The integrated and interdisciplinary treatment of chronic lymphedema

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Abstract

Lymphedema is a chronic and incurable disease, deeply disturbing physical and psychical health of affected individuals. Lymphedema is a progressive, if left untreated.

Affected patients are looking for help between physicians and therapists of different specialities. Chronic lymphedema leads to emotional disorders, depression, even to suicidal attempts – especially in young patients. In rare cases it may lead to lymphangiosarcoma – ominous neoplasm with poor prognosis.

Complex interdisciplinary approach of the lymphedema team is the key to the success of lymphedema therapy. Optimal treatment of lymphedema requires close cooperation of all people involved in the therapeutic process including physicians, nurses, physiotherapists and psychologists. International Society of Lymphology (ISL) published its recommendations for lymphedema therapy, which are accepted in many countries around the world. Recommended by ISL physiotherapy consists of a manual lymphatic drainage, compressive bandaging, decongestive exercises and meticulous skin care. It is called a manual lymphedema treatment – complex physical therapy (MLT-CPT).

Key words: chronic lymphedema, therapy.

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A subject of chronic lymphedema is frequently omitted or placed on the margins of chapters devoted to cardiovascular disorders in medical textbooks. Its etiology is frequently insufficiently explained as a simple imbalance between capillary filtration, reabsorbtion and lymphatic drainage. Misunderstanding of the patomechanism of lymphedema and underscoring of significance of the problem of chronic extremity lymphedema are common between primary care physicians and specialists in oncology or cardiology.

Lymphedema is usually not a life threatening disorder and frequently regarded as price for life salvage in cancer survivors. Such a view probably resulted in relative lack of interest in clinical and basic research devoted to disorders of lymphatic systems. Fortunately nowadays one can notice significant change in this matter.

Lymphedema is a chronic and incurable disease, deeply disturbing physical and psychical health of affected individuals [1]. Lymphedema is a progressive, if left untreated [2].

Lymphedema is a global problem. Number of people affected with lymphedema is estimated on 140 millions around the world [3]. The real number is probably higher due to insufficient diagnosis of lymphedema. Lymphedema is in many countries, including Poland, underdiagnosed and treated due to insufficient number of properly trained physicians. We do not have also statistical data on prevalence and icidence of lymphedema in Poland. Professor Waldemar Olszewski, a world renown expert in lymphology estimates anywhere between ten and twenty thousands people with lymphedema in Poland [4]. In neighboring Germany the estimates are around 120 000, including 40 000 with primary lymphedema and 80 000 with secondary lymphedema. The most common causes of secondary lymphedema in developed countries are cancer therapy (surgery and radiation, other iatrogenic and noniatrogenic injuries, infections and metastatic cancer [5].

Lymphedema is frequently classified according to its etiology into primary and secondary lymphedema [1]. Primary lymphedema can be further divided into: congenital lymphedema – present at birth, caused by congenital abnormalities

of lymphatic vessels (e.g. aplasia, hypoplasia, megalymphatics, chylous reflux); lymphedema precox – which appears at puberty, rarely occurring as familiar disorder (Meige's disease); lymphedema tarda – that appears after age of 35.

Primary lymphedema usually affects lower limbs (94%), upper extremities (3%), genitals (3%), head (0.5%). Secondary lymphedema is usually diagnosed in upper extremities – postmastectomy lymphedema (66%), lower extremities (31%), genitals (1.5%), head (1%) and breasts (0.5%) [5].

Causes of secondary lymphedema include: surgical lymphadenectomy, injury of lymphatics vessels (mechanical or caused by radiation), bacterial and parasitic infections, lymphatic obstruction (external compression by tumor, fibrosis).

Insufficient lymphatic drainage with initially normal capillary filtration is a primary defect in lymphedema. In other types of extremity edema (venous, cardiac, nephritic) capillary filtration is increased. Lymph stasis may result not only from anatomical defects of lymphatics (aplasia, hypoplasia, megalymphatics), but also may be caused by impaired lymphatic contractility. In chronic lymphedema, regardless its etiology, we observe impaired lymphatic drainage and hypertrophy of skin and connective and fatty tissue in the subcutis [6]. Chronic lymphedema causes secondary musculo-skeletal disorders with impaired limb mobility and strength, recurrent infections (cellulitis) requiring antibiotics and frequently hospitalization, leading to permanent disability.

