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WHAT IS Lymphology?

Lymphology is still a closed book to many. Lymphology is a brand of medical science concerning disorders related to the lymphatic vessels and nodes. It also covers very rare diseases affecting fat tissues such as lipedema, lipolymphedema, and filariasis.

The ones who suffer from the lack of awareness about the various lymphatic diseases are always the patients, who remain uncared for or inadequately managed. This booklet is designed to promote an understanding of lymphology among care providers, and thereby, further improve the level of care.



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Acknowledgement:

A special word of thanks to the patients and staff of the Földi Klinik Hinterzarten (Germany) for their outstanding willingness to provide assistance and support.

Introduction

The blood circulation starts to form in the initial phases of embryonic development and subsequently becomes differentiated into the complex circulatory system. This comprises two systems:

- the closed blood circulation consisting of veins and arteries and
- the half-open lymphatic system which, in addition to its known immunological functions, also has important circulatory functions.

Disorders of lymph flow often lead to serious diseases. The most frequent of these is lymphedema, which affects mainly the extremities.

This booklet presents, using simplified examples, current knowledge of:

- the functional anatomy and pathophysiology of the lymphatic system,
- · lymphedema as a disease state and
- · its management.

Functional anatomy

The lymphatic system can be divided into three functional parts:

- 1. lymph production in the initial lymph vessels,
- 2. lymph transport through the lymph collectors, and
- 3. lymph concentration and filtration in the lymph nodes.

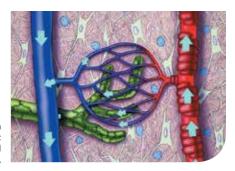




Lymph Production

The supply of lymph to, and its removal from, various body cells and lymph production take place throughout the body's microcirculation according to the same basic principle.

Lymph production takes place in the terminal vessels. Their most important elements are the blood capillaries and initial lymph vessels.



- The blood capillaries are the most important part of microcirculation.
- The interstitial tissue consists of amorphous ground substance, fibers, waterbinding proteoglycans and connective tissue cells, especially fibroblasts. It is permeated by prelymphatic channels.
- Between the blood capillaries are the open-ended initial lymph vessels.

Blood capillaries, interstitial tissue and initial lymph vessels form a functional unit.



Lymph is produced from tissue fluid which flows through the interendothelial gaps into the initial lymph vessel.

The initial lymph vessels form the beginning of the lymphatic system. They are secured to the elastic fibers by anchor filaments.

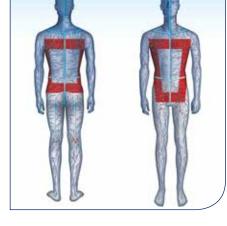
Lymph is produced from tissue fluid which flows through the open interendothelial junctions into the initial lymph vessel.

The initial lymph vessels not only have transport functions - their endothelial cells can also actively phagocytize corpuscular particles. Through various vasoactive substances secreted by their endothelial cells, they also control the tone of the precapillary arterioles and thereby actively regulate the level of ultrafiltration and absorption.

In the functional unit blood capillary/interstitial tissue/initial lymph vessel, extravasation and removal of fluid are usually in a state of equilibrium.

Lymph transport

The initial lymph vessels make up a superficial, valveless network extending throughout the entire body. The lymph is transported from these vessels to the larger, deeper lymph vessels, the lymph collectors.

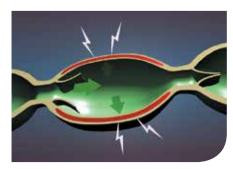


The lymph territories are separated from each other by "watersheds".

The lymph collectors run to the regional lymph nodes. This gives rise to different lymph territories separated from each other by lymphatic watersheds.

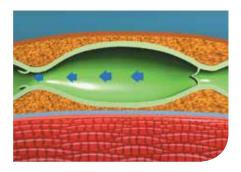
The "driving force" for lymph transport is provided by the lymphangions. These are lymphatic vessel segments bounded by two valves making them contract like small hearts.

Lymphangiomotoricity is controlled mainly by their filling state.



The lymph vessels - like the veins - are equipped with valves. These valves determine the direction of flow. The lymph vessel segment bounded by two valves is known as a "lymphangion" or "lymph pump". Each lymphangion has innervated powerful smooth muscle that makes it pulsate like a small heart. In this way the lymph is transported from one lymphangion to the next.

Lymphangiomotoricity is controlled firstly by the filling state of the vessel. Secondly, the "lymph heart" is now known to have autonomic regulatory mechanisms based on the release of signalling substances. These mechanisms influence both lymph production and the microcirculation, as well as other lymphangions and thereby lymph transport.



