

Fizjoterapia w leczeniu obrzęku limfatycznego

Physiotherapy in the treatment of lymphoedema

Jacek Kultys^(A,B,E,F,G), Teresa Pop^(A,D,E,G), Ryszard Bielak^(A,D,E,G)

Instytut Fizjoterapii Wydziału Medycznego Uniwersytetu Rzeszowskiego, Polska/
Institute of Physiotherapy, Faculty of Medicine, University of Rzeszow, Poland

Streszczenie:

Obrzęk limfatyczny jest przewlekłą chorobą, spowodowaną zaburzeniem działania układu limfatycznego, prowadzącym do gromadzenia się chłonki w przestrzeni śródmiąższowej. Konsekwencją pierwotnego lub wtórnego obrzęku limfatycznego jest nieestetyczny wygląd kończyny, ograniczenie jej czynności, a nieraz znaczne zniekształcenie kończyny z deformacją struktur kostno-stawowych i zanikami mięśniowymi. U chorych często pojawiają się problemy emocjonalne, wpływające na kontakty interpersonalne i jakość życia pacjentów. Kompleksowa fizjoterapia udrażniająca jest nadal uznawana za najskuteczniejszą metodę leczenia zachowawczego obrzęku limfatycznego, a odpowiednie zaplanowanie i stosowanie jej w poszczególnych stopniach zaawansowania choroby przynosi dobre efekty terapeutyczne.

Słowa kluczowe:

obrzęk limfatyczny, leczenie, kompleksowa fizjoterapia udrażniająca

Abstract

Lymphoedema is a chronic disease caused by the malfunction of the lymphatic system, leading to the accumulation of lymph in the interstitial space. The consequence of a primary or secondary lymphoedema is an unesthetic appearance of the limb, limited activity, and sometimes a significant distortion with a deformation of osteoarticular structures and muscle atrophy. Emotional problems often appear, affecting interpersonal relationships and the quality of life of the patients. A complete decongestive physiotherapy is still considered to be the most effective method of conservative treatment of lymphoedema, where a corresponding plan and implementation of the treatment throughout different degrees of severity of the disease brings good therapeutic results.

Key words:

lymphoedema, treatment, complex decongestive physiotherapy

Introduction

The concept of "white blood" in lymph was already described by Hippocrates (460–377 b.c.) in antiquity. However, it was only in the seventeenth and eighteenth century various aspects of anatomy and physiology of the lymphatic system as a whole were discovered and thoroughly researched. In 1627, Gasparo Aselli first provided a detailed description of lymphatic system; Dutch biologist Swammerdam discovered valves in the lymph collectors in the same year. In the eighteenth century, Louis Petit described the phenomenon of breast cancer spreading to axillary lymph nodes [1]. Although the anatomy of the lymphatic system was thoroughly described by the nineteenth century, there are still many issues regarding the lymphatic system that require further research, including controversial concept of ontogenetic origin of lymphatic vessels, whether they develop from veins or de novo from lymphatic angioblasts or are both these processes involved in the development of the lymphatic system [2].

The aim of this paper is to discuss physio-therapeutic programmes used when working with patients with lymphoedema, as recommended by The International Society of Lymphology and Italian Society of Phlebology.

Anatomy and physiology of the lymphatic system

There are many more lymphatic vessels in human body than blood vessels; their main task is transporting lymph to the blood. Lymph is unreabsorbed tissue fluid formed through the filtration process occurring on the microcirculation level. Lymphatic vessels are not found in all tissues, for example central nervous system, bone marrow, striated muscles, thymus and placenta are not part of the lymphatic vasculature [3].

Lymphatic system comprises three basic elements: capillaries, lymphatic collectors and trunks. Lymphatic capillaries are used to drain tissue fluid and produce lymph. They create a non-valvular network of fine vessels located in the direct vicinity of the blood capillaries. Lymphatic capillaries then create the collectors which have valves and function as the actual lymph transporting vessels with a diameter ranging from 0.1 to 2 mm. The valves in the collectors are located every 0.6-2 cm; in the thoracic duct they are more scarce and are found every 6-10 cm. The section of the vessel between two valves is called lymphangion. Lymph flow is forced by spontaneous contraction of individual lymphangions. Lymphangion contracts rhythmically on average 4-6 times per minute, pumping the lymph to the previously emptied lymphangions above. The valves prevent the backflow of lymph to the lower-lying lymphangions.

Lymphatic trunks are the biggest vessels of the system. Their task is to collect the lymph from the collectors and discharge it into the venous system. Lymphatic vessels of the lower limbs comprise deep lymphatic system and surface lymphatic system which transports about 80% of the lymph from the leg. Deep lymphatic system contains vessels located along the saphenous vein. Both systems are joined in the popliteal and inguinal lymph nodes [4]. Lymph from the lower limbs is discharged through the left and right lumbar trunk, which join each other and the intestinal trunk, creating the thoracic duct, 40 cm long and 2-5 mm wide. It is located on the spine in the retroperitoneal space, from the L1 vertebra, where baggy enlargement with the diameter of 0.5-1.5 cm is found, referred to as chyli receptaculum, to which intestinal trunk discharges. In the upper half of the body the lymph is transported by the cervical trunk (head and neck), subclavian trunk (upper limb, upper and side of the chest, breast) and broncho-mediastinal trunk (lungs, bronchi and mediastinum). On the right side these three vessels create a short right lymphatic duct; on the left side they discharge into the thoracic duct. Both of these ducts discharge respectively to the right and left venous angle formed by the merging of the internal jugular vein and subclavian vein and creating the brachiocephalic vein.

