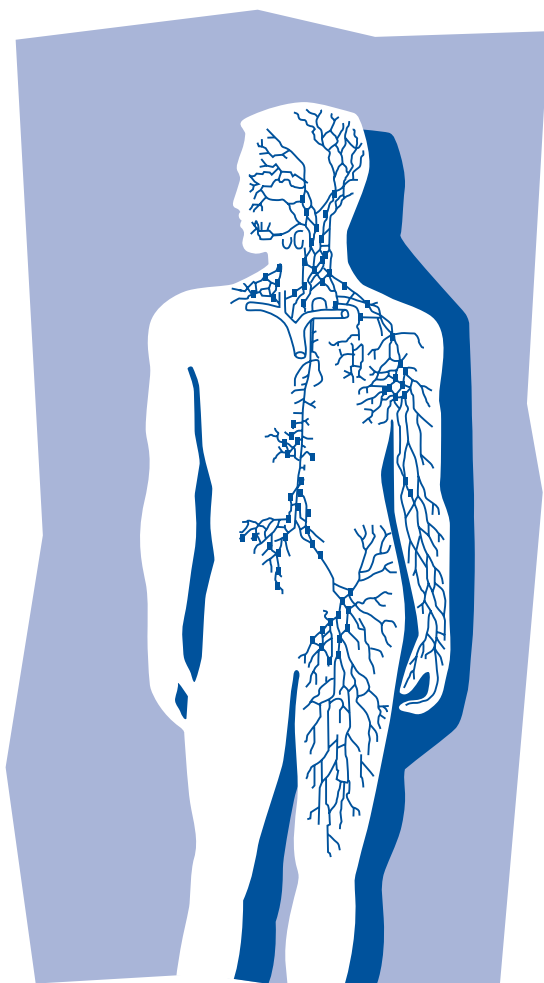


Lymphedema

its cause and how to manage it



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Dear Patient,

Your doctor has told you that you have lymphedema. Perhaps this is the first time in your life that you have ever heard of such a condition. Or perhaps you have already heard something about it from your friends and family. Whichever is the case, you would probably like to know a lot more – what the causes are, what you can do and, above all, the extent to which it will affect your life.

To deal with the most important point first: lymphedema is a long-term condition and you will have to change your lifestyle in certain ways to cope with it. But once you make those changes, you will be able to live an almost normal life without too many restrictions.

The purpose of this information booklet, which is based on the latest medical and scientific knowledge, is to tell you more about living with lymphedema. If your own doctor or therapist suggests a slightly different approach, however, you should follow their recommendations. Only your own clinicians are familiar with your particular situation and are in a position to offer you specific advice.

We wish you all the best!

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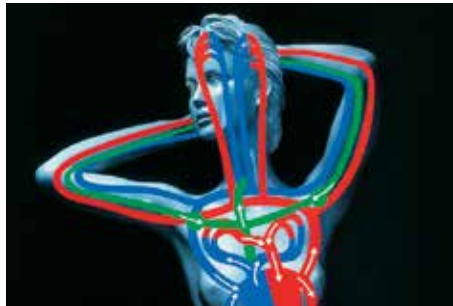
1. Lymphedema... which organ is actually affected?

Whenever we hear “lymph...”, most of us immediately think of lymph nodes. We know that we have them in the neck and groin, for example, and that they can become swollen when inflamed. This is certainly true but not the whole story. The lymph nodes, which are present throughout our body (there are several hundred of them) are part of an interconnected **lymphatic system**.

1.1 The lymphatic system: structure and function

The lymphatic system is a transport system accompanying, and closely associated with, the blood circulation system in our body.

The lymphatic system (shown in green) runs closely alongside the blood circulation system with its arteries (red) and veins (blue).



- As you may know, the blood circulation system consists of arteries and veins and can be compared to a kind of branched pipeline system in which the heart represents the pump. From the heart, the arteries, which are large tubes, carry blood to the organs, where they branch out into a large number of very narrow tubes with porous walls (capillaries). When the capillaries leave the organs, they join up again to form large vessels, called veins, that return the blood to the heart. The capillaries are permeable to allow an exchange of substances

in the organs. The arteries supply the organs with blood which is rich in oxygen and nutrients. In turn, the veins transport metabolic waste products and carbon dioxide away from the organs. These materials are then excreted via the kidney and liver. The blood vessel system is therefore a closed circuit.

