insufficient information on which to base any conclusions. This surgery is not available in Northern Ireland.


Co-morbidity

- Having significant co-morbid conditions is generally thought to add to the risk of developing lymphoedema.¹⁹
 -  • With the exception of two studies,^{5,10} an increased body mass index (BMI), especially >30, is noted as a significant risk factor,^{1,4,6,20-27} and patients with a high BMI should be referred to dietetic services.
 -  • As noted in section 5.3.2, cellulitis is both a risk factor and cause of lymphoedema.^{15,28-31}
- Elevated blood pressure has also been cited as being a risk factor^{2,32} and indeed one study found a reduction in risk in patients on treatment for hypertension.¹⁰

Exercise

- The role of exercise, occupational workload and housework is considered with varying conclusions although a general overview of findings would indicate that exercise does not cause lymphoedema, and individuals who were more sedentary, may be at a higher risk of developing lymphoedema.³³
- Early physiotherapy intervention is noted in several papers to have a positive effect in reducing the risk of developing lymphoedema post surgery.^{34,35}

Education and Awareness

-  Experts in the area discuss at length the importance of raising awareness among health care professionals and patients of the potential risk, signs and symptoms.³⁶⁻⁴⁴
- Prospective monitoring for symptoms of lymphoedema is highlighted as being of vital importance throughout the literature as studies would indicate that early treatment is of benefit.⁴⁵

General advice which may be useful for patients in reducing the potential to develop lymphoedema is provided in Box 8.1.

Other Factors

- Air travel may trigger or worsen lymphoedema, therefore the avoidance of such may reduce this risk. This statement is based on anecdotal evidence.^{2,46}
- Anecdotal evidence also links venepuncture to lymphoedema. There are a limited number of papers which have studied this retrospectively.⁴⁷
- Although not found to be of significance in all studies which considered age as a potential risk factor for the development of secondary lymphoedema,⁵ a small number of studies (post breast cancer), indicate a higher incidence of lymphoedema in younger women.^{10,21,48}
- Anecdotal evidence from both professionals and patients would also point to trauma caused by sphygmomanometers or needle stick injury as being potential causes of lymphoedema. No evidence was found relating to either of these areas.

Box 8.1

General advice to reduce the risk of lymphoedema

- Avoid blood pressure readings and injections in at risk limbs if at all possible. Consider wearing an alert bracelet, carry an alert card;
- Avoid heavy lifting if the arm is affected;

- Use protective gloves in the garden etc;
- Avoid scratches, bites etc, use electric razor;
- Maintain good skin and nail care; take care with manicures, see a podiatrist;
- Maintain ideal weight;
- Exercise but don't fatigue the affected limb;
- Early detection – be aware of and act on early symptoms;
- Avoid hot baths, saunas and steam rooms;
- Use high factor sunscreen;
- Wear good fitting shoes;
- Avoid tight clothing (bra straps, belts etc);
- No tight jewellery;
- Don't smoke, avoid too much alcohol (general advice);
- Avoid hot or cold compresses; be cautious of heat producing therapies;
- Avoid standing or sitting for long periods.

Key Recommendations: Reducing the Risk of Developing Lymphoedema

- Improving knowledge about prevention is key to reducing the impact and burden of lymphoedema. More long term research studies are required to investigate risk factors for developing lymphoedema in patients undergoing surgical or radiotherapy treatment.
- As infection is intrinsically linked to lymphoedema, prompt assessment and treatment is vital to prevent recurrence and hospital admission. (Refer to CREST Cellulitis Guidelines).
- Awareness of the risk of lymphoedema should be raised with all patients prior to undergoing any form of treatment which is likely to cause lymphoedema and should be included in the consent process.
- Awareness of the signs and symptoms of lymphoedema and appropriate referral pathways should be cascaded to all relevant health care professionals, particularly within primary care and the specialties of oncology, palliative care, vascular surgery, genetics and dermatology.
- Patients with an increased body mass index (BMI), particularly over 30, may be at a higher risk of developing lymphoedema and as such, should be referred to dietetic services.
- Adequate in-depth education regarding lymphological disorders and correct treatment should be taught at undergraduate level to all relevant health care professionals.

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9 Lymphoedema Services in Northern Ireland

The 2004 Review of Lymphoedema services in Northern Ireland made a number of recommendations to establish a service to “ensure that people with lymphoedema in Northern Ireland receive cost effective, equitable and high quality lymphoedema services, based on the best evidence available.”

The review recommended that an integrated network of services should be established across the province to provide equal access to appropriate diagnosis, assessment and management by trained practitioners, along with an accompanying programme of activity to improve education and awareness, and support research.

The following recommendations have been made by the CREST Lymphoedema Group to assist with the implementation of the clinical guidelines in parallel with addressing the review recommendations.

Service Recommendations: Lymphoedema Services

- Each trust should establish a dedicated lymphoedema service to identify and address the needs of patients with, or at risk of developing, lymphoedema. A suggested profile of how the teams should be configured is outlined in Appendix 9.
- All trusts should ensure that the lymphoedema guidelines are implemented in their area.
- All patients with a diagnosis of lymphoedema should be referred to the trust lymphoedema team. A proposed service referral form is provided in Appendix 10. Patients with lymphoedema should be referred to the lymphoedema service as per the referral pathway outlined in Appendix 11.
- All patients with a medically complex presentation should have access to appropriate consultant input via regional complex lymphoedema clinics (referral criteria - Appendix 12).
- Ongoing information should be collated on patients to support service evaluation and future planning. A review form is provided in Appendix 13.
- An education and awareness programme about lymphoedema should be established for all relevant health care professionals.
- A regional clinical network should be formed to co-ordinate service development, research, education, patient support and other governance activities.
- The lymphoedema network should promote and support the implementation of the CREST Clinical Guidelines for Lymphoedema across Northern Ireland.
- A database of patient information should be established to support service evaluation, research and future planning.

10 Educational Competencies

10.1 Competency for Those Required to Have a General Awareness of Lymphoedema

The following outlines the educational competencies required by health care professionals who should have a general awareness of lymphoedema so that they ensure patients at risk of or with the condition are appropriately identified, treated and managed.

- To be able to identify those at risk of lymphoedema;
- To be capable of providing general advice on management of the condition and risk factors;
- To be able to provide the patient with advice on the importance of skincare;
- To be able to identify when a garment is poorly fitting;
- To be capable of applying and identifying problems with the garments (not measuring, selecting or fitting garments);
- To know where local services are based and be aware of the referral pathway for the assessment and management of the lymphoedema.

10.2 Specialist Lymphoedema Practitioners Should:

- Be fully trained, with a formal qualification from one of the recognised schools of lymphoedema management;
- Carry an active lymphoedema caseload;
- Have appropriate training and experience in the management of complex lymphoedema;
- Ensure knowledge and skills are kept current;
- Should be trained and competent in the use of the SF36v2 assessment tool for psychological assessment;
- Provide all lymphoedema treatments including skincare, exercise advice, garment measuring and fitting, multilayer short stretch lymphoedema bandaging and manual/medical lymphatic drainage;
- Competently treat severe and complicated lymphoedema;
- Provide education on lymphoedema;
- Work with the key workers (eg. vascular link nurse, breast cancer link nurse);
- Offer advice and support to levels all health care professionals and to liaise with other members of the multidisciplinary team as appropriate;
- Partake in audit of their practice;
- Review critical incidents and patient complaints.

Key Recommendation: Education and Training

- All health care professionals who may come into contact with patients with lymphoedema or at risk of developing the condition should be aware of the signs and symptoms along with the relevant diagnostic and treatment pathways.

11 Audit and Research

Improving the knowledge base for lymphoedema is vital in terms of gaining more accurate information to underpin service development as well improving clinical aspects to diagnosis and treatment.

11.1 Audit of Lymphoedema Services and Clinical Practice

Audit is one of the key implementation tools in monitoring the improvement of Lymphoedema services.

