


Improving the Treatment of Lymphoedema



**Are We Neglecting the Greatest Opportunity to
Promote Quality of Life in Lymphoedema Care?
How Lymphassist Would Help Both Patient and Carers**

**The Lymphatic System, its Organs and its
Connection with Veins and Arteries**

Drugs, Antibiotics and Lymphoedema

**The Complex Causes of Limb and
Joint Swelling and Pain**

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Summary

Foreword

The lymphatic system is a network of tissues and organs that help rid the body of toxins, waste and other unwanted materials. The primary function of the lymphatic system is to transport lymph, a fluid containing infection-fighting white blood cells, throughout the body¹.

WE USE the expression 'soft as a baby's bottom' to express our idea of the ideal softness and suppleness of skin. Skin, as our protective barrier, needs to be moistened and nurtured. Skin under the pressure of oedema hardens. Hardened skin can become scaly and it can crack. Lichen is a type of moss with a distinctive appearance and when skin looks 'lichenified' it is clearly unattractive, but is now also a risk to health as the cracks and fissures allow pathological bacteria to get beneath the epidermis leading to cellulitis. This phenomenon of cellulitis is so common in lymphoedema that we should perhaps manage those with lymphoedema of a limb with the same care as we now advise for those who have diabetes and who need foot care. We live in a world in which we no longer want to bandy about the use of antibiotics indiscriminately. Fortunately, there is guidance with regard to lymphoedema and cellulitis from the British Lymphology Society².

Part of the hardening process in the skin is caused by hyperkeratosis³, a phenomenon you will see with many of your psoriasis patients⁴. Keratin is a substance that causes thickening of the stratum corneum (the horny layer of the skin) with associated thickening of the granular layer.

Clearly it would be better to help your patients before the skin hardens. Pooled water needs a pump system to remove it and the solution for this could be said to have powered the industrial revolution. Traditional coalmines had a problem of pooled water and a steam pump perfected by James Watt in the late 1700s solved that problem. A 'pumping' system can mechanically ensure that the flow of lymph is re-directed to the head and the latest devices allow this to be done in the comfort of your patient's home without the risks of hospital-based infections and complications.

Lymphoedema amongst your patients is relatively common, whether from genetic causes or associated with disease or co-morbidities. Surgery, infection, radiation, drugs, obesity, heart conditions, venous thrombosis and cancer can all adversely affect lymph flow. You have the tools to help patients and improve the quality of their life.

Dr Charles Easmon
Editor

Dr Charles Easmon is a medical doctor with 30 years' experience in the public and private sectors. After qualifying as a physician, he developed his interests in occupational medicine, public health and travel diseases.

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Are We Neglecting the Greatest Opportunity to Promote Quality of Life in Lymphoedema Care? How Lymphassist Would Help Both Patient and Carers

Sylvie Hampton, *Independent Tissue Viability Consultant Nurse.*
Wound Care Consultancy Ltd. Hailsham

Introduction

Lymphoedema is a condition of the lymphatic system and is seen as tissue swelling of body parts, especially the limbs, as a result of reduced lymphatic transportation. This debilitating condition may result in morbidity and immobility, incapacitating and causing economic burdens as well as affects the individual's physiological and psychological wellbeing. (Agbenorku, 2014).

Lymphoedema is certainly fundamentally an under-resourced, and under-recognised condition and its impact is underestimated (Cooper, 2010; Moffatt et.al. 2003) leaving the UK with insufficient lymphoedema services (Morgan and Moffatt, 2007). The prevalence for chronic oedema/lymphoedema is found to be 1.33 per 1000 population, increasing to 5.4 per 1000 population in the >65 age group (Moffatt et.al. 2003).

This lack of recognition of such a serious condition makes lymphoedema, along with tissue viability, "Cinderellas" of healthcare. Lymphoedema, as a chronic condition, requires input from specialists in the assessment, management and specialist treatment (Cherney et al.). Frequent and lengthy appointments are required to dress and bandage the limbs in order to manage chronic exudate levels, (NHS Choices 2016).

Lymphoedema is a chronic, debilitating condition that has traditionally been seen as incurable (Warren, et al. 2007) and that is true. However, quality of life can be greatly improved for sufferers of this chronic problem. Florence Nightingale (Notes on Nursing 1859) spoke a great deal of sense on the issue of healing and

care when she wrote: "The goal of nursing is to place the patient in the best possible condition for nature to act." This suggests that all aspects of care should be considered in order to place the patient in the best possible condition and all therapies investigated in the best interest of the individual.

What is Lymphoedema?



LEG OF DISEASED PATIENT WHO SUFFERS FROM EDEMA

Chronic oedema is due to either a vascular or a lymph problem and has generally been present for more than 3 months and does not reduce when the legs are raised on stool or in bed (Moffatt et al 2003) and chronic oedema can lead to lymphovenous disease.