In summary, the thoracic duct collects lymph from both legs, abdomen, lower pelvis, left upper limb, left half of the chest, neck and head. Right lymphatic duct drains lymph from the right arm, right half of the chest, neck and head.

An important element of the vessels transporting lymph are the nodes, which, as clusters of lymph tissue involved in the processes of the immune response, also function as filters. Humans have approx. 600-700 lymph nodes, single or arranged in groups, which receive lymph from specific areas of the body [5].

Pathology of the lymphatic system

Humans produce approx. 2.5 litres of lymph daily. This amount may increase even 10-fold if certain interstitial fluid production increasing factors occur, including blood stasis in the superior vena cava, heart failure, immobilisation of certain parts of the body and failure of the peristaltic function of the gastrointestinal tract. All the above may lead to the increased lymph flow and the formation of lymphatic oedema or lymphoedema. It can also be caused by expansion of lymphatic vessels due to blood stasis or their underdevelopment as well as occlusion of lymphatic vessels after major surgery and bacterial or parasitic infections [3].

Lymphoedema is a chronic disease caused by malfunctioning lymphatic system. One of the symptoms is lymph accumulation in the interstitial space, at first mainly in the suprafascial area, and then in the subcutaneous tissue [6]. It is experienced by approximately 300 million people worldwide and is recognised as a serious health problem which tends to be ignored by the affected [4, 7].

From the point of view of the physiopathology, lymphatic system failure may be defined as dynamic or mechanical. Dynamic or large capacity failure occurs when a properly functioning lymphatic system needs process the amount of protein which exceeds its capacity. Overproduction of the filtrate is caused by failure of other systems, for example circulatory system [7]. Mechanical or low volume failure occurs due to primary or secondary damage to the lymphatic system, which struggles to cope with normal level of protein content [8]. It can be caused by infectious processes, surgeries, radiotherapy, congenital malformations of the lymphatic vessels and injuries. This leads to imbalance between the rate of lymph production and its drainage [7]. Proteins cause the flow of mastocytes and neutrophils as well as non-specific granulation process, which consequently results in fibrosis occurring within the interstitial space, leading to irreversible tissue damage [9].

Well-functioning lymphatic system depends largely on the efficiency and flexibility of the endothelium. Lymphoedema may also be caused by excessive permeability of the endothelial walls of which lymph vessels are built. This reaction is principally triggered by histamine released in a number of situations, for example injuries or allergic reactions, which may develop into a full inflammatory response. The endothelial permeability is affected by other factors too, including ischemia - reperfusion, atherosclerosis, sepsis, diabetes, thermal injury, angiogenesis and cancer metastasis.

Aberrations of the endothelial function cause the release of not only histamine but also of serotonin, thrombin, bradykinin, substance P, PAF, cytokine, vascular endothelial growth factor (VEGF) and reactive oxygen species [10]. Research has also been carried out on the genetic background of lymphoedema. It has been shown that a mutation in a transcription factor FOXC2 gene, also referred to as the lymphoedema-distichiasis syndrome, causes lymphoedema of the lower limbs and distichiasis (additional row of eyelashes) [11, 12].

Types of lymphoedema

There are two main types of lymphoedema: primary, which occurs mainly in the lower limbs (94% of cases) and secondary, which means both lower (31%) and upper (66%) extremities are affected.

Primary lymphoedema is a great majority of cases is experienced by female patients. It has been categorised due to the reasons for its occurrence:

- a) Primary hereditary lymphoedema. It is caused by the lack of or insufficient underdevelopment of the lymphatic vessels, present from birth. It may have genetic background, or be inherited (Milroy disease);
- b) Primary lymphoedema praecox. It affects people between 1 and 35 years old, and is in most cases caused by infection in an extremity, trauma or rapid growth causing retardation of lymphatic vessels (Meige disease);
- c) Primary lymphoedema tarda. It occurs in patients over 30 years of age and might be triggered by pregnancy [13].

Secondary lymphoedema may be caused by:

- a) Occlusion of lymphatic vessels caused by:
 - External factors, including pressure exerted by a tumour;
 - Internal factors, including bacterial infections (streptococcus) and parasitic infections (filariasis), for example a disease caused by a Wuchereria Bancrofti infection, occurring mainly in Asia).
- b) Damage to ducts and lymph nodes as a result of:
 - An injury, as part of Sudeck syndrome, including fractures and dislocations;
 - Surgical removal of the lymph nodes, radiation;
 - Lack of limb function due to paralysis, paresis or prolonged immobilization [14].

Lymphedema, due to their severity, may be divided into the following stages:

Stage 0: Subclinical or latent oedema. No visible swelling can be observed, however irregularities in lymphatic drainage can be detected in the lymphographic examination.