- The lymphatic system has a similar structure: it also has vessels and capillaries (and at repeated intervals the lymph nodes already mentioned). The lymphatic system, however, is only a semi-circulatory system. It begins with its capillaries directly in the organs where they can absorb tissue fluid and proteins before removing them. This protein-containing tissue



The large conducting vessels (arteries in red, veins in blue, lymph vessels in green) branch out into tiny, tubes (capillaries). These are responsible, among other things, for the exchange of nutrients and metabolic waste products. Lymph fluid is also produced in this region.

fluid is known as **lymph fluid** or **lymph** and – in contrast to blood – is colorless. The lymph is transported by the capillaries to the conducting vessels of the lymphatic system. These vessels finally empty into the veins, connecting the lymphatic system with the blood vessel system. The flow of lymph fluid is caused by the rhythmic pulsation of the lymph vessel walls (alternately contracting and relaxing).

Both the veins and lymph vessels, therefore, have the job of removing waste substances. The veins cannot do this alone, as some substances can only be removed by lymph, particularly larger molecules such as proteins. The lymph capillaries have large pores to “pull” in these substances dissolved in water. The excess absorbed water is filtered

out of the lymphatic system in the lymph nodes, which thus have an important regulating function.

The lymphatic system has other functions, such as a role in the body's immune defense system, which will not be discussed here. Even so, it is clear that damage to the lymphatic system will mean that proteins and tissue fluid simply remain where they are in the tissues – between the cells. The resulting swelling of tissues caused by accumulation of lymph tissue fluid is known as **lymphedema**.

When the lymph tissue fluid can no longer be completely removed, lymphedema develops. This occurs most often in the arms and legs.



2. Impairment of the lymphatic system and its consequences

There are completely different reasons why the functioning of the lymphatic system may be impaired (insufficient). Two main causes can be identified:

- The lymphatic system may be abnormal from birth. When a lymphatic system of this type is no longer able to cope with the demands placed on it, sooner or later during that person's life, **primary lymphedema** develops. It is called "primary" because the cause of the impairment is to be found in the structure of the lymphatic system itself.

- The lymphatic system is fully adequate and functions perfectly; however due to some external influence (such as an injury, surgery or radiation), it becomes damaged. This can adversely affect its function and ultimately lead to **secondary lymphedema**.

2.1. Primary lymphedema

Primary lymphedema can have a variety of causes. For example, the lymph capillaries may be absent (aplasia of the initial lymph vessels); in such cases only modest amounts of lymph fluid can be produced. Sometimes there are simply too few lymph vessels in the body, or they are too narrow (hypoplasia). The lymph transport capacity is then insufficient to remove the accumulating lymph fluid.



Primary lymphedema is usually caused by lymph vessel malformations present from birth.

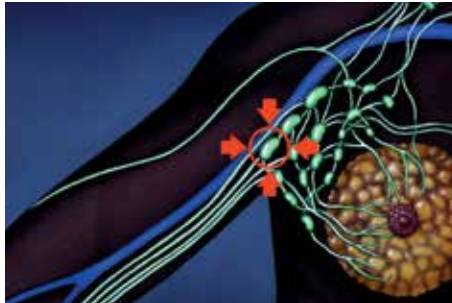
The most obvious sign of decreased lymphatic function is a swelling – lymphedema. However this does not necessarily appear immediately and may only develop gradually due to a continuous overload on those lymph vessels that are still functioning. If primary lymphedema first appears between the age of one and 35 years, doctors refer to it as *Lymphedema praecox* (= premature lymphedema), and if it develops after the age of 35 it is known as *Lymphedema tardum* (late onset lymphedema). In some cases the swelling is already apparent at birth (congenital lymphedema). Primary lymphedema may occur with an increased frequency in certain families or may develop sporadically, as a random event.

2.2. Secondary lymphedema

Anything that injures a healthy lymphatic system can potentially result in permanent damage leading to secondary lymphedema. These harmful factors may be contusions or dislocations, or infections with fungi, bacteria, viruses or worms, particularly in some tropical countries.

In the Western world, however, the most common cause is the treatment of malignant tumors. Many cancer cells can migrate through the lymph vessels to the nearest lymph nodes where they form metastases. In such cases it is important to surgically remove some of the lymph nodes in the affected area, as well as the tumor nodes in the affected area, thereby damaging the lymphatic system. The lymph nodes that have been removed are examined for cancer cells, indicating whether surgery has to be followed up by chemotherapy or radiation therapy.