Lymph fluid is in all body tissues and comes from the tiny blood vessels into the body tissues. Normally it drains fluid and proteins back into the blood stream through channels called lymph vessels which are similar to veins. These are part of the lymphatic system and, when this system stops functioning, this lymph, along with the proteins, will build up in the tissues leading to chronic oedema which can progress to lymphoedema. This leads to an accumulation of

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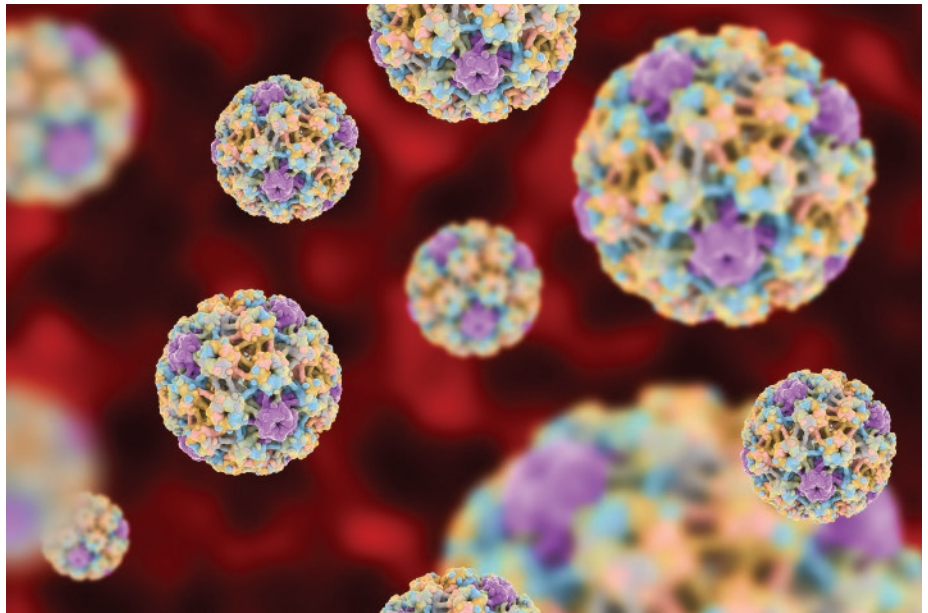
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This condition is not necessarily a disease of old age as it can occur at any age, particularly if it is congenital and is more often seen in females than males



PAPILLOMA

protein rich fluid in the tissues and this excess of protein means that the force of osmosis attracts more fluid into the tissue, creating more swelling, preventing the exchange of necessary oxygen and nutrients to the skin. It then changes the condition of the skin, causing hardening, heat and even papilloma (fig).

This condition is not necessarily a disease of old age as it can occur at any age, particularly if it is congenital and is more often seen in females than males (Dale, 1985). However, the risk to relatives of male patients is about 50% higher than for relatives of females (Dale, 1985).

Lymphoedema can be primary or secondary (Hardy, 2012). Primary is congenital in nature, or related to an abnormal functioning of the lymphatic system due to vascular disease, trauma, immobility and/or infection. Secondary lymphoedema develops as consequential damage or disruption following trauma to the lymphatics either through surgery used to treat cancer, injury, infection, or secondary to other underlying pathologies such as obesity or venous disease (Mortimer, 2013).

Primary lymphoedema is due to genetic abnormalities that cause malformation of the lymphatic system Browse et al (2003) and can be present at birth or develop later in life. Swelling of the limb, caused by the lymph not functioning in the normal way can be exacerbated by obesity and immobility (Ryan 2002; Mortimer, 2013) and although obesity, is not directly involved in how the lymphatic system operates (Scallan et al. 2016), the heavier limbs can act as a reservoir for lymph fluid (Cemal, et al. 2011) and, indirectly, the obesity does have a negative effect on development of lymphoedema. Therefore, controlling weight may help to reduce the amount of lymph that collects in the limb.

There is evidence in one study (Moffatt et al. 2003) that there are less than 64% of patients with lymphoedema receiving treatment with 29% of those developing acute infection in the affected limb being admitted for intravenous antibiotics. However, this was more than a decade ago and we need to believe that more people are being identified with the disease and receiving treatment. Nevertheless, cellulitis is a very real risk for patients with lymphoedema.

Various risk factors have been shown to be associated with cellulitis, with lymphoedema showing the strongest association (Dupuy et al, 1999) and, therefore, it is catch 22. Cellulitis can cause lymphoedema and lymphoedema can lead to cellulitis. Moffatt et al (2003) identified that out of 823 patients, 28% of patients with lymphoedema had had an episode of cellulitis within the previous 12 months (Moffatt et al, 2003). Lymphoedema has also been shown in several studies to be the strongest risk factor for cellulitis (Dupuy et al, 1999).

The Mean length of hospital stay for this condition was 12 days at an estimated mean cost of £2,300 (Bates and Sedgewick 2013). The cost of inpatient lymphoedema treatment in the UK between 2009 and 2010 was £9 million (it would be far higher today) and the cost of tissue viability in the UK (Posnett and Franks 2008) will be well over £3billion today particularly with inflation. Therefore, the NHS should be investigating why so much money is being wasted when expert treatment can improve quality of life and good therapy can greatly reduce costs for the NHS in the long term.