The audit trail should follow the patient on their treatment journey from diagnosis to treatment and lifelong maintenance of lymphoedema. Service structures and processes along with clinical practice and outcomes should be interrogated and audited against the standards derived from the CREST guidelines.

The objectives of developing an audit programme for lymphoedema services across Northern Ireland are as follows:

- To improve services for patients with lymphoedema and identify and address gaps in the current system;
- To address current inequalities in access to services;
- To support future commissioning and planning of services;
- To standardise clinical practice through supporting the implementation of evidence based guidelines;
- To support increased awareness and education initiatives;
- To strengthen multidisciplinary working;
- To improve patients experiences of treatment and care.

Audit priorities for lymphoedema services have been identified by the guideline group and are outlined in Box 11.1. In order to facilitate standardised audit across Northern Ireland, a range of audit proformas are in the process of being developed and will be accessible on the Lymphoedema Network.

Box 11.1 - Audit Topics

Service Structure:

- Environment where lymphoedema is treated;
- Access to assessment clinics;
- Availability of equipment and consumables;
- Trained personnel;
- Access to complex clinics.

Process:

- Triage of referrals;
- Patient diagnosis and assessment;
- Patient having complex decongestive therapy (CDT) and reasons for modifying CDT;
- Patient follow up;
- Waiting times for diagnosis assessment and treatment;
- Attendance for treatment;
- Outcomes of treatment.

Box 11.1 - Audit Topics	
<p>Information:</p> <ul style="list-style-type: none"> • Access to timely information; • Pre surgery risk assessment: Audit of cancer data sets for baseline limb volume assessment. 	<p>Standards and Guidance:</p> <ul style="list-style-type: none"> • Implementation of CREST Guidelines.
<p>Patient orientated outcomes:</p> <ul style="list-style-type: none"> • Patient satisfaction; • Incidence of cellulitis; • Audit of patient support group telephone helpline. 	

11.2 Research

The guideline development group has identified the following areas as priorities for further research in Northern Ireland.

- Epidemiological study into the incidence and prevalence of lymphoedema in Northern Ireland;
- Further research into treatment options including patient preferences on complex decongestive therapy, optimal length of treatment and considering the impact of early versus later treatment;
- Further research into compliance and opinions of compression garments used for treatment;
- Continued research into the reliability and validity of diagnostic methods such as bioimpedance analysis.

Key Recommendations: Audit and Research
<ul style="list-style-type: none"> • Audits should be undertaken using standardised proformas developed by the Lymphoedema Network where available. • Partnerships between relevant institutions such as the Lymphoedema Network, Research and Development Unit and universities, should initiate research activity to address identified research priorities for lymphoedema.

12 Information Resources for Patients

Local Support Groups		
Foyle Lymphoedema Support Group	028 71351010	Ask for the lymphoedema support group
Northern Ireland Lymphoedema Support Group	Tel: 028 90667570, e-mail: info@nilsg.co.uk http://www.nilsg.co.uk	Aims to provide support for people with lymphoedema, provide information about lymphoedema and its treatment, work towards the availability of better resources for lymphoedema treatment and work in partnership with local healthcare professionals involved in the management of lymphoedema - NILSG offers a telephone helpline and support group meeting twice a year.
National Support Groups		
British Lymphology Society	http://www.lymphoedema.org/bls/	BLS aims to promote awareness about lymphoedema to the public and healthcare professionals.
UK Lymph.com	http://www.uklymph.com/	This site has been set up in conjunction with the East Kent Lymphoedema Support Group to offer online support and advice to lymphoedema sufferers.
Lymphoedema. Support Network	http://lymphoedema.org/lsn	The Lymphoedema Support Network (LSN) provides information and promotes self help to educate and support patients with this condition.

Footwear Resources		
Molly Pollys	http://www.mollypollys.com/	A family business that designs, manufactures and supplies specialist footwear and protectors for people that suffer with disorders including lymphoedema.
Cosy Feet Ltd	http://www.cosyfeet.com	Providers of extra roomy footwear for swollen feet, they also provide specialist socks and hosiery. Range includes foot and leg care products.

Information Leaflets

All information leaflets are available to download from the Lymphoedema Network - Northern Ireland website:

- What are emollients?
- Reducing the risk of developing lymphoedema following treatment for cancer;
- Cellulitis (acute inflammatory episodes);
- Exercise and mobility;
- Holidays and travel – advice for patients with lymphoedema;
- Skin care for patients with lymphoedema;
- What is lymphoedema?
- Treatment and management of lymphoedema;
- Taking care of swollen feet;
- Early years lymphoedema (primary lymphoedema);
- Primary lymphoedema;
- Reducing the risk of developing lymphoedema;
- Skin care for patients at risk of developing lymphoedema;
- Holidays and travel patient advice for those at risk;
- Reducing the risk of developing lymphoedema following treatment for breast cancer.

Copies of the information booklet, ‘Lymphoedema – your questions answered’, are available free of charge from the Northern Ireland Lymphoedema Support Group (NILSG).

Appendix 1: Guideline Development Group

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Mr	Paul	Bateson	Vascular Surgeon Director of Surgery	Royal Hospital, Belfast Trust
Ms	Karen	Chambers	Senior Occupational Therapist	Belfast City Hospital, Belfast Trust
Ms	Emma	Christie	Specialist Physiotherapist	North & West, Belfast Trust
Dr	Jackie	Clarke	Oncologist	Belvoir Park Hospital, Belfast Trust
Ms	Jill	Curry	Dietitian	Altnagelvin Hospital, Western Trust
Mr	Stephen	Dobbs	Gynaecological Cancer Surgeon	City Hospital, Belfast Trust
Dr	Julie	Doyle	Consultant in Palliative Care	Mater Hospital, Belfast Trust; N.I. Hospice
Ms	Celene	Duffy	Breast Care Nurse Specialist	Altnagelvin Hospital, Western Trust
Dr	Shauna	Fannin	GP	Ballymoney Health Centre
Ms	Paula	Fearon	Breast Care Nurse	Craigavon Area Hospital Southern Trust
Mr	Joe	Feeney	CREST Secretariat	DHSSPS
Ms	Caroline	Graham	Tissue Viability Nurse	South & East, Belfast Trust
Ms	Rose	Greene	Hospice Nurse	Foyle Hospice Home Care
Dr	Hilary	Jenkinson	Dermatologist	Antrim Area Hospital, Northern Trust
Mr	Stephen	Kirk	Consultant Surgeon	Ulster Hospital, South Eastern Trust
Mr	Bernard	Lee	Vascular Surgeon	City Hospital, Belfast Trust
Ms	Jill	Lorimer	Specialist Physiotherapist	Belfast City Hospital, Belfast Trust
Mr	Joe	Magee	Secondary Care	DHSSPS
Ms	Maria	Magee	District Nurse/ Macmillan Education Facilitator	Eglinton Health Centre

Ms	Patricia	Magee	Specialist Physiotherapist	South & East, Belfast trust
Dr	Adrian	Mairs	Public Health Medicine	NHSSB
Ms	Caroline	Mason	Specialist Registrar	WHSSB
Ms	Claire	McGartland	AHP Commissioning	WHSSB
Ms	Roisin	McKeown	Tissue Visability Nurse	Belfast Trust
Ms	Cliona	McCotter	Senior Physiotherapist	Marie Curie Cancer Centre Belfast
Prof	Noel	McHale	Physiologist	Dundalk Institute of Science/Queens University Belfast
Ms	Pearl	Millar	Physiotherapy	Altnagelvin Hospital, Western Trust
Dr	Barry	Mitchell	GP	Lodge Health
Dr	Liz	Mitchell	Deputy Chief Medical Officer	DHSSPS
Dr	Mary	Morris	Clinical Psychologist	Altnagelvin Hospital, Western Trust
Ms	Margaret	Napier	Senior Physiotherapist	NI Hospice
Ms	Marianne	O'Donnell	Service User	Foyle Hospital Lymphoedema Support Group
Ms	Anne	O'Hagan	Palliative Care Nurse	Antrim Area Hospital, Northern Trust
Ms	Ena	O'Hanlon	Service User	NI Lymphoedema Support Group
Ms	Breige	O'Kane	Service User	NI Lymphoedema Support Group
Ms	Jane	Rankin	Superintendent Physiotherapist	City Hospital, Belfast Trust
Ms	Sigi	Refsum	Breast Cancer Surgeon Consultant	City Hospital, Belfast Trust
Ms	Heather	Reid	Project Manager	EHSSB
Ms	Mary	Waddell	Director of Nursing	EHSSB
Ms	Anne	Witherow	Assistant Director of Nursing	Altnagelvin Hospital, Western Trust
Dr	Michael	Young	Consultant Urologist	Craigavon Area Hospital, Southern Trust

CREST would also like to express their thanks to the many other health care professionals who contributed to the document.