Stage I - Mild oedema of the foot and lower leg can be observed, especially at the end of the day. It is greatly reduced spontaneously following elevation of the limb.

Stage II - Spontaneously reducing all-day swelling, almost completely disappearing after a night's rest, positive Stemmer's sign (thickening of the skin folds on the second toe, skin difficult to lift).

Stage III - Permanent swelling, not reducing after limb elevation. Swollen tissues become increasingly hard.

Stage IV - Permanent, limb deforming swelling, often associated by inflammatory lesions of the skin (erysipelas, eczema, lymph fistula).

Stage V - Also known as elephantiasis, a significant limb deforming swelling, accompanied by skin thickening, muscular dystrophy, disabling the limb function. As a late complication of this oedema, malignant degeneration may occur in the form of lymphangiosarcoma [7, 15].

Diagnostics

Thorough physical examination may reveal the cause of lymphoedema and malfunctions of the lymphatic system may be recognised. Comprehensive additional testing must be scheduled. Although the aetiology of primary and secondary oedema differs significantly, clinical image and the symptoms of both diseases tend to be similar [16]. An interview with the patient can provide information about basic diseases, including heart and kidney problems along with data regarding the length of the disease and its course.

Patient suffering from lymphoedema must be examined in an upright position and lying down. The examination must include the assessment of skin condition of the skin, oedema location, occurrence of spontaneous pain or pain on palpation, depth of skin folds, pleural effusion, varicose veins or lymphatic vessels, enlarged nodes, signs of inflammation of the lymphatic vessels or other skin lesions, current or healed. Palpation allows to assess the tenderness and consistency of swelling. A doctor or physiotherapist should evaluate to which extent the swelling is prone to shaping, as well as assess the Stemmer's sign and measure the limb precisely, while assessing the motion range of the joints. All accessible lymph nodes should be examined. Body mass and height of the patient must be recorded too [17].

Lymphoscintigraphy and ultrasonography are currently the most common techniques in the diagnosis of the lower limbs lymphoedema. Lymphoscintigraphy is the basic non-invasive examination with a significant repeatability. It provides the possibility of quantitative interpretation of the obtained images, enabling to obtain results with high sensitivity and specificity [18, 19]. Other diagnostic methods include: vein Doppler ultrasound, microlymphangiography, computed tomography (CT) and magnetic resonance imaging (MRI), venography, lymph nodes biopsy and, in some cases, lymphangiography [4, 20].

Lymphoscintigraphy performed using Technetium-99, as a radiolabeled colloid, is intended to allow morphological and functional assessment of the lower limb lymphatic system [21, 22]. Ultrasound examination of soft tissue presents the lymph fluid in the interstitial and suprafascial areas [23].

Other tests are used mainly to complement the diagnosis. Computed tomography with contrast, venography, lymphography and biopsy of lymph nodes allow for the exclusion of hereditary malformation of the lymphatic system and cancer [24].

Fluorescent microlymphangiography is a minimally invasive method for an assessment of spontaneous lymphatic drainage of certain substances administered intradermally [25, 26].

The diagnostic tests listed above can be complemented by bacteriological examination of skin flora and histopathological tests of the skin [4].

Treatment

The treatment of lymphoedema is not easy. It is still considered to be a condition which cannot be completely cured. It is possible, however, to stop the progress of the changes, as well as a significantly reduce the swelling and limit the complications associated with the lymphatic swelling [4]. The majority of treatment procedures are focused on treating symptoms. Surgery methods are reserved for selected cases only when the disease is highly advanced. The therapy should be carried out comprehensively. This means that no form of treatment used separately will bring the desired results, as opposed to combined treatment, involving the use of drugs, limb lifting, manual lymphatic drainage, bandaging, compression techniques, air sequential massage apparatus (Pressure Therapy), and finally surgery (oedema reduction, resection and reconstruction procedures) [5, 27, 28, 29, 30, 31, 32].

Specialised centres providing comprehensive lymphoedema treatment are still in short supply [5, 15, 17, 33, 34, 35, 36, 37, 38].

Comprehensive physiotherapy

As early as the nineteenth century it was discovered that the results of individual therapeutic approaches in the treatment of lymphoedema are limited. In 1885, Esmarch and Kulenkampff published a paper on the benefits of combination therapy which involves manual drainage, compression techniques and physical exercise undertaken by run a cooperating patient. But it was only Stillwell in 1950, and Foldi in 1970 that developed the concept of "combination therapy" and outlined its principles [39].

Currently, the primary method of treating lymphoedema, as recommended by the International Society of Lymphology, is a Complex Decongestive Physiotherapy, which involves

manual lymphatic drainage, compression techniques (initially bandaging, then carefully selected compression products), active and active-passive exercises and skin care. Additionally, supporting methods include: limb elevation and heat treatment (local temperature increase to approx. 41 degrees Celsius reduces the volume of lymph, warm mud compresses can be used for this purpose). On the other hand, heat generally is contraindicated, including hot baths, and sauna.) [5, 36, 40]. Other forms of complementary therapies aimed at supporting the complex decongestive physiotherapy involve kinesiotaping, HBOT (Hyperbaric Oxygen Therapy) and electro-stimulation (using devices that use an electric field to assist in the flow of lymph) [7].