Lymphoedema may affect the limbs, trunk, head and neck, breast or genital areas, and requires life-long management. It can appear at any age, even in the newly born, and results in extensive

physical, functional, psychological and social morbidity, and has a significant impact on quality of life, personal socio-economic status, and the health economy at large. (Cancer Network)

Aetiology of lymphoedema is multifaceted and all of the factors that contribute to the condition and the nature of their interaction have not yet been identified. However, cancer is recognised as being a major contributor to lymphoedema incidence, with the most common cancers involved being breast, melanoma, gynaecological, genito-urinary, sarcoma and head and neck (NHS Wales, 2008).

Lymphoedema is different to Lipodema (see fig) which is an abnormal deposition of fat and is generally a genetic abnormality. Often is sited around the legs or hips and is 'foot sparing' which means that the fat often appears like a pair of trousers on the lower limb. The treatment for lymphoedema, which can be very successful, are not helpful in the case of lipodema as the fluid from lymphoedema can be gently massaged into the system but fat cannot be moved in that way.

Diagnosis of lymphoedema at the earliest possible stage is very important as it is progressive and therefore, early diagnosis can lead to early treatment which can then prevent the progression and lead to a satisfactory outcome.

There are three stages of lymphoedema. The first is the simpler oedema which can show pitting when pressed with a finger or shows deep marks from socks or shoes. If treatment is applied at this point it is possible that it will not progress.

The second stage the skin will be firmer and there may still be some slight pitting although it is more likely there will be an absence of pitting. It may have a spongy feel to the tissue or a hardening – fibroses – of the tissue and an increase in girth.

The third stage is full lymphoedema with extreme hardening of the tissues, grossly enlarged, often known as elephantitis.



DIAGNOSING STEMMER'S SIGN

A simple way of identifying lymphoedema is through the positive Stemmer's sign (fig).

Normally, the thickened fold of skin at the base of the second toe or second finger, can be gently pinched and lifted. With lymphoedema, this skin cannot be pinched and lifted. The skin can only be grasped as a lump of tissue and this is a positive Stemmer's sign for lymphoedema.

In the absence of a cure for lymphoedema, precautions and prevention are emphasized (Petrek et al.2000) and one of those treatments that can aid prevention and treatment is sequential compression therapy.

Sequential Pneumatic Compression

Sequential and intermittent pneumatic compression has formed part of long-term lymphoedema management for several years (Lymphoedema Framework, 2006). Johansson et al. compared manual lymphatic drainage (MLD) with sequential compression therapy in arm lymphoedema in 28 women previously treated for breast cancer. Both treatments were carried out for 2 weeks and arm volume was measured by water displacement. There was no significant difference detected between the two treatment methods and each significantly decreased arm volume. This is an important finding as it can have great impact on lymphoedema services when manual lymphatic drainage is taking up clinicians' valuable time.

The treatment of lymphoedema with sequential compression therapy and compression stockings is associated with long-term maintenance of reduced limb girth in 90% of patients (Pappas, and O'Donnell, 1992). Chang and Cormier, (2013) identified that exercise and intermittent pneumatic compression are effective therapies and can be safely implemented in appropriate patients as an adjunct to complete decongestive therapy. Lymphoedema, if left untreated, or is treated inadequately treated, then the outcome can be irreversible and potential of cellulitis is very high which can lead to death. Therefore, it is vital to begin therapy as soon as possible to reduce the progress. LymphAssist™ is perfectly placed to offer a simple and cost effective treatment.

LymphAssist™

LymphAssist™ provides a sequential milking pattern to the limb through multiple compartments along the length of the garment. This is a method of returning the lymph to the main system.

LymphAssist™ Professional is a 12 Chamber Intermittent Pneumatic Compression System (fig) that was developed with advice and input from specialists in Manual Lymphatic Drainage (MLD), Lymphoedema specialists and Tissue Viability nurses.

The LymphAssist™ Professional system is able to complement MLD and compression

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The beauty of LymphAssist™ is that it not only can optimise the patient's condition, but it empowers them to attend to their own needs as there is a carer's LymphAssist™ that can be applied by the individual or by their carers



bandaging therapy and would be part of a care package that would be required to reverse some of the oedema that causes such devastation to the lives of sufferers. It would also reduce nursing time and offer the patient quality of life that is missing when care has to be so intense as in lymphoedema.

The system comprises a pump and a range of multi chamber arm or leg garments capable of delivering three types of treatment options and the action of the chambers replicates the These three options are:

1. LymphAssist™ Therapy which mimics the basic principles of MLD.
2. Graduated Sequential Therapy which provides a controlled pressure gradient throughout the different chambers.
3. Wave Therapy which produces a gentle peristaltic cycle for those patients who may be unable to tolerate graduated sequential compression.