Affected patients are looking for help between physicians and therapists of different specialities. Chronic lymphedema leads to emotional disorders, depression, even to suicidal attempts – especially in young patients. In rare cases it may lead to lymphangiosarcoma – ominous neoplasm with poor prognosis [7].

Optimal treatment of lymphedema requires close cooperation of all people involved in the therapeutic process including physicians, nurses, physiotherapists and psychologists [2].

Physicians are responsible for the diagnosis of lymphedema, therapeutic plan and supervision of physiotherapy. Pharmacoterapy has only a supplementary role in therapy of lymphedema. Lymphotropic drugs include flavonoids, which were shown to increase lymphatic contractility, decrease capillary permeability and inhibit inflammatory reactions. Diuretics are not recommended, due to its non physiological action and quick rebound of edema.

Surgical therapy is recommended only in a subset of patients with severe lymphedema and no improvement after conventional physiotherapy. For patients with severe lymphedema (elephanthiasis) various modifications of 'debulking' surgery were described [8]. Microsurgical reconstruction of lymphatic drainage, introduced by professor Waldemar Olszewski, can be used in limited cases where afferent lymphatic trunks are functional. Microsurgery can be used separately or in conjunction with 'debulking' surgery. Unfortunately lymphatic anastomoses are relatively short living and many surgeons reported occlusion of majority of lymphatic anastomoses within one year after surgery. It is necessary to use compression garments after the surgery to improve lymphatic outflow and prevent recurrence of lymphedema [9].

Meticulous skin care is indispensable, and should be taught and supervised by the team nurse. Proper skin care is essential in prevention of skin infections. Trained nurses can also apply compressive dressings.

Psychologist is an important member of the lymphedema team. Patients with chronic lymphedema are known to suffer from multiple psychological problems ranging from low-self-esteem, to sexual disorders and depression. Quality of life of cancer survivors with lymphedema is much lower that quality of life of breast cancer survivors without this problem [2].

Physiotherapist is the crucial element of the therapeutic team. Physiotherapy was shown to be very effective and safe in chronic lymphedema. Relative simplicity and cost-effectiveness makes physiotherapy the most common therapy for both secondary and primary lymphedema. Physiotherapy is regarded by some authors to be the most effective therapeutic approach for patients with lymphedema [10]. In the polish literature [11] recommended physical therapy for patients lymphedema includes: classic manual massage, pneumatic massage, water massage, whirlpool therapy, breathing exercises, decongestive exercises, limb elevation and electrical stimulation. Compressive garments are also recommended. A value of intermittent pneumatic compression is pointed out [12].

International Society of Lymphology (ISL) published its recommendations for lymphedema therapy [13], which are accepted in many countries around the world. Recommended by ISL physiotherapy consists of a manual lymphatic drainage, compressive bandaging, decongestive exercises and meticulous skin care. It is called a manual lymphedema treatment – complex physical therapy (MLT-CPT). Manual lymphatic drainage (MLD) is a gentle manual technique (very different from a classic massage) in which by means of delicate skin touching and finger movements lymphatic drainage is stimulated, regional lymph stasis is alleviated, and lymphatic regeneration is accelerated. Manual lymphatic drainage was first mentioned by Alexander von Winiwarter over 100 years ago. In 1932 Emil Vodder developed his own technique of manual lymphatic drainage and presented it in 1936 at The International Health and Beauty Exhibition in Paris [14]. Special maneuvers of MLD are applied along lymph drainage pathways. MLD is performed in a specific sequence, starting from supraclavicular areas bilaterally, and then following distally. Slight increase of local tissue pressure during MLD helps filling initial lymphatics. Gentle tissue pulling activates contractions of lymphatic trunks. Lymphatic system activation is helped by stimulation of vagal nerve during long and gentle massage [14].