Physical treatment should not be limited to one single procedure. It is best if a range of methods are combined, depending on the stage of oedema and treatment accessibility [34, 35]. Complex physical treatment is divided into two categories. The methods classed in the first category are focused on reducing the content of lymph in the interstitial space and, consequently, a reduction in limb volume. The methods form the second category aim at stabilisation and improvement of the obtained effects, mainly by wearing compression products [41].

Manual Lymphatic Drainage (MLD)

One of the basic ways to improve the efficiency of lymph drainage is manual lymphatic drainage. Basics form of MLD was described in the nineteenth century by F.Winiwarter [42], later further developed and perfected by a Danish physiologist Emil Vodder in the 1930s [43]. Vodder technique was then perfected by Foldi, Bouchet and Leduc. With some modifications is still used today [5, 13].

Manual lymphatic drainage is a method of manual operation on the superficial lymphatic system which transports 90% of lymph. Specific movements of the therapist's hands result in slow, consistent with the physiology, movements of the lymph, without changing the temperature of the tissues [7].

MLD is based on four basic grips, called Vodder grips:

- Circular motion moves,
- Rotary grip,
- Pumping grip,
- Scooping grip.

All four of them are used in specific area of the body. The strength and the manner of the movements, depend on the body part the massage is performed at; force applied to the lower limb differs from that used for the upper limb or neck.

MLD therapy is commenced with the stimulation of healthy lymphatic vessels and nodes above the area covered by oedema, considering the direction of lymph flow. The grips are not to be used more than 5-7 times in the same part, so to prevent skin overheating. The movements are normally performed in a 1 second rhythm. Each grip has two

components: a shifting phase, during which lymph production, movement of lymph and draining increases, and relaxation phase, during which the emptied vessels perform their suction action. Contrary to common belief, sliding movements are the least effective form of the drainage therapy, they should therefore be avoided. For this therapy no oils are necessary as hands should not slide on the skin. In most areas the grips are to be done with a full hand, laid flat, parallel to the patient's skin. The general aim of manual lymphatic drainage is emptying the most proximal segment of the lymphatic system and then letting it fill with the lymph held up on the lower levels, for example emptying the thigh, moving the oedema from the shin to the thigh, repeating emptying the thigh, working on feet, moving the oedema to the shin, and then from the shin to the thigh, etc. [44].

MLD should be performed 2-3 times a week for the first month and then 1-2 times a week [13].

Contraindications for MLD can be divided into two categories: systemic and local [39].

1. Systemic Contraindications:

a) Absolute:

- Heart failure;
- Recent venous thrombosis;

b) Relative:

- Acute skin inflammatory diseases;
- Swelling in active cancer.

2. Local Contraindications:

a) On the neck side:

- Hyperthyroidism;
- Carotid sinus hypersensitivity;

b) On the abdomen cavity side:

- Pregnancy;
- Menstruation;
- Ileus;
- Active phase of inflammatory bowel disease, including ulcerative colitis, Crohn's disease and irritable bowel syndrome;
- Aneurysms of large arteries in the abdominal cavity and post-their surgery condition;
- Advanced atherosclerotic lesions of the aorta;
- Advanced post-radiotherapy lesions;
- Post-thrombosis of the deep veins in the pelvis condition [7, 39].

Pressure Therapy

Pressure Therapy enables objective determination of the pressure the device has on the tissues of the swollen limbs [45, 46]. This method uses pneumatic tubes, divided into chambers, which are to be worn on the swollen upper or lower limbs. Chambers are sequentially filled with air, from the most extreme to the most proximal part of the limb, in an ascending wave. The pace of filling and emptying the chambers as well as the air pressure is determined on case by case basis,

depending on the oedema size and type. According to Olszewski, when determining treatment parameters elasticity of skin tissue should take into consideration. If the skin is hard and fibrotic, relatively high pressure of over 150 mmHg can be applied [46]. Other authors report that safe pressure should not exceed 60 mm Hg, as lymphatic vessels are delicate and fragile; higher pressure might result in tissue damage and consequently even higher level of lymph low irregularities[45].

Olszewski and his colleagues have shown that long-term pneumatic compression causes the formation of new lymphatic channels [47].

Bandaging

The importance of bandaging in the treatment of lymphoedema of the lower limbs was already recognised in antiquity, as evidenced by the paintings from over 4000 years ago found on Sahara's rocks. When using this treatment method an appropriate choice of the bandage type should be made, as well as correct technique and the clinical condition of the patient [13]. Currently short stretch bandages are primarily used. They generate resting pressure which remains low and well tolerated by patients when limbs are not mobile, as opposed to the high resting pressure generated by the more elastic bandages. They also create working pressure, produced primarily by the strength of muscles during physical exercise [7]. To treat lymphoedema, multi-layer bandaging (2-3 layers) is recommended [13].