The lymphangions are stimulated to pulsation by the activity of the muscle pumps.

Lymph transport is influenced by several auxiliary mechanisms. The most important of these in the limbs are the muscle pumps that come into effect during movement. The lymphangions are stimulated to pulsation by the interplay of contraction and relaxation.

Lymph vessels can adapt very readily to the amounts of lymph produced in different situations. The upper limit of its ability to compensate is known as "transport capacity".

The amount of lymph transported in a vascular segment per unit of time is known as "lymph volume over time".

The lymph is returned to the circulation through the major lymphatic trunks such as the thoracic duct. The lymph trunks empty into the large veins in

the collar bone region.

Every day about 2 liters of lymph - and with it a large amount of differing, vitally important proteins and immune defense cells - flow into the blood circulation

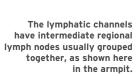


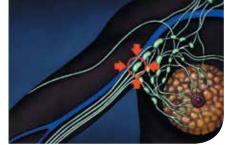
Lymph concentration and filtration in the lymph nodes

On the way into the blood circulation the lymph passes through several lymph nodes frequently arranged in groups. The first lymph node of these groups is known as the sentinel lymph node.

The lymph nodes also have important immunologic functions. They act as biological filter stations. Bacteria and cell debris are filtered out of the lymph and phagocytized by immune defense cells, such as macrophages, present in the

lymph nodes.





The lymph nodes also contain specific cellular defense substances which can destroy cancer cells.

Lymph undergoes further concentration in the lymph nodes, usually by absorption of water by the nodes. The water is removed through the veins.

In venous hypertension, however, fluid is also added to the lymph.

The scarcely pea-sized lymph nodes act as filter stations for disease pathogens, foreign bodies and cell debris and for concentration of lymph.

The absorbed water is carried away through the veins, while the filtrate is transported to the next lymph node station via the efferent lymph vessels.



Lymphedema

In this complex system of lymph production, transport, concentration and filtration, disorders can develop which can have serious consequences. The most prevalent of these disorders is lymphedema.



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Lymphedema is a chronic disease with a tendency to progression. It can affect all parts of the body, but most often the limbs are affected.



Left untreated, lymphedema can assume gigantic proportions.

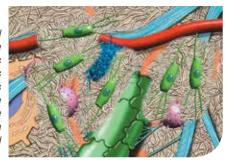
If left untreated, lymphedema can become disabling and at the very worst undergo sarcomatous degeneration.

Pathophysiology

When a disorder of lymph drainage develops, protein-rich fluid accumulates in the tissue. The interstitial amorphous ground substance swells.

As in a chronic inflammation, increased amounts of mononucleated inflammatory cells, such as monocytes, migrate into the edematous area. The monocytes transform into macrophages and multiply.

Protein-rich edema is characterized by secondary tissue changes: the accumulation of lipoperoxides attracts defense cells. Activated macrophages (colored blue) secrete various cytokines (e.g. interleukin-1) which stimulate the fibroblasts (colored green) to produce collagen. Connective tissue proliferation results in fibrosclerosis usually associated with fatty degeneration of tissue.



Especially the macrophages produce increased amounts of signalling substances, which stimulate the fibroblasts to divide and synthesize more connective tissue.

Some fibroblasts transform into lipid-storing adipocytes. The late complication is the fibrosclerosis typical of lymphedema and proliferation of fatty tissue.

Causes

The causes of lymph drainage disorder and thus also of lymphedema may be found in

- · lymph production,
- · lymph transport or
- lymph nodes

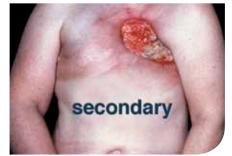


Lymphedema can have a primary cause such as congenital defect, or secondary causes such as acquired damage or disease of the originally intact lymphatic system.

Disorders of lymph production

Disorders of lymph production can develop as a primary phenomenon associated with aplasia of the lymph capillaries, or as a secondary consequence of damage

to the lymph capillaries during surgical procedures or diseases.



The causes of secondary lymphedema are an acquired obstruction or destruction of efferent lymphatic vessels, due for example to cancer treatment or severe injury.

Disorders of lymph transport

Lymphovascular lymphedema can also be of primary or secondary origin.

Primary limb lymphedema usually begins distally and spreads in proximal direction.

It can be present at birth or develop later on during the patient's lifetime. When left untreated, it can sometimes assume gigantic proportions.



Extensive scarring, e.g. after a severe burn, can also cause lymphedema.