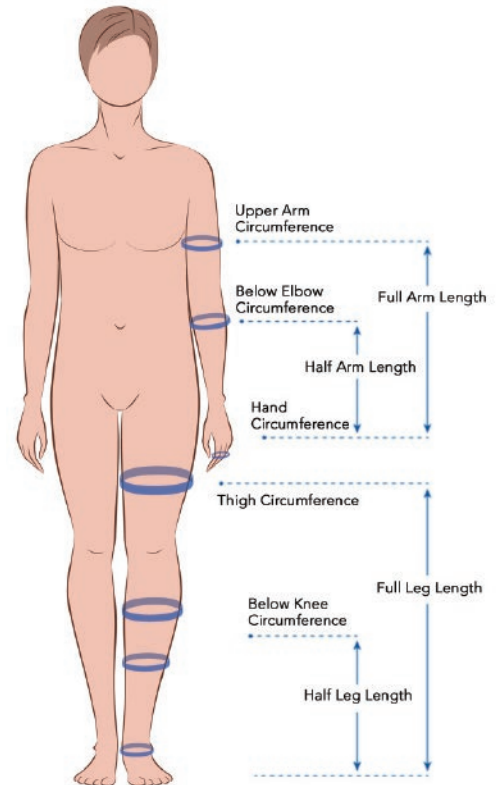
The beauty of LymphAssist™ is that it not only can optimise the patient's condition, but it empowers them to attend to their own needs as there is a carer's LymphAssist™ that can be applied by the individual or by their carers. This would be applied two or three times a day for 35 minutes during which they can sit comfortably, watching TV or reading etc.

Between the treatments with LymphAssist™ the patient would wear a compression garment and continue their activities of daily living and social activities.

If limbs are reduced in girth, then there is less need for bandaging and hosiery can be applied. Also, as the fact that the patient and/or carer can apply the garment without nursing input, then the cost savings in bandage cost and nursing time could be very great, especially as nursing input of MLD and or bandaging takes up great periods of time and many bandages.

A further cost would be the reduction in potential of cellulitis that is so closely linked with lymphoedema.

Measuring for a LymphAssist™ Garment



Selecting the correct size of garment for the individual patient is quite simple.

Any bandage, hosiery or bulky dressings should be removed and the points shown in fig 9 should be measured and recorded.

The measuring tape should be applied without pulling it tightly.

It is important that the measurement for length goes beyond the area that requires treatment.

Education of the Patient

It is vital that the patient has an understanding of why the treatment is required and why it should be consistently used. One they see the difference that occurs when the treatment results in smaller and lighter limbs then concordance is far more likely.

Conclusion

LymphAssist™ provides an enhancement of the therapeutic response particularly when used with compression therapy such as hosiery. The availability and ease of use of LymphAssist™ will reduce nursing time and release the patient from the constant need for nursing care. They would then be lighter in weight as the limbs reduce in size and would be more likely to return to their normal life style.

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The Lymphatic System, its Organs and its Connection with Veins and Arteries

Joanne Greenwood, Medical Correspondent

Lymphatic damage is not as deadly as arterial or venous damage but it is disabling and disfiguring and can significantly reduce quality of life

IN THE Western world we owe the discovery of the circulatory system to William Harvey¹ but who should be credited for the lymphatic system, which is often not discussed? We are an outline of veins, arteries and lymph vessels and all three interact with the rest of the body. Since the three vesicular systems carry fluid, a disruption in any one can lead to abnormal swelling of the soft tissues of cells, muscles and ligaments, which are structured around bone. If an artery bursts the effects are seen quickly and need to be remedied early before they lead to death or disability. An artery bursting in the enclosed area of the brain leads to brain ischaemia and anywhere else in the body because the cardiac output is almost 5 litres² per minute in adults, within 5 minutes a person can exsanguinate and it only takes 4 minutes of lack of supply to the brain³ to cause irreparable brain damage⁴.

If the veins become enlarged then varicose veins become visible. If they become blocked then a thrombosis develops which may be fatal. If they leak then swelling and local tissue damage occur.

Lymphatic damage is not as deadly as arterial or venous damage but it is disabling and disfiguring and can significantly reduce quality of life.

We know that we are more than 50% fluid⁵. We are aware about blood, whether venous or arterial. Diagrams of the arterio-venous system depict arterial (oxygenated) blood as red and venous (de-oxygenated) blood as blue and both systems give us an outline of ourselves, which we can easily imagine super-imposed on Leonardo Da Vinci's Vitruvian Man⁶. The vessels of the lymphatic system can also be super-imposed on this but in an almost truncated version. The lymphatic system additionally carries the protein rich fluid, lymph, and has its own organs of supply and function.

The drainage system of the lymph differs from the arterio-venous system in terms of flow and symmetry. The lymph flows only one way

and that is towards the head. On both sides of the body the lymph drains into the right or left brachiocephalic vein but in an asymmetric manner. Hence, on the right side the drainage into the right brachiocephalic vein is from the right lymphatic duct and is only draining the lymphatic vessels of the right arm and the right side of the head, neck and thorax (the right arm and right upper part of the body), all the rest drains into the left brachiocephalic via the thoracic duct which connects the lymphatic vessels of the abdomen, both legs and the left side of the head, neck, and thorax.

Lymph Is Major Part of The Body's Immune Defence System

It is easy to confuse, lymph, plasma and chyle. The word 'lymph' derives 'from the Latin word *lymphā*, which means "connected to water⁷.'" Plasma is fluid that has left the body's cells having delivered its nutrients and removed debris. Most of this plasma returns to the venous circulation through venules to continue its journey as venous blood. The remainder becomes lymph⁸.