A frequent cause of secondary lymphedema is the surgical removal of lymph nodes, e.g. from the armpits of breast cancer patients.



Radiation therapy can damage the lymph vessels, but the successful treatment of the cancer must be given precedence when deciding on the therapeutic approach.

Removing the lymph nodes means that lymph drainage is interrupted. When the lymph nodes in the armpit are removed because of breast cancer, lymph drainage from the arm is impaired. If lymph nodes in the groin are removed, drainage from the affected leg and from the genitals is impaired.

In some cases, the body finds ways of compensating for this damage to a certain extent. For example, it can stimulate existing lymph vessels to increase their performance or create connecting branches (anastomoses) to other lymph vessels or veins. However, a degree of impairment will always remain due to the fact that lymph nodes cannot be regenerated.

Depending on how well this compensatory mechanism works and the degree of stress to which the particular person's lymph system is subjected, it may be that no lymphedema occurs at all. On the other hand, lymphedema may develop either very soon after the operation, or after a delay of several years.



Secondary arm lymphedema resulting from the removal of lymph nodes and/or radiation therapy for breast cancer.

2.3. The different stages of lymphedema

We have already seen that lymphedema does not always develop overnight as the body has certain compensatory mechanisms at its disposal. Once it has started to develop, however, complicated mechanisms come into play which cause the lymphedema to progress over a period of time. The accumulation of protein-rich fluid in the tissues causes cells to be activated that produce new connective tissue substance. Doctors refer to this process as **fibrosis**.

The area affected by lymphedema also becomes increasingly hard; this is known as **sclerosis**.

The processes occurring in the skin and subcutaneous tissue in untreated lymphedema also show certain similarities with chronic inflammation. Although not recognizable as an inflammation from outside the body, the altered conditions weaken the skin's immune defense system. As the severity of the lymphedema increases – the skin becomes more susceptible to bacterial infections such as **erysipelas** and to fungal infections. These infections in turn affect the lymphatic system and worsen the lymphedema, setting off a vicious cycle.

The clinical stages of lymphedema are defined according to these individual developmental steps:

- **Stage 0 (latent stage):** This is the situation in which the lymphatic system is already functioning inadequately, but it is still coping – by means of compensatory mechanisms – with the lymph that is produced. No edema is present.
- **Stage I (reversible stage):** The lymphatic system is overburdened; a protein-rich soft swelling develops in the affected area. It can be indented by applying pressure. When the affected arm or leg is raised, the swelling reduces of its own.



Stage I (reversible stage) of lymphedema: the tissue is still soft; this seen during examinations when it can still be indented by applying pressure.

- **Stage II (spontaneously irreversible stage):** The swelling is already characterized by the presence of excess connective tissue; fibrosis and sclerosis have developed. Indentations can only be produced by applying strong, deliberate pressure; raising the limb no longer reduces the swelling.



Stage II (spontaneously irreversible stage) of lymphedema: excessive connective tissue has already formed and begins to harden

- **Stage III (elephantiasis):** The swelling is extreme, the skin is hardened and shows wart-like growths. Sometimes large bulges are present. The risk of inflammation (erysipelas) is high, and the skin is vulnerable to the development of deep, poorly healing wounds.



Stage III of lymphedema (elephantiasis): The skin is hardened and exhibits growths. Sometimes large bulges are present.

3. Management of lymphedema

Because of the mechanisms described above, there is little point in simply ignoring the condition and allowing it to take its course. It will then almost inevitably become steadily worse.

The sooner you begin a suitable program of therapy, the better are your prospects of preventing disease progression and of improving your condition, and returning to a less severe stage. If therapy is begun at Stage I, for example, it is often possible to return to Stage 0. (Although you would still need to remain under close medical supervision, you would probably not need therapy for a long time.)

For a specialized physician, a clinical examination is often enough to diagnose lymphedema of the arms or legs. This should be done early on to prevent the disease progressing.