In lymphatic oedema of the lower limb elastic bandages of short to medium level of elasticity as should be used as the pressure material (70-130%). If a bandaging is to last longer, it is a good practice to use one layer of 8 cm width and one layer of 10 cm width bandage up to the height of the knee joint. For the thigh a foam bandage (sponge) with a width of 12 cm should be used. Then a bandage of low elasticity can be used, 10 to 12 cm wide; the foam bandage underneath provides good protection against slipping. Other products that can be used include adhesive or sticky bandages which can extend in both directions and zinc or gel bandages (coagulating).

Bandages must be applied by a trained physiotherapist and changed every 24 hours [7].

Another form of treatment is pressure therapy and physiotherapy.

Pressure Therapy

Pressure Therapy enables objective determination of the pressure the device has on the tissues of the swollen limbs [45, 46]. This method uses pneumatic tubes, divided into chambers, which are to be worn on the swollen upper or lower limbs. Chambers are sequentially filled with air, from the most extreme to the most proximal part of the limb, in an ascending

wave. The pace of filling and emptying the chambers as well as the air pressure is determined on case by case basis, depending on the oedema size and type. According to Olszewski, when determining treatment parameters elasticity of skin tissue should take into consideration. If the skin is hard and fibrotic, relatively high pressure of over 150 mmHg can be applied [46]. Other authors report that safe pressure should not exceed 60 mm Hg, as lymphatic vessels are delicate and fragile; higher pressure might result in tissue damage and consequently even higher level of lymph low irregularities[45].

Olszewski and his colleagues have shown that long-term pneumatic compression causes the formation of new lymphatic channels [47].

Compression garments

Compression garments (compression sleeves, tights, stockings and knee-high socks) should be used in the second stage of the complex decongestive physiotherapy in the treatment of lymphoedema. It is important to choose products with the level of pressure adequate to the degree of swelling.

According to the German classification (RAL), there are four pressure levels:

- 1st degree (18-21 mmHg),
- 2nd degree (23-32 mmHg),
- 3rd degree (34-46 mmHg),
- 4th degree (>46 mmHg).

For upper limb oedema, 1st or 2nd degree of pressure is primarily used [7], while in the advanced stage of lymphoedema of the lower limb 3rd or 4th degree of compression is usually applied.

Compression garments should be flatly woven, not very elastic, allowing for the moisture to be moved away from the skin. It must be chosen by the physician or physiotherapist based on a precise measurement of the affected limb. Failure to select appropriate garments may mean that the effect obtained in the first stage of the therapy will not be sustained, or the oedema can even increase [7].

Contraindications of the compression therapy:

a) Absolute:

- Advanced peripheral arterial disease, including narrowing and occlusion;
- Congestive heart failure;
- Septic (infected) phlebitis;
- Phlegmasia coerulea dolens.

b) Relative:

- Seeping dermatitis;
- Inappropriate compression products;
- Allergic reaction to the material;
- Advanced peripheral neuropathy, eg. Diabetes;
- Primary chronic polyarthritis, including rheumatoid arthritis, chronic progressive rheumatism.

Physiotherapy

One of the pillars of decongestive therapy are physical exercises which trigger the muscle pump activating deeper lymphatic vessels. Patients should perform active exercises, optionally supported exercise should the swollen limb be too heavy. The limb during exercise should be bandaged or protected by compression garments. It must also be placed in a way facilitating effective lymph drainage [7]. Increased pressure in fascial compartments and the pressure exerted by the elements of the ligament system on the lymphatic vessels stimulate their own mobility, facilitating drainage of the stagnant lymph. Active muscle contraction and active-passive bending of the joints have similar effect on the vessels. On the other hand, the flow of lymph is not observed during isometric exercise [39].

Breathing exercises constitute another important element of the therapy. They create negative pressure in the chest, as well as affect the abdominal muscles, stimulating the lymph drainage from chyli receptaculum.

The patient should perform series of exercises of the affected limb several times a day, breathing exercises should be performed in-between them [7].

Therapeutic positioning

Therapeutic positions aim to facilitate the outflow of lymph from the swollen limb and should be used by the patient during sleep, rest and exercise. Patient with oedema of the lower limb should keep the leg above the hip, for example using a folded blanket or pillow, in a way that the whole length between the heel and the popliteal fossa is supported, bearing in mind that popliteal fossa lymph nodes should not be pressured. The best drainage position for upper limb is using the spline and holding it next to the chest [7].

Modified recommendations of the Italian Society of Phlebology for physiotherapy in the treatment of lymphoedema [17] are as follows:

Phase 1*Clinical condition:*

Swelling is soft, reduced after a night's rest. The diameter of the affected limb is 1-2 cm larger than the healthy limb. The skin is supple, with a positive Stemmer's sign. The patient claims not to have been suffering from lymphangitis in the past. The limb is functional and is only moderately affected by the condition. No lymph exudate observed.

Physiotherapy:

- a) MLD: two sessions per week, in cycles of 12-15 sessions, followed by supporting therapy (at least two sessions per year);
- b) Compression garments: elastic bandages creating pressure of 18-21 mmHg;
- c) Additional therapy: physiotherapy, therapeutic positioning carried out daily.