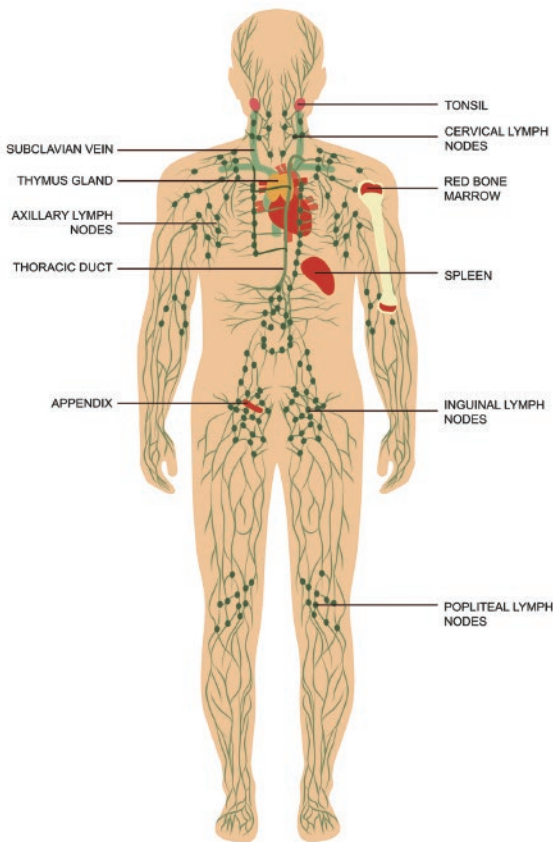
Therefore lymph, which derives mostly from the fluid between cells (interstitial) is a white or clear colour and contains white blood cells (mainly lymphocytes) and from the intestinal area it acquires protein and fats (known as 'chyle', triglycerides are absorbed by the intestinal villi to create this). Lymph itself is estimated to be about '90% water and 10% solutes such as proteins, cellular waste products, dissolved gases, and hormones⁹'. In pathological situations the lymph contains bacteria and cancer cells and can be a means of propagation of infection and metastatic cancer. Its organs include the lymph nodes, ducts, the tonsils, the adenoids and the spleen.

The Lymphatic Organs and Tissues

These include:

- 1) The lymph nodes. The number of these in the body is estimated to be between 6-700.

LYMPHATIC SYSTEM



Each node has a capsule and has a vessel that takes lymph to them (afferent) and vessels that takes lymph from them (efferent). The soft, small, round or bean-shaped lymph nodes may be superficial or deep. They are located in the neck, armpit, groin and inside the centre of the chest and abdomen. They make the immune cells that help the body fight infection. They act to filter the lymph fluid and also to remove foreign material such as bacteria and cancer cells. Once bacteria are recognized in the lymph fluid, the lymph nodes make more infection-fighting white blood cells. This causes the nodes to enlarge and we can sometimes feel the swollen superficial lymph nodes in the neck, under the arms, and groin¹⁰.

2) The Spleen, which is the largest lymphatic organ. This can be damaged by trauma or disease. It helps fight infection by filtering bacteria and viruses and its functions includes both lymph, lymphocyte and blood production in that it controls both the amount of red cells and blood storage in the body. If the spleen is removed or damaged, then encapsulated bacteria become a problem hence the advice to protect by vaccination against Meningitis, Haemophilus and

Pneumonia¹¹. Sickle cell disease leads to infarction and hyposplenism¹² (functional splenic removal). A spleen damaged by trauma can lead to a rapid death just from the volume and speed of the blood loss.

3) The tonsils are large clusters of lymphatic cells found in the pharynx at the base of the tongue and posteriorly¹³. We have tonsil tissue at the posterior root of our tongue (lingual) and the ones we see enlarged when we look at the back of the throat with a torch and spatula (palatine). The American Academy of Otolaryngology – Head and Neck Surgery, describes the tonsils as the body's 'first line of defense'¹⁴ as part of the immune system in that they sample bacteria and viruses that enter the body through the mouth or nose. This part of the immune system was abused for years within most of our lifetimes by a lack of evidence-based surgical removal (tonsillectomy¹⁵) which fortunately has stopped as a routine procedure. However, a full-blown peritonsillar abscess or Quinsy¹⁶ may need post infective surgical removal.

4) The adenoids 'are glands at the back of the nose, where it meets the back of the throat. The adenoids are also called the nasopharyngeal tonsils'¹⁷.

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The lymph system is complex with organs and vessels which are all a key part of our immune system

The tonsils and the adenoids can be regarded as guardians at the doorways to the gut and the lungs, which protect the body from viruses and bacteria.

- 5) The Thymus gland. This organ in the throat area is where T-cells mature. The organ can be affected by autoimmune diseases and has links with myasthenia gravis¹⁸. It is important to be aware about malfunction of the Thymus because it is a contraindication to giving the yellow fever vaccine¹⁹.
- 6) Peyer's patches are named after the Swiss pathologist Johann Conrad Peyer²⁰ who described them in the 1600s. These are small masses of lymphatic tissue found in the ileum of the small intestine²¹. They are

deadly in the disease Typhoid since our inflamed immune response to the bacillus can cause perforations in the gut wall that then lead to death.