Secondary lymphedema is often the result of a cancer operation. But lymph vessels can also be injured or destroyed by other surgical interventions.

Extensive scar formation after severe injury can also cause lymphedema. Distal to the drainage obstruction caused by the injury, the lymph stasis extends into the lymph capillaries. Lymph production is also affected.

Malignant tumors infiltrating into the lymph system or compressing the lymph vessels can also cause lymphedema.

Filaria that penetrate into and damage the lymph vessels cause a special form of secondary lymphedema. Filariasis is a widespread disease in the tropical regions, where the threadworms are transmitted by mosauito bites.





A disease endemic in tropical regions, where it is transmitted by mosquito

Another form of secondary lymphedema is artificial lymphedema: for various reasons some patients deliberately create a lymphedema by applying a tourniquet to their upper arm.

Damage to lymph nodes

Lymph stasis can also be of nodular origin. The swelling then usually begins centrally and spreads in distal direction.

The congenital absence of lymph nodes can give rise to primary lymphedema.



In lymph node fibrosis, the lymph node tissue is transformed into scar tissue for reasons that are still unknown. Lymph flow through the lymph node is thereby impeded.

In the treatment of breast cancer, lymph nodes are usually resected or damaged by radiotherapy. The result is secondary

arm lymphedema.



In cancer therapy, e.g. for breast cancer, several lymph nodes – depending on the tumor size and its histology – are either resected or damaged by irradiation. Lymphedema can then occur as an acute effect or as a secondary phenomenon after a latent period.

Diagnosis

Limb lymphedema can be diagnosed by specialized clinicians on the basis of a clinical examination.

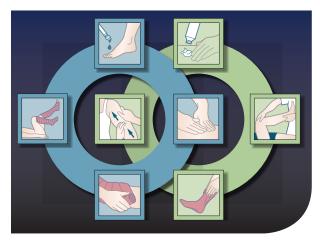
Modern imaging procedures can also be used if necessary.



Lymphedema Management -

Complete Decongestive Therapy (CDT) as the method of choice

The aim of lymphedema therapy is to mobilize the congested tissue fluid, reduce the proliferation of connective tissue and fat and achieve freedom from symptoms.



Complete Decongestive Therapy (CDT) consists of two phases, symbolized here by the two circles. In phase I (left) decongestion is predominant. Phase II (right) serves to maintain and optimize the results of this management. The following measures are applied in both phases: 1) compression therapy, 2) manual lymph drainage, 3) remedial exercises, 4) skin care.

An increasingly popular management technique is "Complete Decongestive Therapy" (CDT) which has practically no side effects.

It consists of two phases: the first phase is usually performed intensively. Lymph drainage is the main component in this phase.

The second, phase, is aimed at maintaining and optimizing the lymph decongestion achieved in phase I.

Both phases - integrated into an overall program of medical care - are adapted to meet actual requirements and comprise the following measures:

- compression therapy: in Phase I with compression bandages and in Phase II with medical compression garments.
- · manual lymph drainage
- · remedial exercises
- · skin treatment and skin care

Complete Decongestive Therapy (CDT) acts on lymph production, lymph transport and on the lymph nodes themselves.

The interplay of manual lymph drainage, compression therapy and movement in Complete Decongestive Therapy (CDT) reduces the pathologically elevated pressure in the lymph capillaries typically present in lymphedema.

Lymph is removed in increased amounts from the edematous region via the still

intact lymph vessels.



Complete Decongestive Therapy (CDT) loosens the fibrosclerosis, and indurated tissue and fat are reduced.

CDT interrupts the processes leading to chronic inflammation, and the indurated tissue softens. The indurated tissue and the fat are gradually broken down by metabolic processes.

Improvement of lymph transport

Improving lymph transport in the lymph vessels is a major objective of CDT. This can be achieved by the following therapy:

· Manual lymph drainage

In the valveless network of lymph capillaries of the skin that extends throughout the surface of the body, and which is not separated by watersheds, lymph can be propelled, by the manipulations of manual lymph drainage, from the edematous

areas of skin into non-edematous regions, from where they are removed by healthy lymph vessels.

The lymph territories are of major importance for lymphedema therapy. In the normal state, lymph is drained from the individual territories separated by lymphatic watersheds into the regional lymph nodes.



Lympho-lymphatic anastomoses connect the lymph territories and bridge over the watersheds. Usually only small amounts of lymph flow through these anastomoses.

In manual lymph drainage, which begins in healthy, non-edematous territories, a suction effect is generated by the increased lymphangiomotoricity. The anastomoses dilate and the lymph is transported from the edematous into the edema-free territory.