Appendix 2: Key Questions

Prevention Management and Assessment of Lymphoedema

Prevention/Risk Reduction

1. Is there any evidence that giving advice on prevention is effective in patients with increased risk of developing lymphoedema?
2. Is there any evidence that involving patients in treatment decisions reduces the risk of developing lymphoedema?
3. What evidence is there on the value of various types of information – leaflets, booklets etc?
4. Is there any evidence that increasing awareness among professionals will prevent complications of lymphoedema?
5. Is there any evidence of the effectiveness of current routine lymphoedema prevention advice?
6. What evidence is there available to suggest how patients make decisions regarding lifestyle and prevention?
7. Is there evidence to suggest that variations in surgical techniques (for cancers such as head and neck, breast, gynae or melanoma) may prevent the development of lymphoedema?
8. Is there any evidence to suggest that patients are routinely given advice on the prevention of lymphoedema prior to surgery?
9. Is there evidence to suggest that treatments such as radiotherapy may be adjusted to prevent the development of lymphoedema?
10. Is there any evidence to suggest the best time in a patient's treatment to achieve optimum compliance with advice?
11. Is there any evidence to suggest that a reduction in weight reduces the risk of developing lymphoedema?

B Diagnosis/Assessment

12. Is there evidence to suggest what skills are required to make an appropriate assessment and diagnosis of lymphoedema?
13. Is there any evidence that early diagnosis of lymphoedema makes a difference to outcome?
14. Is there any evidence of what information is required to make a diagnosis?
15. Is there evidence of a specific list of specific clinical features or symptoms which would indicate a diagnosis of lymphoedema?
16. What evidence exists to indicate the most robust method (including clinical investigations) of diagnosis?
17. Is there evidence to indicate what stage referral to a 'specialist' is necessary – pre formal diagnosis, assessment, and treatment?
18. Does evidence exist that support patient education in recognising symptoms in preventing complications?
19. Does evidence exist that support patients education in recognising the development of cellulites if they have lymphoedema?

20. Is there evidence to support the assessment of staging for lymphoedema?
21. Is there evidence to suggest which outcome measures are important in terms of diagnosis?

C Management of Patients with Lymphoedema

22. What evidence is there about the effectiveness of good skin care in relation to the treatment of lymphoedema?
23. Is there any evidence to suggest the types of skin care products to be used?
24. Is there any evidence to suggest what skin care products should be used if a patient is particularly allergic?
25. Where interruption of skin integrity is necessary, is there any evidence to suggest precautions to be taken to prevent the development of lymphoedema or complications?
26. What evidence exists about the effectiveness of maintaining a healthy weight in improving outcomes?
27. Does any evidence exist regarding the benefit of taking regular exercise?
28. Does any evidence exist regarding the amount or type of exercise that should be undertaken?
29. Is there any evidence to suggest that weight management is useful in the management of lymphoedema?
30. Does evidence to suggest if patients have poor nutritional status (and in particular low protein levels), that they are at risk of developing lymphoedema?
31. Does evidence exist regarding the benefit of improving nutritional status in patients with lymphoedema improves outcomes?
32. Is there any evidence to suggest that good nutritional status will help prevent complications associated with lymphoedema?
33. Is there any evidence to suggest that nutritional support will help alleviate complications such as leakage at the site of lymphoedema?
34. Does any evidence exist regarding the benefit of early versus late intervention in regard to treating lymphoedema?
35. Does any evidence exist regarding the effectiveness of manual lymphatic drainage?
36. Does any evidence exist regarding the skills required to undertake manual lymphatic drainage?
37. Does any evidence exist regarding the effectiveness of simple lymphatic drainage?
38. Does any evidence exist regarding the skills required to undertake simple lymphatic drainage?
39. Does any evidence exist regarding the effectiveness of bandaging and compression?
40. Does any evidence exist regarding the skills required to apply bandaging and compression techniques?
41. Is there any evidence to support the use of compression pumps in the treatment of lymphoedema?
42. Is there any evidence to support the use of combined modalities in improving outcomes?
43. Is there any evidence to suggest the optimum length or intensity of treatments for lymphoedema?

44. Is there any pharmacological evidence for the treatment of lymphoedema (particularly in relation to diuretics, benzopyrones and antibiotics)?
45. Is there any evidence to suggest what antibiotics should be used for the effective treatment and prevention of infection, including cellulitis?
46. Is there any evidence to suggest how long antibiotic therapy should continue for at full dose in treating lymphoedema related cellulites?
47. Is there any evidence regarding the most effective treatment of infection?
48. Is there any evidence regarding the impact of social and psychological support for patients with lymphoedema in improving quality of life?
49. Is there any evidence to suggest the most effective type of social or psychological advice?
50. Is there any evidence to support specific models of care in improving patient outcomes?
51. Is there any evidence to support the role of support groups in the treatment of patients with or at risk of developing lymphoedema?
52. Is there any evidence to identify the most appropriate clinical settings in which initial treatment and follow up should take place?
53. Is there evidence to suggest that taking blood pressure or vunenpuncture on an affected limb affects outcomes in patients at risk of developing lymphoedema?

D Patient Advice

54. Is there any evidence to suggest that giving patients advice on prevention at time of treatment will decrease the incidence of lymphoedema?
55. Is there any evidence on the most effective way of communicating information on prevention to patients?
56. Is there any evidence to indicate how patients make decisions on compliance versus implications on quality of life?
57. Is there any evidence to suggest what advice content should be included to prevent the development of lymphoedema or to improve outcomes?

E Education and Training

58. Is there evidence of established educational competencies for those diagnosing, assessing and treating patients with lymphoedema?
59. What is the best method of ensuring widespread knowledge of the guidelines in primary care and secondary settings?

F Service Standards

60. Do standards for service provision exist?
61. What evidence exists in relation to cost benefit analysis of the provision of lymphoedema services?

Appendix 3: Literature Search Strategy

In order to identify evidence relevant to the research questions set by the CREST steering group, literature searches were undertaken of electronic databases of published studies.

The following databases were used:

- Medline
- Embase
- EBM Reviews/Cochrane Library
- Cinahl
- Psychinfo
- HMIC
- Pubmed

Whilst CREST is responsible for producing clinical guidelines for use in Northern Ireland, it is not supported by a specific information unit skilled in undertaking evidence reviews.

The following steps were taken to ensure the review of evidence used in the development of the recommendations was as robust as possible.

Advice was sought from organisations recognised for their experience in developing clinical guidelines such as SIGN and NICE and their processes studied. Those involved in undertaking the literature review have all had prior experience in this area.

In order to optimise the number of relevant studies, search strategies relevant to the research questions were discussed by the project manager and information specialists at the Queens University Belfast medical library to elicit key word and identify alternate terms.

Literature searches for a number of key questions were undertaken by the information specialist at the medical library as well as the project manager to increase the sensitivity of the overall search. The lists of records generated were then compared for relevant papers.

The project manager sifted the initial list of records generated by both individuals and made the final study selections. Abstracts of the identified papers were read to ensure relevancy and a list of studies were identified for which the original papers were accessed and critically appraised.

The clinical questions covered a wide number of topic areas. For some areas, a large volume of studies were identified. In specific topic areas the search strategies were rerun with the addition of syntax filters to identify only RCTs, systematic reviews or interventional studies. Filters compiled by the Cochrane centre were used.