3.1. Complex decongestive physiotherapy (CDP)

Complex decongestive physiotherapy (CDP) consists of two phases. In Phase 1, the aim is to achieve the best possible result in terms of reducing the swelling. The purpose of Phase 2 is to maintain and further improve the situation with respect to any existing fibrosis and sclerosis. CDP is divided into two phases:

- **Phase 1 edema reduction**
- **Phase 2 maintenance**

Complex decongestive physiotherapy is a combined form of therapy, both phases consisting of four elements. All the elements have particular contributions and can only be fully effective if applied together. The four elements of CDP are:

- **Skin care**
- **Manual lymph drainage (MLD)**
- **Compression therapy**
- **Exercise therapy**



Complex decongestive physiotherapy consists of two phases. Phase I (blue) is devoted mainly to edema removal, phase II (green) to maintaining and further improving the treatment outcome. The following measures are applied in both phases:

- Skin care
- Manual lymph drainage
- Compression therapy
- Exercise therapy

CDP Phase 1 usually takes about 4 weeks or less if treatment is commenced at Stage I of the illness. It can take much longer (several months) if the disease has already reached Stage III. Phase 2 of CDP is usually continued indefinitely – one exception, as mentioned above, is when the condition can be returned to Stage 0.



Even in severe forms of lymphedema (here a Stage III) complex decongestive physiotherapy can provide considerable benefits (left: before, right: after).

3.2. The four elements of CDP

- **Skin care:**

Regular protective skin care in lymphedema is essential as the skin is vulnerable to inflammations and infections. As already explained (see 2.3), infections can further aggravate the disorder. To help prevent bacteria from penetrating through the skin, therefore, it is important to keep the skin as intact as possible.

Regular protective skin care is indispensable since skin affected by lymphedema tends to develop infections and inflammations.



Skin care should be carried out twice daily, in the morning and evening. You can choose a product suitable for your personal needs in consultation with your doctor.

- **Manual lymph drainage (MLD):**

Manual lymph drainage (MLD) is performed by therapists specializing in this treatment. It should not be confused with conventional massage. The purpose of MLD is to stimulate the pulsation of the lymph vessel walls, which accelerates drainage of the lymph in the lymph vessels that are still functioning.



Manual lymph drainage can move lymph fluid from the swollen region into parts of the lymphatic system that are still functioning, from where the lymph can then be transported away.

You may be surprised to find that your therapist starts the MLD much higher than where you see the swelling. This is because the visible lymphedema always has its cause somewhere else. You can compare it to a traffic jam where the actual cause, such as the road being blocked by an accident, is sometimes located some distance away. The problem can be solved by clearing the accident site or by creating a bypass, but not by attempting to push one's way forward from behind. MLD is performed once or twice daily in Phase 1 of CDP; in Phase 2, this can then be reduced to a frequency of once to three times a week.

- **Compression therapy:**

Manual lymph drainage increases the flow of lymph in the still functioning lymph vessels, allowing more of the accumulated fluid to be carried away by the lymph capillaries. This process can be greatly enhanced by suitable compression therapy. This therapy has a positive influence on the pressure conditions in the tissue. Since we have not considered this aspect so far, here is a brief explanation:

The exchange of fluids between the capillaries of the blood circulation and lymphatic system and the organs, such as the skin, is dependent on differences in pressure. The direction of flow is always from the area of higher pressure to the area of lower pressure. When everything is intact, nature has provided a combination of varying vessel diameters, different protein and glucose concentrations in the fluids and special pumping mechanisms to ensure that the flow goes in the right direction. In other words, from the arteries into the tissue, and from the tissue back into the veins and lymph vessels. In lymphedema, the lymph vessels are no longer working properly and,

these pressure conditions are disturbed. As a result, too much fluid accumulates in the tissue. The idea behind compression therapy, therefore, is to assist the tissue by applying pressure from outside, which allows the fluid to flow back into the lymph vessels and veins. Depending on the severity of lymphedema, different degrees of pressure are needed for this purpose. In Phase 1 of CDP (edema reduction) compression bandages, together with padding materials, are used for compression therapy. The advantage of a compression bandage is that it can be continuously adjusted as the swelling decreases. In this phase of treatment, low-stretch materials (such as the Comprilan® short-stretch bandage) provide optimal results.

In Phase 1 of the therapy a compression bandage made from low-stretch bandages is applied.