In limb lymphedema, the root of the limb is treated in the second step. The limb itself is then treated, beginning proximally and progressing distally.

Compression therapy

The decongestion achieved by manual lymph drainage is maintained and optimized by the compression bandage



A firm compression bandage suited to the prevailing drainage situation maintains the edema removal achieved in manual lymph drainage.

Remedial exercises to increase lymph drainage

During movement, the firm bandage acts as a non-yielding abutment for the contracting muscles and thereby enhances the effect of the muscle pumps.

The pulsation of the lymphangions is stimulated and the lymph volume transported over time is increased.

Further effects of lymphologic compression therapy

Compression bandages and medical compression garments increase the interstitial pressure. The fluid balance in the functional unit capillary, interstitial tissue/initial lymph vessel is optimized.

Compression bandages with suitable padding and medical compression garment with a textured surface in combination with movement have been shown to produce a micromassage effect that improves lymph transport.

The skin temperature increases by up to 5°C especially under heavily padded compression bandages. This effect enhances lymphangiomotoricity. A temperature increase also occurs under firm compression garments.



Diverting lymph flow around damaged lymph nodes

The lymph nodes are also influenced by the complex physical drainage therapy. If they are overloaded, fibrotic or have been removed, lymph stasis develops in the afferent lymph vessels in this region and spreads distally.

In these cases the lymph is conducted through the collateral vessels - through bypasses so to speak - around the obstruction.

The lymph volume transported over time in the collateral lymph vessels increases and the edema is reduced.

Skin treatment and skin care

Complete Decongestive Therapy (CDT) also commences with skin management procedures.

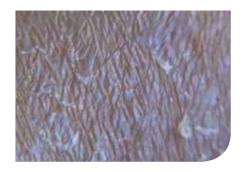


Thorough skin management is given before starting CDT. Interdigital spaces and skinfolds are especially vulnerable to mycoses and erysipelas.

Lifelong careful skin care is imperative, since the skin in the edematous region is particularly vulnerable to infections.

Especially interdigital spaces and skinfolds promote the development of mycoses and frequently also erysipelas.

During therapy the skin is subject to mechanical stress and its natural hydrolipid film is impaired.



The dry skin is subject to continuous mechanical stress and requires attentive care to restore the protective hydrolipid film.

Regular skin care, preferably with acid-buffered dermatologically tested skin care products, before applying a compression bandage and after removing a compression garment protects the skin and promotes restoration of the protective hydrolipid film.

Remedial exercises

Remedial exercises adapted to the phase of Complete Decongestive Therapy (CDT) and the patient's general condition, increase the effectiveness of compression therapy by activating and enhancing the muscle and joint pumps.

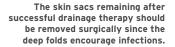
Furthermore, specific exercises reduce the risk of impairment of the musculoskeletal system due to immobilization.

Functional deficits of the muscles and joints can be remedied or improved by special remedial exercises.



Concomitant surgical treatment

After successful management of extreme lymphedema such as elephantiasis, the remaining empty sacs of skin greatly hinder continued therapy.





Particularly the wearing of made-to-measure compression garments becomes difficult. Furthermore, the skin in the skinfolds can macerate and become infected.

These skin sacs should therefore be removed surgically if possible.

Lymphostatic papillomatoses and lymphocutaneous fistulas can also be treated surgically or - in suitable cases - by laser therapy.



Lymphocutaneous fistula can be treated with lasers.

Management of combined forms

Lymphedema can also occur in combination with other diseases.

Often a disorder of fat distribution occurs in combination with lymphedema and is known as "lipolymphedema".

These combined forms, like lymphedema, are also managed, and in many cases compression therapy is the primary therapeutic approach.

Lymphedema is often combined with lipedema. Lipedema is a disorder where there is enlargement of both legs due to deposits of fat under the skin. Once again, CVT is the first-line approach.



Lymphedema is often mistaken for obesity, a typical lipedema patient is one who struggles with large hips and legs, usually out of proportion to the rest of their body.

Structure of a compression bandage for lymphologic indications

Compression therapy is indispensable in phase I and phase II of complex physical drainage therapy.





In the edema removal phase the volume reduction is so great that the compression bandages have to be adjusted daily to the altered size of limb. Only compression bandages are therefore suitable for this phase of therapy.

Because of their major therapeutic importance, the structure of multi-layered lymphologic compression bandages will now be explained:

- · first skin care is provided;
- then skin protection is applied in the form of soft-tube bandage; (eg: Tricofix®)



A soft tube bandage is fitted to protect the skin.

 toes and fingers are usually bandaged as well, even if not edematous. This prevents the edema from spreading to the fingers or toes. Contraindications are neuropathies such as microangiopathies.