Relevant papers in each of the key question areas were summarised and an evidence level attached. Tables summarising the review were submitted to the wider steering group for discussion along with an overview of the literature relating to the questions posed originally by the group.

The wider group was also asked to identify if they knew of any key relevant papers which were missing, based on their own knowledge of the literature or searches that they had carried out.

Where evidence was lacking or inconclusive, and if insufficient expertise was available on the group, additional appropriate input was sought to ensure the recommendations would be locally feasible as well as reflecting consensus of opinion.

Evidence tables are available on the GAIN website.

Levels of Evidence	
1++	High quality meta analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+	Well conducted meta analyses, systematic reviews of RCTs, or RCTs with a low risk of bias
1 -	Meta analyses, systematic reviews of RCTs, or RCTs with a high risk of bias
2++	High quality systematic reviews of case-control or cohort studies High quality case-control or cohort studies with a very low risk of confounding, bias, or chance and a high probability that the relationship is causal
2+	Well conducted case control or cohort studies with a low risk of confounding, bias, or chance and a moderate probability that the relationship is causal
2 -	Case control or cohort studies with a high risk of confounding, bias, or chance and a significant risk that the relationship is not causal
3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion

Appendix 4: Overview of Quality of Life Tools

Overall conceptual and methodological rigour is lacking when looking at assessment tools in the literature. There is a need for an encompassing, unequivocal tool to assess quality of life / psychological issues in individuals with lymphoedema for all areas of the body.

Quality of Life Tools – an Overview

- There are no specific quality of life measures for patients with lymphoedema. The two most commonly used questionnaires on this target population are SF-36 and FACT-B. Both questionnaires are short, yet have been found to be sensitive and reliable.
- SF-36 is a general health measure which numerous studies have found useful in assessing QOL in patients with cancer.^{1,2}
- FACT-B focuses specifically on QOL in breast cancer patients, but is not primarily aimed at those who suffer from lymphoedema.
- The FLQA-1 was found to be a reliable and valid measure of QOL³
- One study found the SF-36 particularly useful in measuring QOL in patients' with lower limb lymphoedema, in comparison to other questionnaires.²
- The Functional Assessment of Cancer Therapy has five subscales. These measure functional, social, physical and emotional factors and a patients' relationship with their doctor in relation to QOL. One study found that women who had lymphoedema scored significantly lower on all subscales than those who did not.⁴
- The FACT-B was specifically developed to measure QOL in patients with breast cancer. The presence of lymphoedema was significantly associated with decreased quality of life.⁵
- The FACT-B tool was found to be valid and reliable after the addition of a four-item subscale as well as being sensitive to rehabilitation over time,⁶ thus making it a useful measure for longitudinal studies.
- The Nottingham Health Profile (NHP-1) measures QOL in terms of emotional, social and physical factors. One study found that after treatment participants scored significantly higher in physical mobility subscale and lower on the one measuring pain.⁷
- The Upper Limb Lymphoedema questionnaire (ULL-27) was found it to be much more sensitive than previous scales in measuring QOL in patients' with upper limb lymphoedema.⁸
- The Dermatology Life Quality Index is another measure of QOL. Scores on this index significantly improved after a hygiene and skin care regimen was introduced to treat lymphoedema.⁹
- In comparisons of the RAND 36 and the Functional Living Index Cancer (FLIC) in breast cancer patients with and without lymphoedema. Both instruments focused on different aspects of QOL. The FLIC demonstrated greater sensitivity to emotional wellbeing.¹⁰

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Appendix 5: SF-36v2™ Health Survey

This survey asks for your views about your health. This information will help you keep track of how you feel and how well you are able to do your usual activities.

Answer every question by selecting the answer as indicated. If you are unsure about how to answer a question, please give the best answer you can.

1. In general would you say your health is:				
Excellent	Very Good	Good	Fair	Poor
2. Compared to one year ago, how would you rate your health in general now?				
Much better now than one year ago	Somewhat better now than one year ago	About the same as one year ago	Somewhat worse now than one year ago	Much worse now than one year ago
3. The following questions are about activities you might do during a typical day. Does YOUR HEALTH NOW LIMIT YOU in these activities? If so, how much?				
		Yes, limited a lot	Yes, limited a little	No, not limited
a	Vigorous activities , such as running, lifting heavy objects, participating in strenuous sports			
b	Moderate activities , such as moving a table, pushing a vacuum cleaner, bowling, or playing golf			
c	Lifting or carrying groceries			
d	Climbing several flights of stairs			
e	Climbing one flight of stairs			
f	Bending, kneeling, or stooping			
g	Walking more than a mile			
h	Walking several hundred yards			
i	Walking one hundred yards			
j	Bathing or dressing yourself			

4. During the past 4 WEEKS, how much of the time have you had any of the following problems with your work or other regular daily activities AS A RESULT OF YOUR PHYSICAL HEALTH?		All of the time	Most of the time	Some of the time	Little of the time	None of the time
a	Cut down on the amount of time you spent on work or other activities					
b	Accomplished less than you would like					
c	Were limited in the kind of work or other activities					
d	Had difficulty performing the work or other activities (for example, it took extra effort)					
5. During the past 4 WEEKS, how much of the time have you had any of the following problems with your work or other regular daily activities AS A RESULT OF ANY EMOTIONAL PROBLEMS (such as feeling depressed or anxious)?		All of the time	Most of the time	Some of the time	Little of the time	None of the time
a	Cut down on the amount of time you spent on work or other activities					
b	Accomplished less than you would like					
c	Did work or activities less carefully than usual					
6. During the past 4 WEEKS, to what extent has your physical health or emotional problems interfered with your normal social activities with family, friends, neighbours, or groups?		Not at all	Slightly	Moderately	Quite a bit	Extremely

7. How much BODILY pain have you had during the PAST 4 WEEKS?						
None	Very mild	Mild	Moderate	Severe	Very Severe	
8. During the PAST 4 WEEKS, how much did PAIN interfere with your normal work (including both work outside the home and housework)?						
Not at all	Slightly	Moderately	Quite a bit	Extremely		
9. These questions are about how you feel and how things have been with you during the PAST 4 WEEKS. For each question, please give the one answer that comes closest to the way you have been feeling. How much of the time during the past 4 weeks...						
		All of the time	Most of the time	Some of the time	Little of the time	None of the time
a	Did you feel full of life?					
b	Have you been very nervous?					
c	Have you felt so down in the dumps that nothing could cheer you up?					
d	Have you felt calm and peaceful?					
e	Did you have a lot of energy?					
f	Have you felt downhearted and depressed?					
g	Did you feel worn out?					
h	Have you been happy?					
i	Did you feel tired?					
10. During the PAST 4 WEEKS, how much of the time has your PHYSICAL HEALTH OR EMOTIONAL PROBLEMS interfered with your social activities (like visiting friends, relatives, etc.)?						
All of the time	Most of the time	Some of the time	Little of the time	None of the time		

11. How TRUE or FALSE is each of the following statements for you?						
		Definitely true	Mostly true	Don't know	Mostly false	Definitely false
a	I seem to get sick a little easier than other people					
b	I am as healthy as anybody I know					
c	I expect my health to get worse					
d	My health is excellent					

Thank you for completing these questions!