Summary

The lymph system is complex with organs and vessels which are all a key part of our immune system. Malfunction can occur at many different stages, some visible (superficial lymphadenopathy, lymphoedema) and some invisible (thymus malfunction, Peyer's patch rupture, deeper lymphadenopathy). It deserves our caution and attention as much as the venous and arterial systems that we think we know so well.

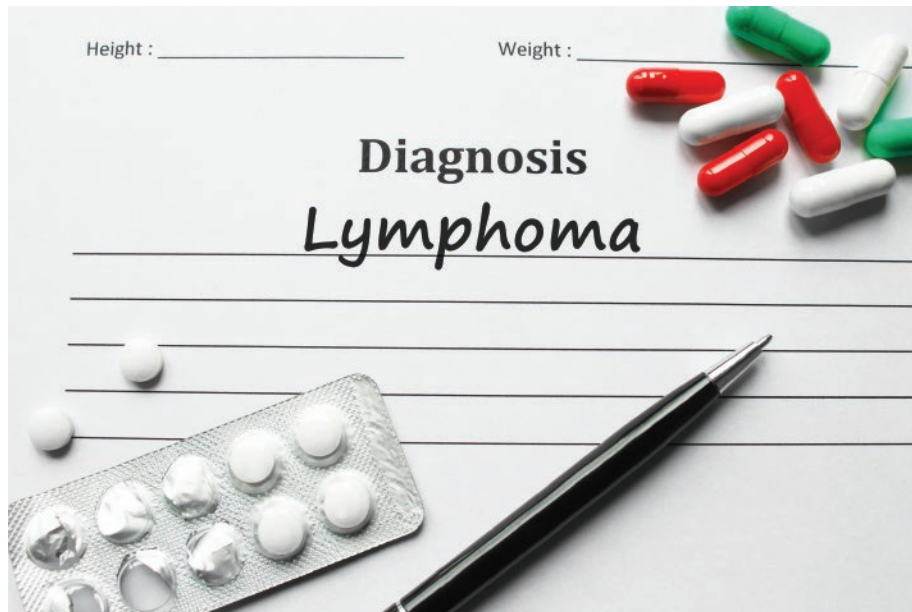
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Drugs, Antibiotics and Lymphoedema

Dr Charles Easmon, Editor

Drugs have good, bad and ugly sides and sometimes they can be all three at the same time in your lymphoedema patient.



MANY DRUGS may cause peripheral oedema or exacerbate existing chronic oedema¹. Drug causes or aggravators of lymphoedema are surprisingly common. When the researcher Vaughan Keeley investigated Summaries of Product Characteristics (SPCs) he found at least 900 drugs reporting oedema side-effects (admittedly some pulmonary oedema) and over 300 drugs in the the adverse reactions sections of Micromedex² (2007). The frequency and severity of oedema caused by drugs is not data readily available and it would need expensive post-marketing surveillance to really appreciate these data. However, the most common ones for a General Practitioner to consider are: calcium channel antagonists; corticosteroids; nonsteroidal anti-inflammatories (NSAIDs); alpha-blockers and sex hormones³ as detailed further below.

The Impact of Drugs

Drugs can increase capillary filtration by increasing the hydrostatic pressure in the capillaries and this might be by 1) increasing blood volume and fluid retention (examples include non-steroidal anti-inflammatory drugs [NSAIDs]), corticosteroids,

and other hormones) or 2) by causing peripheral arteriolar vasodilation (examples include alpha blockers, calcium channel blockers). This effect of drugs can occur in patients with normally functioning lymphatics.

In those patients with existing lymphoedema, the drugs that cause increased capillary filtration will create a 'negative loop' and overload the failing lymphatics even further, causing increased oedema.

The problem for the clinician is when the 'oedema' unwanted effect is a by-product of the mechanism of action required to treat a particular condition as in the use of alpha blockers to reduce hypertension. If necessary, attempts at adjusting the dose to ensure clinical efficacy versus minimal side-effects may be the only practical solution.

The most commonly used drugs that we know cause or contribute to oedema include:

- 1) Calcium channel blockers, which are frequently prescribed and are the first choice of hypertension therapy for black persons of African or Caribbean origin of any age and those over the age of 55⁴. In a dose-dependent manner they frequently cause ankle oedema via the mechanism of peripheral arteriolar vasodilation. This effect may be transient.

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The problem for the clinician is when the 'oedema' unwanted effect is a by-product of the mechanism of action required to treat a particular condition as in the use of alpha blockers to reduce hypertension

Up to 50% of lymphoedema patients experience at least one attack of cellulitis in their lifetime⁵.

- 2) Corticosteroids (e.g. prednisolone, dexamethasone) as used for many conditions have a mineral corticoid effect and lead to peripheral oedema by sodium and water retention and it is observed that the degree of swelling depends upon the dose and duration of treatment.
- 3) Sex hormones and related hormones may cause oedema by fluid retention.

Antibiotics and Lymphoedema

Up to 50% of lymphoedema patients experience at least one attack of cellulitis in their lifetime .