Phase 2

Clinical condition:

Swelling is soft, partially reduced after a night's rest. The diameter of the affected limb is 3-5 cm larger than the healthy limb. The skin is partially supple, with a positive Stemmer sign; no significant trophic disorders. The patient claims not to have been suffering from lymphangitis in the past, although individual episodes possible. The patient can use the limb in the normal or slightly restricted range. Lymphatic effusion is not observed.

Physiotherapy:

- a) MLD: two sessions per week, in cycles of 12-15 sessions, followed by supporting therapy (at least two sessions per year);
- b) Compression garments: elastic bandages creating pressure of 23-32 mmHg;
- c) Additional therapy: physiotherapy, therapeutic postural position out daily.
- d) Heat treatment: twice a day for 40 minutes.

Phase 3

Clinical condition:

Swelling is hard but supple, not reduced after a night's rest. The diameter of the affected limb is 5-8 cm larger than the healthy limb. The skin has lost its elasticity. It has a positive Stemmer sign, thickening of the skin on the outer surface of the limb can be observed. The patient confirms multiple episodes of lymphangitis in the past. The limb function is slightly reduced, there are certain difficulties in bending and straightening. Lymphatic effusion may be observed.

Physiotherapy:

- a) MLD: two sessions per week, in cycles of 12-15 sessions, followed by supporting therapy (at least two sessions per year);
- b) Compression garments: elastic bandages creating pressure of 34-46 mmHg;
- c) Additional therapy: physiotherapy, therapeutic positioning carried out daily.
- d) Heat treatment: three times a day for 40 minutes.

Phase 4

Clinical condition:

Swelling is hard, does not reduce after a night's rest. The diameter of the affected limb is over 8 cm larger than the healthy limb. The skin has lost its elasticity. It has a positive Stemmer sign, thickening of the skin on the outer surface of the limb can be observed together with orange peel syndrome. The patient confirms multiple episodes of lymphangitis in the past, with short remission periods. The limb function is greatly reduced, patient can hardly move the limb due to the swelling and fibrosis.

Physiotherapy:

- a) MLD: is not effective on this stage;
- b) Compression therapy: bandages and stockings of low elasticity creating pressure of 59 mmHg,
- c) Additional therapy: physiotherapy, therapeutic postural position is not effective at this stage;
- d) Heat treatment: three times a day for 40 minutes.

Phase 5*Clinical condition:*

Swelling is hard, limb deformed (elephantiasis), the swelling does not reduce after a night's rest. Deformation of osteoarticular structures can be observed. The skin has lost its elasticity. It has a negative Stemmer sign, large areas of thickened skin with the orange peel syndrome can be observed. No lymphatic effusion. Limb function is significantly reduced, the patient can hardly move it due to its increased mass and extensive fibrosis. This in turn leads to muscle atrophy.

Physiotherapy:

- a) MLD: is not effective on this stage;
- b) Compression therapy: bandages and stockings of low elasticity creating pressure of over 59 mmHg,
- c) Additional therapy: physiotherapy, therapeutic postural position is not effective at this stage;
- d) Heat treatment: three or four times a day for 40 minutes.

The number of sessions on each stage depends on clinical condition of the patient and the severity of oedema and it should be decided on by the treatment team. The treatment of lymphoedema in all five stages can be complemented by benzopyrone group medicines which reduce vascular permeability, as well as are proteolytically active, reducing oedema [48, 49, 50]. If the patient experiences infectious complications, antibiotics should immediately be introduced, for example penicillins and cephalosporins. The use of diuretics is not recommended, as they support elimination of water rather than proteins [51]. Stage 3, 4 and 5, may qualify for surgical treatment [17]. Surgery may be recommended in some cases in order to restore the outflow of the tissue fluid and lymph from the limb. Most commonly micro-surgical lymphoreticular anastomosis is performed and the removal of persistent (recurrent) swollen tissues, in the cases of significant degree of limbs deformation and chronic bacterial complications [4]. The only absolute indication for surgical treatment is diagnosis of angiosarcoma, an aggressive form of the tumour growing within longtime lymphoedema [7].

Summary

Lymphedema is a chronic disease, requiring a specialised treatment based on a thorough knowledge of anatomy, physiology and pathophysiology of the lymphatic system. Quality of life of the patients with lymphoedema depends on early dia-

gnosis and availability of information and treatment options best meeting the needs of the patient. Patient's acceptance and thorough understanding of the implemented treatment strategies is the key to therapeutic success and indicates the ideas of how patient's current model life style needs to be modified in order to reduce the risk of recurrence or exacerbation of symptoms. Comprehensive physical therapy has been proven to bring the best results in the conservative treatment of lymphoedema.