Appendix 6: Lymphoedema Assessment Form

Name: _____	Date of Assessment: _____
Address: _____	Diagnosis/P.C: _____
Health and Care No/Other: _____	_____
DOB: _____	Telephone No: _____
GP: _____	Communication/ Comprehension _____
Source of Referral _____	_____

Consent for Assessment Treatment Digital Imaging

If No, Why? _____

History of oedema (inc onset, progression, patients' perception, management of oedema and outcome)

Date of onset of oedema _____

Current symptoms _____

Recent/Ongoing Investigations _____

Surgery _____

Regional Lymph Node Clearance Sentinel Node Biopsy No. of Nodes removed _____

No. of +ve nodes _____

Radiotherapy _____ Hormonal therapy _____

Chemotherapy _____

Family History of oedema Y N

PMH/General Health (inc CVA, TIA, Skin conditions, Intermittent claudication, night pain into big toe, Peripheral neuropathy)

MANADATORY QUESTIONS:

Diabetes Epilepsy MI/Angina Wgt Loss/Gain Previous Surgery

RA Chest Renal Thyroid

Medication _____

Drug Sensitivities/Allergies (inc Latex) _____

Current/Previous History and Management of Cellulitis

General Contraindications	Y	or No	Contraindications to Neck Treatment	Y	or N
Uncontrolled heart failure	<input type="checkbox"/>	<input type="checkbox"/>	Cardiac arrhythmia	<input type="checkbox"/>	<input type="checkbox"/>
Acute deep vein thrombosis	<input type="checkbox"/>	<input type="checkbox"/>	Hyperthyroid	<input type="checkbox"/>	<input type="checkbox"/>
Acute infections	<input type="checkbox"/>	<input type="checkbox"/>	Hypersensitivity of carotid sinus	<input type="checkbox"/>	<input type="checkbox"/>
Superior vena cava obstruction	<input type="checkbox"/>	<input type="checkbox"/>	Patients over the age of 60 (Relative Contraindication)	<input type="checkbox"/>	<input type="checkbox"/>
Renal failure	<input type="checkbox"/>	<input type="checkbox"/>			
Acute dyspnoea	<input type="checkbox"/>	<input type="checkbox"/>			
Malignancies (Relative Contraindication)	<input type="checkbox"/>	<input type="checkbox"/>			
Contraindications for Deep MLD of abdominal region					
Pregnancy/Menses	<input type="checkbox"/>	<input type="checkbox"/>	Diverticulitis	<input type="checkbox"/>	<input type="checkbox"/>
Recent abdominal surgery	<input type="checkbox"/>	<input type="checkbox"/>	Abdominal aortic aneurysm	<input type="checkbox"/>	<input type="checkbox"/>
DVT in pelvic region	<input type="checkbox"/>	<input type="checkbox"/>	Unexplained abdominal pain	<input type="checkbox"/>	<input type="checkbox"/>
Inflammatory bowel disease	<input type="checkbox"/>	<input type="checkbox"/>	Previous radiotherapy over abdomen	<input type="checkbox"/>	<input type="checkbox"/>

Social History

Occupation _____

Hobbies _____

Smoker Y N

Lives alone Lives with _____

Type of Accommodation Flat House Bungalow

Sheltered Dwelling Residential home Nursing home

Ownership Own NIHE Private rent Other _____

Bathroom Upstairs Downstairs

Toilet Upstairs Downstairs

Steps/Stairs Internal steps External steps Stairs

Lift Ramp

Sleeps in Bed Chair

Community Services/Carer Support _____

Benefits _____

Functional activities and limitations in personal/domestic ADL, transfers/Hx of falls, mobility aids.

SYMPTOMS

Main Complaint _____

Location of oedema _____

Numbness/Tingling/Altered sensation _____

Any pain/night pain/discomfort associated with lymphoedema: Yes No

Location of pain _____

Duration of pain: Constant Intermittent N/A

Overall Condition: Improving Worsening Static

Any occurrence of genital oedema:

Yes No N/A

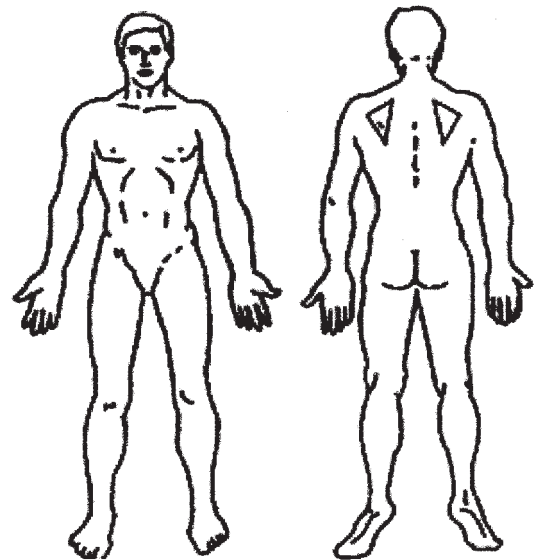
Symptoms of lymphoedema relieved by:

Elevation Exercise Massage

Compression Hosiery

Unable to relieve symptoms

Patient's expectations of treatment: _____



OBJECTIVE EXAM

Observation (posture/scars)	Dominant side R <input type="checkbox"/> L <input type="checkbox"/>
	Non pitting oedema <input type="checkbox"/>
Limb shape distorted <input type="checkbox"/>	Pitting oedema <input type="checkbox"/>
Deepened skin folds <input type="checkbox"/>	Tissue thickening <input type="checkbox"/>

SKIN ASSESSMENT

Discoloration: _____

- Temperature: Normal Cold Warm
- Sensation: Intact Altered
- Subcutaneous Tissue Normal Fibrotic Fatty Rubbery
- Non Pitting Pitting
- Skin: Broken Fragile Blisters Hyperkeratosis
- Taut/Shiny Ulceration Lymphorrhoea Dry
- Fungal infection

STEMMER SIGN: _____

WEIGHT (Kg): _____ HEIGHT (m) _____ BMI _____

- Doppler/ABPI Yes No Pulses R L
- To be ordered Yes No Sensation R L

RANGE OF MOVEMENT / STRENGTH (Full / Limited)

UPPER LIMB	ROM	Strength	LOWER LIMB	ROM	Strength
Neck			L/SP		
Shoulder			Hip		
Elbow			Knee		
Wrist/Hands			Ankle/Foot		

SHORTENED RIVERMEAD MOBILITY SCORE:

ITEM	INITIAL ASSESSMENT	DISCHARGE
Turning over		
Lying to sitting		
Sitting balance		
Sitting to standing		
Standing unsupported		
Transfers		
Walking indoors		
Stairs		

- 0 – Unable to perform
- 1 – assistance of 2 people
- 2 – assistance of 1 person
- 3 – requires supervision
verbal instruction
- 4 – requires aid/appliance
- 5 – independent



CIRCUMFERENTIAL/VOLUME MEASUREMENTS (see attached sheet)
 BIOIMPEDENCE MEASUREMENTS (see attached sheet)

EVALUATION

LYMPHOEDEMA TYPE: Primary Lymphovenous
 Secondary Dependency

GRADING/STAGING

Stage 0 (Latency) Stage 2 (Non-pitting spontaneous irreversible)
 Stage 1 (Pitting reversible) Stage 3 (Elephantiasis)

FACTORS AFFECTING OUTCOME

Venous complications		Functional difficulties	
Arterial complications		General immobility	
Pain		Lack of domestic support	
Lipoedema		Active Cancer	
High BMI		Sedentary lifestyle	
Recurrent AIE		Appropriate Rx not available	
Chronic skin condition eg psoriasis, eczema		Appropriate Rx not accepted	
Other uncontrolled symptoms eg nausea, fatigue			

AGREED SHORT TERM GOALS

- Reduce Limb Volume Improve AROM of _____ UL/LL
- Improve Strength of _____ UL/LL
- Patient Education – Skin Care, HEP, Adherence to Lymphoedema precautions
- Pain Reduction Other/Functional Goals _____

AGREED LONG TERM GOALS

- Reduce limb volume Improvement in pitting oedema
- Restore normal limb shape Patient to be independent with self bandaging
- Improve AROM of UL/LL Improve strength of UL/LL
- Reduce pain Tissue softening
- Patient to be independent with lymphoedema management / skin care
- Patient to be independent with home exercise programme
- Referral for compression garment and instructions for donning / doffing garments
- Other/ functional goals _____



PLAN

Intensive Phase		Maintenance Phase	
MLD		Skin Care	
MLB		SLD	
Skin Care		Exercise	
SLD		Garment	
Exercise		Self Bandaging	
Teach Self Bandaging		Other	
Education			
Other			

Frequency: _____ Duration: _____

CDT Modified due to:

Co-morbidities (please state) _____

Patient Choice

Level of resources

Other

CONSENT

Consent Form 3 to be completed

Blue copy given to patient Y N

Assessors Name: Print _____

Signature _____

Profession _____

Visual analogue scales

Functioning

Where are you now functioning for Home activities (H), Work activity (W) and Self Care (SC)?