The toes and fingers are usually bandaged as well.

 Padding made of highly textured padded bandages (eg: Artiflex[®]) is applied on top of the soft tube bandage and, in suitable cases, various uneven padding

materials.



Highly textured padded bandages provide good cushioning.

• The actual short stretch compression bandage (eg: Comprilan®) takes the form of textile-elastic bandages.

These bandages have very little elasticity. They are short-stretch and thus exert very low resting pressure.

During movement they provide a firm abutment to the muscle and thereby increase the effect of the muscle pumps. As a result, they have a high working pressure and therefore highly decongesting effect.



The actual compression bandage is applied in the form of textileelastic short-stretch bandages.

Compression bandages are applied according to the extent of edema. Usually they extend as far as the next large joint. For ankle edema, therefore, the bandage extends as far as the knee.



Compression bandages should always extend as far as the next large joint. For ankle edema, extend as far as the knee.

Contraindications for compression therapy

Ischemia (e.g. advanced arterial disease), uncontrolled congestive heart failure, untreated septic phlebitis, Phlegmasia cerulea dolens.

Compression therapy with medical compression garments

When drainage is largely complete and the limb is as free as possible from edema, phase II is commenced which serves mainly to sustain and optimize the therapeutic benefits already achieved.

In phase II, compression is carried out with medical compression garments. Lower pressures may be indicated for arm lymphedema.



Compression garments are indicated for the management of lymphedema.

Because of the high pressures required and the frequently abnormal shape of the extremities in lymphedema, most often made-to-measure medical compression garments are used. (eg: Elvarex®).

These garments have to be made of a firm fabric and should have similar properties to a textile-elastic short-stretch bandage.



Arm lymphedema is managed with compression sleeves.

To ensure an exact fit - essential for effectiveness and comfort - patients should only be measured for compression garments by trained specialized personnel applying defined criteria.



The precondition for a perfect fit and thus for optimal therapeutic success is accurate measurement of the limb according to defined criteria.

Two garments should always be used alternately, both to ensure that a garment is always being worn and for hygienic reasons. One is worn while the other is being washed.

Compression garments that fit poorly or have become slack through wear, or damaged garments undermine the success of therapy and are even harmful.



Slack and/or damaged compression garments impair the success of the therapy. The garments therefore have to be replaced every six months.

Garments should be replaced at least every six months.

Patient education and instruction promotes compliance

The provision of thorough education and instructions to the patient concerning the lifestyle modifications required by the illness are as indispensable a part of therapy as close medical supervision.

In all phases of therapy, the patients' compliance, their active cooperation, is essential to ensure continued therapeutic success.

Only when patients understand what the individual measures achieve will they actively cooperate in their therapeutic regimen.

Lymphedema is a chronic disease which tends to progress if left untreated. The consistent application of Complete Decongestive Therapy (CDT), however, can prevent this progression and improve the therapeutic outcome to an extent where patients can lead a virtually normal life – provided that patients continue their therapy, wear their made-to-measure medical compression garments as directed and adapt their lifestyle accordingly.

Are there alternatives to CDT?

At present there appears to be no pharmacological management of lymphedema that can compete with the therapeutic success provided by Complete Decongestive Therapy (CDT).

The microsurgical methods of management performed in only a small number of patients in highly specialized centers also require a lifelong post-operative phase II Complete Decongestive Therapy (CDT).

SUMMARY

Only the precisely coordinated interaction of the individual measures comprising Complete Decongestive Therapy (CDT) can yield success:

Compression therapy, manual lymph drainage, remedial exercise, skin treatment and skin care are only effective in combination.

Used individually, they have no therapeutic effect.

Complete Decongestive Therapy (CDT) of lymphedema is a therapy based on the latest scientific knowledge, but its fundamental concept has been applied successfully for more than 100 years. Many thousands of successfully treated patients can testify to the effectiveness of this treatment.



Even in extreme cases, Complete Decongestive Therapy (CDT) can achieve considerable success.

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30-40 mmHg*



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30-40 mmHg*



30-40 mmHg*

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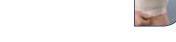
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This booklet is addressed to all health care professionals involved with lymphedema and its treatment. The booklet was produced in cooperation between Dr. med. Ethel Földi, the Földi Klinik, Hinterzarten (Germany) and BSN medical GmbH, Hamburg (Germany) / JOBST GmbH, Emmerich (Germany).

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^{*}The mean compression for average ankle size.