Adres do korespondencji / Corresponding author



Teresa Pop

Instytut Fizjoterapii Wydziału Medycznego
Uniwersytetu Rzeszowskiego
e-mail: mailto:apopter@interia.pl

Piśmiennictwo/ References

1. Skandalakis J.E.: I wish I had been there: highlights in the history of lymphatics. *Am. Surg.* 1995; 61: 799-808.
2. Wilting J., Neeff H., Chris B.: Embryonic lymphangiogenesis. *Cell Tissue Res* 1999; 297: 1-11.
3. Maśliński S., Ryżewski J.: Patofizjologia. Wyd. Lekarskie PZWL, Warszawa 2002; 376-379.
4. Olszewski W.: Choroby naczyń chłonnych. W: Pasierski T., Gaciąg Z., Torbicki A., Szmidt J.(red.): Angiologia. Wyd. Lekarskie PZWL, Warszawa 2004; 59: 472-480.
5. Chęciński P.(red.): Choroby naczyń – wybrane problemy. Wyd. Medyczne Termedia, Poznań 2006; 12: 177-193.
6. Bergan J.J.: Lymphoedema: introduction and clinical evaluation. W: Handbook of venous disorders, guidelines of American venous forum. Chapman-Hall Medical, North Way 1996; 33: 569-579.
7. Jaszczur I., Batorycka M., Rybak Z., Wrzosek Z.: Aktualny stan wiedzy na temat leczenia obrzęków limfatycznych. *Przegląd Flebologiczny* 2012; 20(1-4): 19-27.
8. Foldi M., Casley-Smith J.R.: Lymphangiology. Schattauer Verlag, Stuttgart-New York 1983.
9. Rutili G., Arfors K.E.: Protein concentration in interstitia and lymphatic fluids from the subcutaneous tissue. *Acta Physiol. Scand.* 1989; 99: 1-8.
10. Feletou M.: The Endothelium Part 1: Multiple Functions of the Endothelial Cells-Focus on Endothelium-Derived Vasoactive Mediators. San Rafael: Morgan & Claypool Life Sciences 2011.
11. Sutkowska E., Gil J., Stembalska A., Hill-Bator A., Szuba A.: Novel mutation in the FOXC2 gene in three generations of a family with lymphoedema-distichiasis syndrome. *Gene* 2012; 498(1): 96-99.
12. Sutkowska E., Bator A., Trompeta K., Szuba A.: Different lymphoscintigraphic patterns in patients with lymphoedema distichiasis. *Lymphology* 2010; 43(2): 73-77.
13. Ruciński A., Janczak D., Szyber P.: Etiologia, rozpoznanie i leczenie obrzęku limfatycznego kończyn dolnych. *Terapia* 2000; 8(8): 43-46.
14. Ruciński A., Pupa A., Rybak A.: Pourazowy obrzęk limfatyczny kończyny dolnej. Opis przypadku. *Przegląd Flebologiczny* 2003; 11(4): 107-109.
15. Cavezzi A., Michelini S.: Phlebolympoedema. From diagnosis to therapy. P.R.Comm. Bologna 1998.
16. Antignani P.L., Poli L., Amato B., Riba U.: Il Duplex scanner ed il color Doppler nella patologia vascolare. Torino: Centro Scientifico Editore, II edizione, 1998.
17. Agus G.B., Allegra C., Arpaia G., Botta G., Cataldi A., Gasbarro V., Mancini S.: Włoskie Towarzystwo Flebologiczne: Zalecenia diagnostyczne i lecznicze w chorobach żył i naczyń chłonnych. Blackhorse Publishing International Sp. z o.o., Warszawa 2002; Standardy diagnostyki i leczenia chorób naczyń chłonnych: 93-109.
18. Marotel M., Cluzan R., Ghabboun S. i wsp.: Transaxial computer tomography of lower extremity lymphoedema. *Lymphology*, 1998; 31: 180-185.
19. Case T.C., Witte C.L., Witte M.H. I wsp.: Magnetic resonance imaging in human lymphoedema: comparison with lymphangiography. *J. Mag. Reson. Imaging*, 1992; 10: 549-558.
20. ISL, Executive Committee: The diagnosis and treatment of peripheral lymphoedema. Consensus Document, *Lymphology* 1995; 28: 113-117.
21. Taylor G.W., Kinmonth J.B., Rollinson E. et al.: Lymphatic circulation studied with radioactive plasma protein. *B. Med. J.* 1957; 1: 133-137.
22. Kinmonth J.B.: Lymphangiography in man: a method of outlining lymphatic trunks at operation. *Clin. Sci.* 1952; 11: 13-20.
23. Vettorello G.F., Gasbarro V. et al.: L'ecotomografia dei tessuti molli degli arti inferiori nella diagnostica non invasiva dei limfedema. *Minerva Angiol.* 1992; 17: 1-3, 35.
24. Pecking A., Cluzan R.: Explorations du systéme lymphatique: epreuve au bleu, lymphographies directs, lymphoscintigraphies, autres methodes. *Encycl. Med. Chir. (Elsevier, Paris) Angiologie* 1997; 5: 19-1130.
25. Bollinger A., Insemer I., Franzeck U.K., Jager K.: Fluorescence microlymphography in various forms of primary lymphoedema. In: Bollinger A.G., editor: The initial lymphatics: new method and findings. Stuttgart, Thieme Verlag 1985: 140-146, 218-219.
