

**Lymphoedema** results from a failure of the lymphatic system. Consequences are swelling, skin and tissue changes and predisposition to infection. It most commonly affects the lower or upper limbs, but may also affect midline structures such as the head and neck, trunk, breasts or genitalia.

**Oedema** results from an imbalance between capillary filtration into, and lymphatic drainage from, the interstitial space.

**'Chronic Oedema'** is a term used to describe a group of conditions characterised by the presence of swelling within tissues of the body, caused by the accumulation of excess fluid within the interstitial space of the affected area and lasting more than 3 months. It is often used interchangeably with the term 'Lymphoedema'. Although the term 'lymphoedema' suggests that the oedema is caused by a lymphatic abnormality, in every case of chronic oedema there will be some impairment of lymphatic drainage, either through an underlying abnormality ('primary' or 'secondary') of the lymphatic system, or through 'lymphatic failure' as a result of the capacity of the lymphatics being overloaded.

**LYMPHOEDEMA MAY BE PRIMARY OR SECONDARY**

In Primary Lymphoedema there is an abnormality of the lymphatic system which has been present from birth. In some people it will be evident at birth, but in others it may not be identified until later in life, possibly after an event that has caused further damage to the system, such as an injury, infection or surgical procedure. Some primary lymphoedemas are hereditary, e.g. Milroy's Disease. There are a number of genes associated with syndromes in which lymphoedema features, offering some hope of gene therapy in the future.

Secondary Lymphoedema arises as a result of damage to a normally functioning lymphatic system. Such damage may arise from surgery, trauma, radiotherapy or infection in areas that are rich in nodes and vessels, e.g. the axilla, groin, pelvis and around joints, risk being increased if there are a combination of these factors. Damage to vessels may have a more insidious onset as a consequence of obesity, venous disease, obesity, lack of activity, advanced stages of various conditions or lipoedema.

**PREVALENCE OF LYMPHOEDEMA**

A study in 2013 reported a prevalence of 3.93 per 1000 (Moffat et al 2016). However, data from referrals to the national lymphoedema service in Wales reveals a

prevalence of 6 per 1000. This suggests lymphoedema affects between 200,000 and 420,000 people of all ages in the UK.

**LYMPHATIC FUNCTIONS AND IMPLICATIONS OF FAILURE**

The lymphatic system has 3 main functions which are disrupted when the system is compromised and give rise to particular clinical features and complications that differentiate it from other forms of oedema as shown in the table below.

Function	Implication of failure
1. Maintenance of fluid balance by collection and transport of fluid from tissue spaces back to the circulation	Peripheral swelling which does not lessen overnight or with elevation.
2. Nutrition by fat absorption from the gut and transport back to the circulation	Swelling may be soft and pitting initially. Over time, and without treatment, the interstitial fluid is replaced by fibrosis and adipose tissue, which is unevenly distributed and pitting is difficult or impossible. Subcutaneous tissues become thickened and increasingly firm. Deep folds develop, e.g. at the ankle. Appropriate treatment will minimise common complications:
3. Immunity and defence by removal of dying or mutant cells, transporting antigens and immune cells, generating immune responses to infection and malignant cell antigens	<ul style="list-style-type: none"> <li>• predisposition to cellulitis, which in turns further damages the superficial lymphatic vessels.</li> <li>• skin becoming dry and cracked as cellular particles that would normally be removed from the interstitial spaces draw moisture from the skin.</li> <li>• lymph blisters developing, becoming fibrosed over time, giving a 'warty' appearance.</li> <li>• areas of lymph leakage or lymphorrhoea through cracks in the skin.</li> </ul>

# DEVELOPMENT AND PROGRESSION OF LYMPHOEDEMA

The physiological changes that occur over time as a consequence of lymphatic failure mean that early recognition and intervention is always easier and more effective, than interventions initiated at later stages. The International Society of Lymphology (ISL) has identified the following stages of lymphoedema based on the relative “softness” or “firmness” of the limb (reflecting fibrotic soft tissue changes) and the outcome after elevation (see right).

Stage	Characteristics
0 (latent/pre-clinical/at risk)	Swelling not present despite impaired lymph transport.
I	Early presentation with visible swelling that is soft and pitting and may subside with elevation.
II	Increased swelling and elevation alone rarely reduces oedema. Tissues becoming firm with pitting only possible with strong sustained pressure.
III (lymphostatic elephantiasis)	Severe swelling with changes in skin and tissue texture. Tissues increasingly fibrotic, no pitting. Deep skin folds. May be hyperkeratosis (thickening of skin) and /or papillomatosis (fibrosed lymph blisters).

## References

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### About the British Lymphology Society (BLS)

The BLS is a membership charity run by and for its members who share its mission:

To actively promote professional standards and the study, understanding and treatment of lymphoedema/ chronic oedema.

Through support of its membership, the Society seeks to achieve high standards of care and equitable access to treatment across the UK, raise awareness of the condition, promote early detection and intervention with supported self management. We work with other stakeholders, advise government, NHS and other professional bodies and organisations to effect change and influence practice.

See [www.thebls.com](http://www.thebls.com) for helpful resources and the benefits of membership.

### About Lymph Facts

Lymph Facts are a series of documents produced, reviewed and monitored by BLS Members. Please feel free to use these to support your education/ awareness raising activities. Every effort is made to ensure the content of Lymph Facts is accurate, up-to-date and appropriately acknowledged or referenced. We would be very grateful to receive feedback on anything that seems inappropriate or incorrect. Please see the website for the full range of Lymph Facts available. We would also welcome offers of contributions to extend the range of Lymph Facts.

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