Unable to function \longrightarrow No limitations of activities

Home Activities
0  10

Work Activities
0  10

Self Care Activities
0  10

Pain

Pain Intensity
0  10
(No Pain) (Worst Pain Imaginable)

Appendix 7: CREST Guidelines for the Management of Cellulitis in Adults

Diagnosis

Flu-like symptoms, malaise
onset of unilateral swelling, pain, redness

Decide Classification

Class I	Class II	Class III	Class IV
Patients have no signs of systemic toxicity, have no uncontrolled co-morbidities and can usually be managed with oral antimicrobials on an outpatient basis.	Patients are either systemically ill or systemically well but with a co-morbidity such as peripheral vascular disease, chronic venous insufficiency or morbid obesity which may complicate or delay resolution of their infection.	Patients may have a significant systemic upset such as confusion, tachycardia, tachypnoea, hypotension, or may have unstable co-morbidities that may interfere with a response to therapy or have a limb threatening infection due to vascular compromise.	Patients have sepsis syndrome or severe acute life threatening infections such as necrotizing fasciitis.

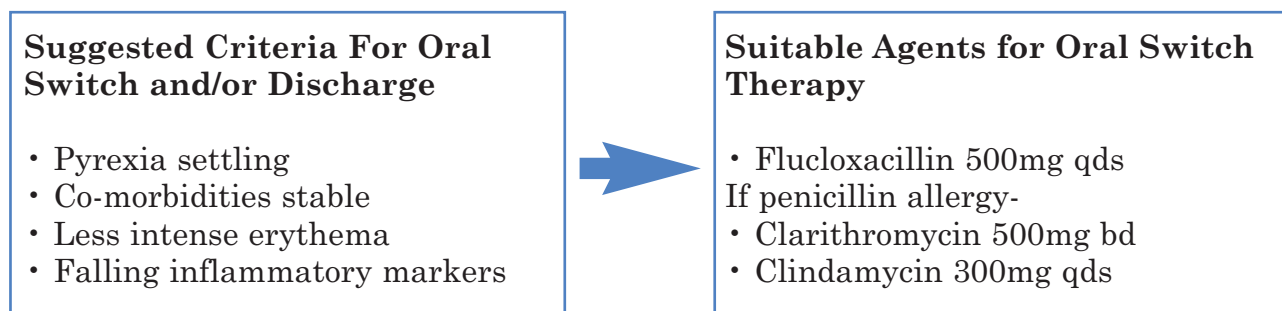
Lab Investigations

Class II - IV	Selected Patients
FBC ESR or CRP U+E Culture any ulceration or blister fluid.	<ul style="list-style-type: none"> • Blood cultures only Class III or Class IV. • Streptococcal serology only in refractory cases where diagnosis is in doubt. • Skin biopsy where differential diagnosis includes other inflammatory lesions.

Treatment

	First line	Second line
Class I	Flucloxacillin 500mg qds po	Penicillin allergy - Clarithromycin 500mg bd po
Class II	Flucloxacillin 2g qds IV or *Ceftriaxone 1g od IV (OPAT)	Penicillin allergy - Clarithromycin 500mg bd IV or Clindamycin 600mg tds IV
Class III	Flucloxacillin 2g qds IV	Penicillin allergy - Clarithromycin 500mg bd IV or Clindamycin 900mg tds IV
Class IV	Benzylpenicillin 2.4 g 2-4 hourly IV +Ciprofloxacin 400mg bd IV +Clindamycin 900mg tds IV (If allergic to penicillin use Ciprofloxacin and Clindamycin only). NB Discuss with local Medical Microbiology Service	

***Must not be used in penicillin anaphylaxis**



<p>Prophylaxis for Recurrent Cellulitis</p> <ul style="list-style-type: none"> • 2 or more episodes at the same site • Penicillin V 250mg bd or Erythromycin 250mg bd for up to 2 years
--

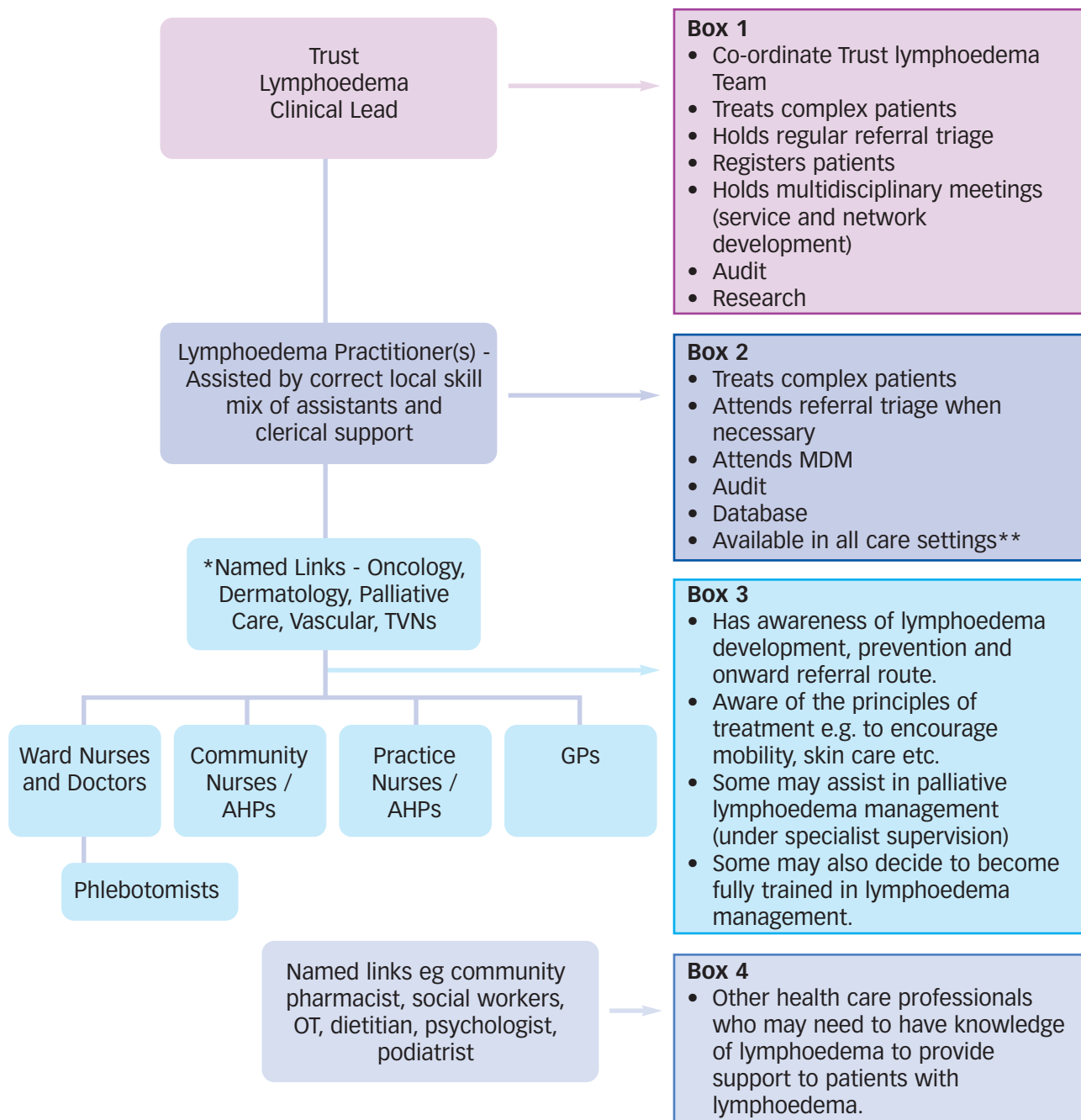
Appendix 8: Suppliers of Lymphoedema Garments