26. Allegra C., Bartolo M.Jr., Sarcinella R.: Morphological and functional characters of the cutaneous lymphatic in primary lymphoedema. *Eur. J. Lymph.* 1996; 6: 24.
27. Głowiczki P.: Surgical treatment of chronic lymphoedema and primary chylous disorders. In: Rutherford R.B.(ed.), *Vascular Surgery*, 6th ed. Philadelphia: Elsevier Saunders; 2005. pp. 2428-2437.
28. Rybak Z., Wojciechowski R., Rybak W.: Dwudziestoletnia obserwacja chorej z jatrogennym uszkodzeniem układu chłonnego prawej kończyny dolnej. Skuteczność leczenia paliatywnej liposukcji chorej kończyny. *Przegląd Flebologiczny* 2005; 13(1): 25-27.
29. Sankowski A., Skórski M., Łoń S.: Operacje redukcyjne w leczeniu skrajnie zaawansowanych postaci przewlekłego obrzęku chłonnego. *Przegląd Flebologiczny* 2002; 10(1): 5-10.
30. Schaverien M.V., Munro K.J., Baker P.A., Munnoch D.A.: Liposuction for chronic lymphoedema of the upper limb- 5 years of experience. *J. Plast. Reconstr. Aesthet. Surg.* 2012; 65(7): 935-942.
31. Warren A.G., Bronson H., Slavin S.A.: Lymphoedema a comprehensive review. *Ann. Plast. Surg.* 2007; 59: 464-472.
32. Wiktor M., Chęciński P.: Obrzęk limfatyczny – niedoceniany problem kliniczny i terapeutyczny. *Praktyczna Fizjoterapia i Rehabilitacja* 2010; 10: 30-36.
33. Bouchet J.V., Richard C., Carpentier P.H., Franco A.: Reeducation en pathologie lymphatique. *EMC, Angiologie* 1997; 19-3660.
34. Clodius L., Foldi M.: Therapy for lymphoedema to day. *Int. Angiol.* 1984; 3: 207-213.
35. Oliva E., Sarcinella R.: Effectiveness of the physical therapy in lymphoedema's treatment. *Eur. J. Lymph.* 1996; 6: Sp. Co I, 23.
36. O'Donnell T.F., Howrigan P.: Diagnosis and management of lymphoedema. In: Bell P.R.F., Jamieson C.W., Rukley C.V.(red.): *Surgical Management of Vascular Disease*. WB Saunders, Philadelphia 1992; 1305-1327.
37. Casley-Smith J.R., Foldi M., Ryan T.J.: Lymphoedema. Summary of the 10th International Congress of Lymphology Working Group Discussions and Recommendations. Adelaide, Australia, August 10-17. *Lymphology* 1985; 18: 175.
38. McHale N.G., Hollywood M.A.: Control of lymphatic pumping: of Daflon 500m. *Phlebology* 1994, suppl.1: 23-25.
39. Zapalski S., Oszkinis G.(red.): Ambulatoryjne leczenie chorób żył kończyn dolnych. Gabriel M., Pawlaczek K.: Kompleksowa terapia fizykalna obrzęków limfatycznych. Wyd. Medyczne Via Medica, Gdańsk 2001; 20: 323-343.
40. Głowiczki P.: Treatment of secondary lymphoedema. In: Ernst C.B., Stanley J.C., editors: *Current Therapy in Vascular Surgery*, 2nd ed. Philadelphia: B.C.Decker, 1991: 372-378.
41. Foldi M., Kubik S.: Lymphologie. Wyd. 3. Gustav Fischer Verlag. Stuttgart 1993; 469-526.
42. Winiwarter F.: Die chirurgischen Krankheiten der Haut und des Zellgewebes, Billroth Chr., Deutsche Chirurgie, Lieferund 23. Verlag Ferdinand Enke, Stuttgart 1892; 152-292.
43. Wittlinger G., Wittlinger H.: Textbook of Dr. Vodder's manual lymph drainage (5th ed.). Brussels, Belgium: Haug International 1992.
44. Foldi M., Strossenreuther R.: Podstawy manualnego drenażu limfatycznego. Urban & Partner, Wrocław 2005.
45. Sutkowska E. et al.: Choroby układu limfatycznego. In: Sieroń A. (ed): *Podręcznik angiologii*. Bielsko-Biała: Alfa Medica Press; 2009. pp. 189-197.
46. Taradaj J.: Pneumatyczny drenaż limfatyczny. *Rehabilitacja w Praktyce* 2009; 2: 36-37.
47. Olszewski W.: Obrzęki limfatyczne kończyn – klasyfikacja, diagnostyka i leczenie. Materiały do konsensusu. *Przegląd Flebologiczny* 2005; 13(5): 1-8.
48. Krukowska J., Terek M., Macek P., Woldańska-Okońska M.: Metody redukcji obrzęku limfatycznego u kobiet po mastektomii. *Fizjoterapia* 2010; 18(4): 3-10.
49. Casley-Smith J., Casley-Smith J.R.: Hight-proteins oedemas and Benzopyrones. Sidney and Baltimore: Lippincott, 1968.
50. Casley-Smith J.R., Morgan R.G., Piller N.B.: Treatment of lymphoedema of the arms and legs with 5,6-benzo-[alpha] pyrone. *New Engl. J. Med.* 1993; 329: 1158-1163.
51. Casley-Smith J., Casley-Smith J.R.: Modern treatment for lymphoedema. Adelaide, Australia: LAA Ed., 1994.