Cellulitis is frighteningly common in lymphoedema and this is not surprising since the skin barrier becomes hardened and cracked allowing pathogenic bacteria to enter

below the epidermis and multiply. Fortunately, The British Lymphology Society Consensus statement of 2016 on Cellulitis gives clear guidance as to when to advise hospitals, how to monitor, when to change antibiotics, how to deal with penicillin allergies and which patients to consider for prophylactic antibiotics⁶.

Summary

Always review your lymphoedema patient's drug history carefully since it may be the cause or the reason the condition is getting worse. Follow the British Lymphology Society's guidance on the sensible use of antibiotics in your lymphoedema patients with cellulitis and use the same to assess the need to prevent recurrences.

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Cellulitis is frighteningly common in lymphoedema and this is not surprising since the skin barrier becomes hardened and cracked allowing pathogenic bacteria to enter below the epidermis and multiply

The Complex Causes of Limb and Joint Swelling and Pain

Joanne Greenwood, Medical Correspondent

Is my swelling obesity, my veins, arthritis, my kidneys or lymphoedema, Doctor, and what can be done about it?

Chronic oedema is extremely common in severely obese patients. Sometimes it has the features of lymphoedema with typical skin changes... whereas in others there may be a clear pattern of venous disease. In severely obese patients, immobility may contribute to the picture¹.

THE PATIENT sits in your surgery with their leg or arm up. It is clearly swollen and they expect you to do something about it but you need diagnostic and management help. They complain not just of the swelling but also the feeling of fullness or heaviness, tightness and stretching of the skin, reduced movement of the joints, thickening and dryness of the skin, discomfort and/or pain², clothing, shoes or jewellery (i.e. rings or watches) feeling tighter than usual and, in addition, they may have anxiety and a sense of malaise.

Examine the Patient and the Records

Most frequently the answer is on your record card. If the female patient has had a mastectomy and the swelling is in the arm of the affected side and with breast cancer being the most common cancer in the UK you can expect to see several cases in your surgery in your working life time.

As with many clinical situations, from the history and the examination you can gather a lot of information before the need to do any tests.

You look at the patient, they are obese and you ask yourself might this itself be the cause? Well, it might be because of the abnormal hydrostatic pressures created in the lower limbs if the problem is there. Similarly, if they have a mobility problem caused by a co-morbidity condition such as a stroke, this may be the cause of the swelling.



If they are slim and young, you should enquire if there is any family history to indicate an autosomal dominant hereditary cause. These are relatively rare.

You look at the distribution of the swelling and, if it is unilateral, this might suggest an obstruction, trauma, surgery or a possible post-infective or post-thrombotic cause on the affected side. There may be an associated history of deep vein

Following the removal of lymph nodes and radiation therapy, Secondary Lymphoedema or swelling of the arm affects about 25% of breast cancer patients².

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As with many clinical situations, from the history and the examination you can gather a lot of information before the need to do any tests

thrombosis, cellulitis in the affected limb, breast surgery or traumatic damage to the lymph vessels in the affected area.

You note that the swelling is symmetrical and you try Stemmer's test⁴ and it is positive. You consider drugs as possible causes or aggravators of the lymphoedema. You know that more than 900 drugs can cause this but the main culprits are calcium channel blockers, Non-Steroidal Anti-inflammatory drugs (NSAIDs) and of course steroids themselves.

Your urine test for protein rules out nephrotic syndrome. Your basic blood tests of Full Blood Count, Liver, Kidney and cholesterol levels add no useful new information.

Having considered primary lymphoedema, then secondary you have ruled out obesity as the main cause and have a working diagnosis based on some of the options considered above, but how now to treat and manage?

Treatment and Management of Lymphoedema

Since swelling indicates trapped fluid, simple logic would suggest the use of diuretics but sadly these do not work in lymphoedema⁵ since you are dealing with compartmentalised lymph and making your poor patient pass urine more frequently is not the solution.

Skin care and antibiotics (correctly used) are both important and these are discussed in another article in this series.

Pump-action and Manual Lymphatic Drainage (MLD)

The latest American President is quoted as saying that he would 'drain the swamp' of Congress of political elites. Drainage in lymphoedema is important and this can be done manually or by power assisted devices.

A utilitarian pump designed by Scotsman, James Watt^{6,7}, in the 1700s to pump water out of mine shafts can be said to have started the industrial revolution and to have been one of the first truly useful products of the steam age. From that utility we ended up with pump-action firearms openly carried in Texas, which voted in such a law in 2015⁸.

On a more positive note, pumps apply or provide intermittent pressure, which can propel items, especially fluids from point A to point B and this action can and is used to move lymphatic fluid from where it has collected back into the body circulation.

A human version of the 'pumping' action is part of the process in manual lymphatic drainage⁹ (MLD) but this advanced process and procedure must ideally be conducted by someone trained in it so as not to damage delicate skin, risk infections or complications. The origin of this technique is credited to Dr Emil Vodder¹⁰ in the 1930s.

Summary

Problems of lymphatic drainage causing lymphoedema are most frequently secondary rather than primary (genetic). The primary (genetic) causes are mostly single gene defects and autosomal dominant so a good history is important when assessing your patient.