Compression helps to maintain positive MLD results. Importance of compression was known in ancient times, as documented by 4 thousands years old paintings on the rocks of Sahara [15]. Compression helps to avoid lymph stasis decreasing capillary filtration, improving lymphatic transport and improving function of venous muscular pump. It also helps in remodeling of thickened fibrous tissue. Application of compression dressing requires sophisticated technique, special low-stretch bandages and proper assessment of patients health status to avoid complications. Multilayered bandaging with low and medium stretch bandages is recommended in lymphedema [15]. Compression garments (sleeves, stockings etc.) are used to maintain the results of MLT, and are much more comfortable

Table 1. **Lymphedema therapy according to the disease stage [20]**

Stage	Symptoms	Phase I intensive	Phase II optimalization	Phase II b maintanence
Stage 0	Asymptomatic stage (possible detection with special imaging techniques only)	–	–	–
Stage I	A soft edema, reversible with eleveation or after a night rest	MLD: 1 x daily, compression banadging, decongestive exercises; duration of treatment 14-21 days	–	MLD: in cycles, exercises, compression: compressive garments occasionally or permanently
Stage II	An organized edema with tissue proliferation, no help with elevation	MLD: 2 x daily, compression banadging, decongestive exercises; duration of treatment 24-28 days	MLD: 1-2 x a week for 2-5 years, compression therapy: compressive garments, bandaging, decongestive exercises; Intesive phase should be repeated 2-3 x	MLD: in cycles or 1 x a week, decongestive exercises, compression: compressive garments permanently
Stage III	A hard, organized edema with extremity deformation (elephanthiasis), frequently skin changes (inflammation, severe hyperkeratosis, ulcerations)	MLD: 2-3 x daily, compression: banadging, decongestive exercises; duration of treatment 28-35 days	MDL: 2-3 x a week for 5-10 years, compression therapy: compressive garments, bandaging, decongestive exercise; Intesive phase should be repeated 3-8 x, consider surgical treatment	MLD: in cycles, or 1-2 x a week, decongestive exercises, compression: compressive garments permanently

MLD = manual lymphatic drainage, meticulous skin care is mandatory in all stages of lymphedema

than bandaging. These garments provide proximally decreasing compression of the extremities, and are available in four compression classes. Garments should be carefully chosen for the individual patients, and occasionally have to be custom made. Wear-off time differs for different garments, but generally they have to be replaced every 6 months.

Intermittent sequential compression utilizes pneumatic one or multichamber sleeve and a special pump to deliver intermittent pressure to the extremity. Chamber pressures, inflation and deflation time are regulated and individually adjusted depending on the type of edema and patient's tolerance. Usually compression time is longer than deflation time, and generally maximal chamber pressure should not be higher than patients diastolic pressure. For harder, fibrotic edemas, compression times are usually shorter and pressure lower. It should be remembered that high chamber pressure can damage delicate skin lymphatics [12]. Guidelines of The German Lymphological Society [16] do not recommend intermittent pneumatic compression as a sole therapy, and if prescribed should be always preceded by manual lymphatic drainage, otherwise it is not effective and can lead to several complications [17,18].

Decongestive exercises are important in lymphedema therapy [19]. Specific exercises improve lymphatic transport in edematous extremities (isometric, respiratory), enhance function of muscular venous pump (active), improve lymph transport in proximal lymphatic trunks (isometric and active neck exercises) and in a thoracic duct (respiratory exercises). All the exercises should performed in positions allowing help of gravitation. Exercise should not be too intensive in order to avoid accumulation of lactic acid and mauscular hyperemia leading to aggravation of edema. Elevation of the extremity allows gravitational drainage of lymphatic fluid and can supplement physiotherapy of lymphedema.

Therapy of lymphedema requires close cooperation between the patient and physiotherapist and is crucial for satisfactory therapeutic results. Patients should be well educated in the chronic nature and undulant course of the disease. Positive motivation and patient awareness are crucial to the success of decongestive therapy. According to Boris [17], active cooperation between the patient and therapist improve results of the therapy by 51% in patients with a lower extremity lymphedema, and by 36% in patients with an upper extremity lymphedema. Results of physiotherapy in lymphedema depend on the clinical stage of the disease and a presence of coexisting disorders (e.g. venous obstruction, decongestive heart failure, malignancy, neurological disorders, arterial ischemia, clotting disorders, collagen diseases etc.). Number and frequency of treatments depends on the phase of therapy (intensive, maintenance) and stage of lymphedema (*Tab. 1*) [20].

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