Company Name	Web Address
Activa	www.activahealthcare.co.uk
Haddenham Healthcare	www.hadhealth.com
Jobst Elvarex	www.jobst.com
Juzo	www.juzo.com
Medi	www.mediuk.co.uk
Sigvaris	www.ganzoni.com

Company & Compression Class*	ULmmHg	LLmmHg
Activa		
Class 1	18-21	18-21
Class 2	23-32	23-32
Class 3	N/A	34-46
Haddenham		
Class 1	18-21	18-21
Class 2	23-32	23-32
Class 3	34-46	34-46
Class 4	N/A	49-70
Jobst Elvarex		
Class 1	14-18	18-21
Class 2	20-25	23-32
Class 3	25-30	34-46
Class 4	N/A	49-70
Class 4S	N/A	60-90
Juzo		
Class 1	18-21	18-21
Class 2	23-32	23-32
Class 3	34-43	34-43
Class 4	N/A	49+
Medi		
Class 1	18-21	18-21
Class 2	23-32	23-32
Class 3	34-46	34-46
Class 4	N/A	49+
Sigvaris		
Class 1	12-16	18-21
Class 2	18-22	23-32
Class 3	N/A	34-46
Class 4	N/A	49+

NB * Compression grades (i.e. mm Hg levels describing compression grades) vary from company to company

Appendix 9: Trust Lymphoedema Teams



All named Practitioners should meet for regular multidisciplinary meetings to discuss patient treatment and outcomes.

*Each associated speciality or profession should have a named link to the Trust Lymphoedema Team e.g. oncology, vascular, dermatology, psychologist and dietitian etc.

*Each Trust Lymphoedema Team should have formalised medical links with the associated specialities (vascular/dermatology/oncology and palliative care) to facilitate specific non-routine consultations as required.

**Care settings to include: acute; domiciliary; voluntary and nursing homes

Appendix 10: Lymphoedema Service Referral Form

Patient Details

Name	Referred By:
Address	Date of referral
	Weight:
	Height:
Post Code	BMI
DOB	Chair Bound Y <input type="checkbox"/> N <input type="checkbox"/>
Contact Tel	House Bound Y <input type="checkbox"/> N <input type="checkbox"/>
Hospital/Health and Care No	Transport Required Y <input type="checkbox"/> N <input type="checkbox"/>
GP	<input type="checkbox"/> Chair <input type="checkbox"/> Stretcher

Lymphoedema History

Type:	Primary <input type="checkbox"/>	Secondary <input type="checkbox"/>	Mixed <input type="checkbox"/>
UL/LL/Other:			
Commenced:			
Past Treatment:			
Prioritisation:	Routine <input type="checkbox"/>	Lymphorrhoea <input type="checkbox"/>	Palliative <input type="checkbox"/>

Reason for Referral

- Swelling
- Heaviness
- Pain
- Cellulitis
- h/o cellulitis
- Lymphorrhoea

Duration of symptoms:

Relevant History

(inc family history if appropriate)

Investigations to Date

Lymphoscintigraphy:

Carried out

Ordered

ABPI (if available)

Carried out

Ordered

Lymphoedema Secondary to Cancer:

Cancer Diagnosis:
Surgery:
Chemotherapy:
Radiotherapy:
Active Treatment:
Palliative

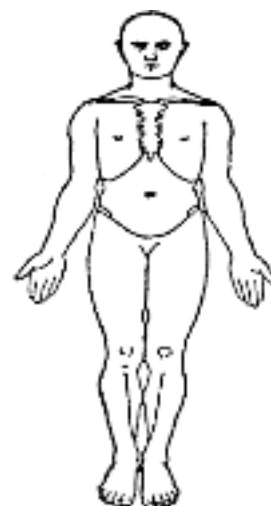
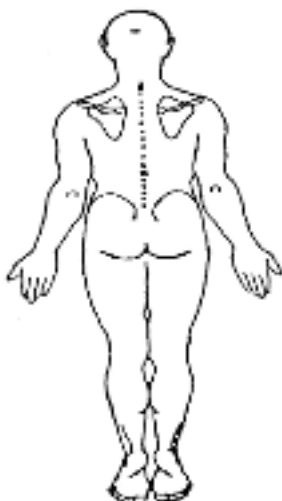
Lymphoedema Secondary to:

Vascular Condition Lipoedema

Skin Condition

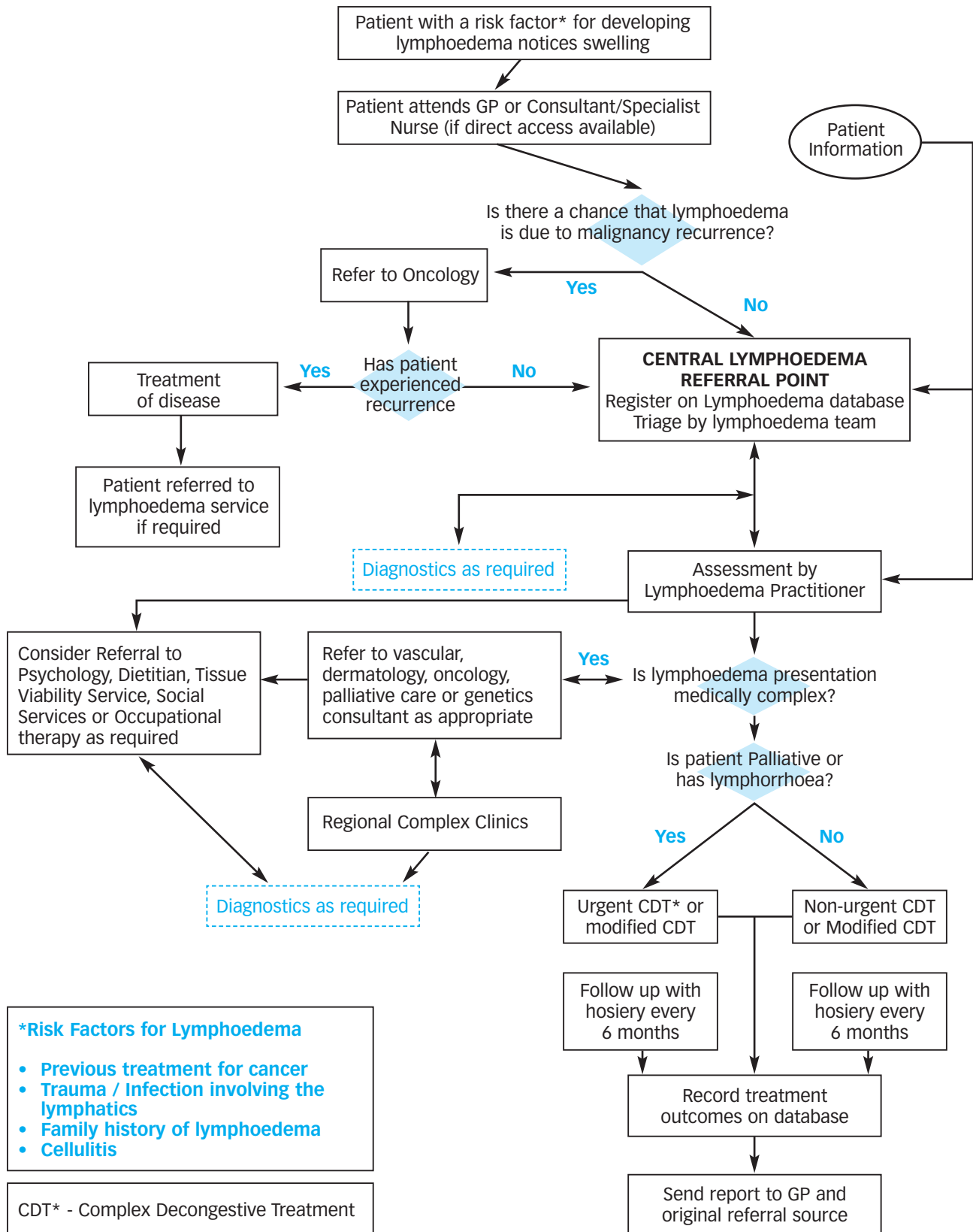
Other:

Indicate Area of Swelling



Medical History:	Yes	No	?		Yes	No	?	Medication:
Hypertension				Venous thrombosis				
Heart Failure				Varicose veins				
Hemiplegia				Chronic renal failure				
PVD / Arterial embolism				Chronic skin disorder				
Phlebitis				Rheumatoid arthritis				
Diabetes				Osteoarthritis				
Obesity				Thyroid				
Allergies				Sleep Apnoea				
Other relevant information:								

Appendix 11: Lymphoedema Referral Pathway - Patient with Identified Risk Factors



Appendix 12 : Criteria for Referral to Complex Regional Lymphoedema Clinics

1. Chronic limb swelling where there is some doubt about the diagnosis of lymphoedema. For example some clinical features of lymphoedema but lymphoscintigraphy is negative.
2. Diagnosis of lymphoedema but patient has complex vascular problems which will effect treatment plans.
3. Diagnosis of lymphoedema but complex dermatological issues which will effect treatment plans.
4. Patients with lymphoedema and complex syndromes on initial diagnosis e.g. Klippel- Trenaunay syndrome or Intestinal Lymphangectasia.
5. Patients with a family history of lymphoedema. (Genetics link).
6. Patients with lymphoedema who are not responding to complex decongestive therapy.
7. Children/young adults with primary lymphoedema on initial diagnosis.
8. Suspected artificial Lymphoedema

NOTES:

Personnel: Vascular surgeon & dermatologist, geneticists, palliative care, local clinical lead for lymphoedema/specialist practitioner.

All patients must have a referral letter from Consultant/Lymph practitioner.

Investigations which may need to be viewed at the clinic:

- Lymphoscintigraphy
- MRI/CT

Equipment required to assess:

- Arterial circulation - ABPI
- Bioimpedance machine



Appendix 13: Routine Lymphoedema Review Form

Name: _____ Therapist: _____

Health & Care No./Other: _____ Date of Assessment: _____

Current Medical History:

Mandatory Questions:

Diabetes Epilepsy MI/Angina Wgt Loss/Gain Previous Surgery
RA Chest Renal Thyroid

Cellulitis Update:

Where are you now functioning for Home activities (H), Work activity (W) and Self Care (SC)?

Unable to function \longrightarrow No limitations of activities

Home Activities
0 _____ 10

Work Activities
0 _____ 10

Self Care Activities
0 _____ 10

Pain Intensity
0 _____ 10
(No Pain) (Worst Pain Imaginable)

Observation of oedematous limb/area (colour, temp, texture etc.)

Weight (Kg): _____ **Height (M):** _____

Bio-impedance reading: _____

Plan

Issue with replacement garments

Ongoing 6 monthly review

Further CDT required

Onward referral required

Please specify _____

Glossary

- **Afferent Lymphatics**

Pre-nodal (that is the ones that enter the lymph node) collecting vessels having smooth muscle in their walls and capable of pumping lymph.

- **Antibody** - A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

- **Antigen** - A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

- **Bariatric** - severe obesity.

- **Bicuspid Valve** - Valve with two flaps or 'cusps'.

- **Bioimpedance** - A newer approach to evaluating tissue changes related to lymphedema is based on measuring or impedance instead of limb volume. Impedance is an electrical property that is similar to resistance but measured using signals of different frequencies.

Within our bodies, fluid can be found within the cells (intracellular fluid) or outside the cells (extracellular fluid). Lymph is a form of extracellular fluid and lymphedema is an excess of extracellular fluid.

Low frequency electrical current travel through the extracellular fluid in the spaces between the cells without penetrating the cell membrane. Higher frequency signals penetrate the cell walls and pass through both intracellular and extracellular fluid. Impedance can be calculated based on current flows at different signal frequencies and used to estimate how much extracellular fluid is present.

Bioimpedance can be measured using a hand-held device and electrodes attached to one foot, each hand and wrist (for upper extremity). The process is fast and painless.

- **Cellulitis:** an infection of the skin and subcutaneous tissues.

- **Complex decongestive therapy (CDT):** the recognised conservative two phased approach to the management of lymphoedema.

- **Compression garments:** are required for life long containment of oedematous limbs:

- 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.

- **Efferent Lymphatics**

Post-nodal (ones leaving the lymph node) muscular collecting vessels similar to the afferent lymphatics but larger and also capable of pumping lymph.

- **Endothelial Cells**

Thin flattened cells found lining blood vessels and lymphatics.

- **Flap Valve** - Overlapping endothelial cells.

- **Hydrostatic Pressure**

The pressure exerted by a fluid at rest. It acts equally in all directions.

- **Initial Lymphatics** – Blind ended sacs with thin walls that consist of a single layer of endothelial cells. These have no muscle in their walls and are not capable of active pumping.
- **International Society of Lymphology** (2003)
- **Interstitial Fluid**
The fluid within the tissues.
- **Interstitial Pressure**
The pressure of the interstitium or tissue spaces. Again, there are two components; interstitial oncotic and interstitial hydrostatic pressure.
- **Interstitial Spaces** - The 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**
The pressure within the lumen (inside) of a vessel.
- **Limb Volume Bioimpedance Method.** Bioimpedance measures tissue resistance to an electrical current, which estimates extra cellular fluid volume.
- **Limb Volume Circumferential Measurements Method** is the most widely used method of calculating limb volume. Measurements are calculated from fixed anatomical points, with repeated 4 cm measurements along the limb.
- **Limb Volume Perometry Method.** This uses an infrared optoelectronic system to measure limb volume.
- **Limb Volume Water Displacement Method.** This method is considered the Gold Standard for calculating limb volume, particularly for hands and feet. It is based on the principle that an object displaces its own volume of water.
- **Lipoedema** can be described as a bilateral, symmetrical, flabby swelling that arises from the deposition of adipose tissue. The cause of Lipoedema is not fully understood. Lipolymphoedema is a combination of lipoedema, obesity and Lymphoedema.
- **Luminal Side**
Inside a lymphatic vessel.
- **Lymphangiomas** - Lymph 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.
- **Lymphorrhoea** - leakage of lymph fluid through the skin surface.
- **Lymphoscintigraphy** is radiological imaging of lymphatics using radioactive tracers (Nanocolloid).
- **Manual lymphatic drainage (MLD):** a specific form of massage to stimulate the lymphatic system.
- **Multilayer lymphoedema bandaging (MLLB):** a specialist bandaging technique used to encourage lymph movement and reduce fibrosis.

- **Oncotic Pressure** Because proteins in blood can't easily cross through the walls of the blood capillary they exert a small osmotic pressure across the wall. This acts to suck fluid back from the tissue spaces into the blood. The hydrostatic pressure in the capillary normally exceeds this oncotic pressure so that net fluid movement is outward.
- **Sentinel lymph node biopsy:** a conservative surgical cancer screening procedure to identify spread of disease to the local primary draining lymph node.
- **Stemmer's Sign (Fig 6.1)** is positive when a thickened skin fold at the dorsum of the fingers or toes cannot be lifted or is difficult to lift. The presence of of this sign is an early diagnostic indication of Lymphoedema. The absence of a Stemmer sign does rule out the possibility of Lymphoedema.
- **Systemic circulation** - Circulation of blood throughout the body through the arteries, capillaries, and veins, which carry oxygenated blood from the left ventricle to various tissues and return venous blood to the right atrium.
- **Pappilomatosis** - a warty appearance of the skin due to fibrosis over dilated lymphatic vessels.
- **Pneumatic compression pump (PCP):** compartmentalised sequential pressure device used in conjunction with CDT.
- **Pulmonary circulation** - The passage of blood from the right ventricle through the pulmonary artery to the lungs and back through the pulmonary veins to the left atrium.