Within the 'secondary' group can be included causes such as obesity, venous problems (post Deep Vein Thrombosis, DVT and varicose veins), heart failure, immobility and advanced cancers¹¹. Iatrogenic causes of secondary lymphoedema include surgery and radiotherapy. Infection (some, of course, iatrogenic) and trauma can be other causes that disrupt this delicate system.

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Lymphatic Filiriasis

Dr Charles Easmon, Editor

ANYONE WHO has a domestic drain blocked by hairs of grease knows how frustrating it is to watch as the dirty water fails to drain after the plug is pulled. A solution is required to degrease pipes and drains usually in the form of some highly toxic substance or a very costly man/woman with a rod. Blockage of the lymphatics is a plumbing problem without a suitable de-cluttering solution. We cannot send a miniaturised Raquel Welch and her team, as in the *Fantastic Voyage*¹ film, to unblock our systems and we have no non-toxic substance to flush through the lymphatics and, even if we had such, the route of flow would make this challenging (towards the neck only). However, we can pump excess fluid back into and through the lymphatics into the relevant veins through mechanical devices designed to do just that.

We can be thankful that we do not live in a part of the world where an unfriendly mosquito can lay a special type of egg on our skin. That special egg then develops into tiny worms, which burrow into our skin and, from there, into our lymphatic system and then rather unhelpfully block it causing chronic and in cases massive swelling of the limbs or genitals affected (as happens in tropical lymphatic filiriasis).

The prospect of filiriasis spreading is concerning enough that some countries such as Kuwait insist on screening for it in all work visa applications².

Worms in the Lymphatics

Lymphatic filiriasis is species of filarial round worms (nematodes) blocking lymphatics. In parts of Asia the worms are *Brugia malayi*³ and *Brugia timori*⁴. In other parts of the world the worms are *Wuchereria bancrofti*⁵ (the cause of an estimated 90% of all infections). The mosquito carriers for these worms can vary from Anopheles mosquitoes in rural environments, especially in Africa and these, of course, are the same mosquitoes that carry malaria as part of their deadly or disabling cargo. In Pacific areas, the mosquito carrier tends to be the white-banded (look at the legs) *Aedes* mosquitoes. In the increasing urban and semi-urban areas of the world affected, the mosquito carrier is *Culex* mosquitoes. A significant

percentage (40%) of those whose lymphatics are damaged also get kidney damage from the same worms.

The early stages of infection may include attacks of fever, characterised by episodic attacks of malaise with chills alongside painful lymph nodes.

The long-term effects are nauseatingly disfiguring causing elephantiasis with swelling of the legs and genitals in both sexes and, more specifically, enormous testicular swellings in men and huge increases in breast size in women.

Epidemiology of Lymphatic Filiriasis

Tropical areas of Africa, Asia and the Americas as well as the Pacific, as previously stated are affected. Worryingly, about 1.4 billion people are estimated to live in areas where Lymphatic Filiriasis is actively transmitted⁷. The Lymphatic Filiriasis problem is endemic in 83 countries (over a third of the world's countries) and it has infected over 120 million people. An estimated two-thirds of the world's infections are clustered in South-East Asia with Africa taking up a third of the burden. The World Bank⁸ devised a composite measure for disability called the Disability Adjusted Life Year (DALY) where 'One DALY can be thought of as one lost year of "healthy" life⁹'. Lymphatic Filiriasis has been estimated to cause 2.8 million DALYs¹⁰.

Public Health Prevention, Treatment and Management for Lymphatic Filiriasis requires at least 'five rounds of annual treatment using a combination of albendazole and ivermectin at 100% geographic coverage and achieving effective treatment coverage of over 65% during each round¹¹'.

These programmes of Mass Drug Administration (MDA) have reached over 900 million people in the last 17 years and are estimated to have reduced the global prevalence of infection by 30%¹².

Studies have shown that, even after all disease seems to be eradicated, the worms can still be present in a sub-population, which then provide the fodder for more mosquito infections.

Lymphatic Filiriasis is regarded as one of the neglected tropical daises and it has been targeted for global elimination (the Global Programme to Eliminate Lymphatic Filiriasis¹³

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The prospect of filariasis spreading is concerning enough that some countries such as Kuwait insist on screening for it in all work visa applications



was launched in 2000). Elimination means just that and is different from eradication¹⁴, since it is about stopping the spread of a disease rather than completely annihilating a microbe or organism from the world¹⁵.

Diagnostic Problems and Night Owls

If you look for something that is not there you will not find it although you might find the trace of its presence. Perversely, the worms themselves (the *Microfilariae* which are the small offspring of the adult worms) only come out at night! No one who went into pathology chose the speciality so that they would have to take blood tests in the evenings but, in developing nations, this is actually, what

they have to do (for intensity of infection) unless an antigen test (for diagnosis only) is used instead (an immunochromatographic rapid test or ICT¹⁶).

Summary

We in the United Kingdom are lucky. We have the mosquitoes but not the toxic payload they carry that blocks lymphatics. About a quarter of the world's population are not so lucky and they need early and multiple mass drug administration to kill the round worms and, if symptoms of swelling develop, they may benefit from early intervention with mechanical pumping devices but this would need a proper research trial to ensure that the worms entering the venous system would not cause further damage there.

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