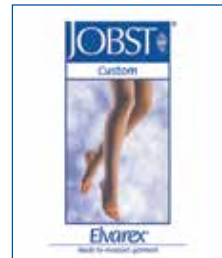
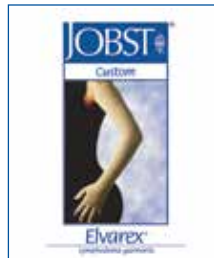
The low-stretch bandages provide a natural resistance to the pressure exerted on the tissue during movement of the muscles. This promotes the return flow of lymph and brings about a reduction in the swelling.

In Phase 2 of CDP (maintaining and improving the result), medical compression garments are generally used.



In Phase 2 of the therapy a compression stocking is prescribed which is often custom-made.

Elvarex® compression garments have proved successful in the management of lymphedema and are available in a style to suit any individual need.



Medical compression garments help maintain the result achieved in reducing the edema. They can be more stretchable than the bandages which makes them more pleasing to wear and allows easier application. Due to the greatly differing degrees of lymphedema, garments often have to be custom-made. Compression garments are also available in various compression classes (compression pressures) and different styles (stockings, open or closed toe, gauntlets, arm sleeves etc.) and can thus be selected to suit individual requirements perfectly.

Exact measurements are essential to achieve a perfect stocking fit.



Compression garments of the **Elvarex®** brand have been an indispensable feature of lymphological care.

- The flat-knitted stocking is made to suit your individual sizes, ensuring the best possible fit. This is extremely important for a product that has to be worn every day.
- The unique-knit fabric prevents the stocking getting caught in skinfolds.
- Some experts believe that the special knitted fabric not only provides compression (acts as “compression aid”), but also exerts a micromassage effect when worn during exercise or movement.

Elvarex® compression stockings contribute to the success of therapy.



Please follow your doctor's advice regarding the garment quality, style and compression class that are most suitable for you. As soon as you are accustomed to putting on, taking off and wearing it, you will soon come to value the relief provided by your compression garment. In use, you should wear your compression garment every day for as long as you are up and about. Take the garment off at bedtime; arm sleeves may be an exception to this rule depending on what your doctor advises.

• **Exercise therapy:**

Exercise therapy is the fourth component of CDP. Compression bandages or compression garments are worn during this treatment. Exercise therapy allows the compression to exert its affect to the fullest and helps drain fluid from the tissue.



Regular physical exercises promote lymph drainage.

You should perform your exercises as often as possible, for example two to three times a day (both in Phase 1 and in Phase 2 of CDP). Which particular exercises are most suitable for you depends on which part of your body is affected by lymphedema. Your therapist will design a program of exercises especially for you. Perform your exercises as diligently as possible and only continue as long as you feel no pain. Include the side of your body not affected by lymphedema in your training.

3.3. Other forms of therapy

At present there is no medicinal alternative to CDP.

4. How can you help make the therapy successful?

- Follow your doctor's and lymph therapist instructions and advice regarding your therapeutic program.
- Should you notice anything unusual (redness and excessively warm skin, inflammations or wounds, pain, an increase in the swelling, or perhaps that your compression garment doesn't fit properly) please consult your doctor immediately.
- Success with CDP depends on your full cooperation: regular skin care, wearing the compression garments, keeping your appointments with the lymph therapist and performing the recommended exercises are all up to you!

Consistent application of the CDP therapy, which includes wearing the compression garment will allow you to lead an almost normal life without restrictions.



Some more useful tips to remember:

- **Your diet...** should be balanced. If you are overweight, you should try to lose weight under your doctor's guidance, as obesity places an additional burden on the lymphatic system.

- **Your responsibility:** Having your blood pressure measured, a blood sample taken, injections, acupuncture and so on, should not be carried out on the affected arm or leg. Please remember to inform whomever it is that you have a lymphatic condition.

- **In your leisure time...** you should protect yourself against overexposure to the sun (which also includes tanning beds), because the warmth causes widening of the vessels, and secondly because sunburn puts even more stress on your already affected skin. It is also advisable to be cautious around insects – their bites or stings can cause inflammations that may worsen the lymphedema.

- **In the home and garden...** try and take precautions against injury (e.g. wear protective gloves) and avoid overstraining (e.g. carrying heavy objects).

- **Your sporting activities...** are important and beneficial – although you should restrict them to a reasonable level. Types of sport involving extreme stress can have an adverse effect on the lymphedema. If in doubt, just ask your therapist or physician.

- **Your clothing...** can still be fashionable but avoid restrictive items such as narrow bra straps and tight underwear.



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**Comprilan® Short
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