Complex Decongestive Therapy in lymphedema: report from an Interdisciplinary Center

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Abstract

Lymphedema is a chronic and worsening disease due to an abnormal accumulation of liquids, with a high protein content in the interstitial space. The disease is characterized by an insufficient flow of lymphatic fluid, which manifests as edema, inflammation, and fibrosis, all the way up to the stiffening of the affected tissues. Because it's a chronic and increasing disease, the treatment is highly complex. The literature shows that the treatment must be multidisciplinary, and it is necessary to combine multiple techniques, such as manual lymphatic drainage, mechanical lymphatic drainage, elasto-compressive bandages, and other complementary techniques up to surgical treatment in the most advanced forms which are not responsive to physical therapy. Furthermore, the disease is characterized by episodes of cellulitis, that may lead to infectious complications because the lymphatic function becomes insufficient. A condition of local immunodeficiency is created due to the crucial role that the lymphatic system covers with immune defenses, therefore creating a fertile ground for infections caused by small skin wounds, insect bites, animal scratches, nail fungus, and blood draws. Therefore, particular attention is paid to skin folds and interdigital spaces for which hygiene is necessary by using neutral detergents, drying by dabbing, and applying emollient creams for skin hydration. Unfortunately, such practices of prevention and care are often underestimated. We provide education to the patient on self-care, such as the self-bandage and the correct application of the elasto-compressed stoking. For wrapping the bandage, it is advisable to wear a glove or a special sock. The multilayer bandages are used in the first decongestant phase, while over time, the elastic stocking is the best aid for the management of lymphedema. Our clinic is a referral center for diagnosis, treatment, and surgical therapy, where patients come for surgical evaluation. Since 2016, we have systematically collected clinical data and volume evaluation of more than 600 cases affected by lymphedema and lipoedema of all stages. They have been treated with complex decongestive therapy for 4 hours a day, 15 consecutive days, subsequently maintenance once a week for 3 months, then once a month for 6 months. Of more than 600 patients treated, only 150 were submitted to surgery (lymphatic venous anastomosis, fasciotomy or liposuction, chylothorax and chyloperitoneal shunt, reconstructive plastic of external genitalia).

Complex decongestive interdisciplinary therapy, when properly performed, can stabilize the lymphedema patient situation, reducing the stage and ensuring a good quality of life.

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Introduction

Lymphedema is a chronic disease with a progressively worsening evolution and appearance of recurrent complications; Dermato Lymphangio Adenitis (DLA) is responsible for a further rapid increase in the volume and consistency of the edema.

The conservative therapeutic methods of medical-physical rehabilitation adopted in the treatment of lymphedema of the limbs allow excellent results to be obtained whether performed by expert hands following precise treatment protocols.

Only patients refractory to conservative treatment should be referred for surgical treatment, a decision exclusively reserved for an experienced lymphologist.

In recent decades, the literature has shown that the advent of surgical techniques such as microsurgery and minimally invasive techniques, including autologous Vascularized Lymph Node Microsurgery (VLNT), Lymphatic Graft (Lympholymphatic Graft), Anastomosis Lymphatic Venous Vein (LVA) and the super-

ficial one performed in super microsurgery, have allowed positive and constantly prolonged results, but they must always be preceded and followed by medical-physical-rehabilitative therapy.

The purpose of the present article is to demonstrate how medical physical rehabilitation therapy is effective in treating lymphedema of the limbs, when is performed in highly experienced centers, so then only a small part of patients need surgery.vOur clinic is a referral center for the diagnosis, conservative treatment, and surgical therapy for lymphedema, where only physiotherapists with a diploma from a Vodder school can work, and are always supervised by an expert lymphologist. Since 2016, we have systematically collected the clinical data and centimeter evaluation of more than 600 cases affected by lymphedema and lipoedema of all stages, treated with complex decongestive therapy for 4 hours a day, 15 consecutive days, subsequently maintenance once a week for 3 months, then once a month for 6 months. Of more than 600 patients treated, only 150 were submitted to surgery (lymphatic venous anastomosis, fasciotomy or liposuction, chylothorax, and chyloperitoneal shunt, reconstructive plastic surgery of the external genitalia). Complex decongestive physical therapy, when properly performed, can stabilize the lymphedema patient situation, reducing the stage and ensuring a good quality of life.

Materials and Methods

Between 2016 and 2022, our center conducted a clinical study involving 600 patients with primary and secondary lymphoedema of the limbs undergoing intensive treatments (Table 1).

Inclusion and exclusion criteria

i) CDT, patients with clinical stage II or III of lymphoedema (ISL stage), lipoedema all stages; patients with NYHA 3 heart failure were excluded from the study; ii) surgery, patients who could not be stabilized after 2 cycles of intensive care or patients who had an absolute indication for surgical therapy, *e.g.* post-surgical lymphocele, lymphangioma, thoracic duct anomalies, plastic surgery of the external genitalia; patients with lipoedema were excluded from surgery.

The study included 606 patients, with a middle age of 54 years: 171 with lipoedema, 113 with lower limbs affection (99 women and 14 men), 58 with duplicated lipoedema of upper limbs (56 women and 2 men), 435 with lymphedemas, 125 primary lymphedemas, 119 lower limbs (73 women and 46 men), 6 upper limbs (2 men and 4 women). About primary lymphedemas, 80 cases were certified for rare disease with exemption code RGG020 (in our region, we have been authorized to issue specific exemptions since 2018), 25 of which underwent genetic testing. Two hundred and ninety-one with secondary lymphedema, 202 lower limbs (123 women and 79 men) for uterine and prostate cancer with lymphadenectomy, a minor part for melanoma with lymphadenectomy, 89 affected by lymphedema of the upper limbs, 82 women (mainly secondary lymphedema to breast cancer) and 7 men (2 for breast cancer and 5 for melanoma surgery).

Patients were classified according to the ISL consensus document: i) primary lymphedema, upper limbs 6 cases stage II, lower limbs 15 stage I, 92 stage II and 12 stage III; ii) secondary lymphedema, upper limbs 69 cases stage II, 20 stage III, lower limbs 171 stage II, 31 stage III. Nineteen patients had other pathologies of the truncular lymphatic system.

Methods

In our center, patients are subjected to a lymphological examination by a specialist with many years of experience. The diagnostic procedure includes the collection of anamnestic data, a clinical examination with venous and arterial Doppler ultrasound examination of the limbs, a complete ultrasound abdominal scan, and in case of suspected cancer pathology, tumor markers are prescribed. Each patient is discussed collectively with the members of the staff (physiotherapists with a Vodder School diploma). All patients underwent lymphoscintigraphic examination of the limbs, but for selected or complex cases, we address them to Magnetic Resonance Imaging (MRI) studies. Indications on skincare and how to prevent possible infectious episodes are always explained. At the beginning of the session, the therapist marks centimeter measurements of limbs, weight, and photographic documentation of the patient. A suitable diet is given to overweight or obese patients. Our protocol provides an intensive cycle of 15 days of Complex Decongestive Therapy (CDT). Each

Table 1. Patients' characteristics.

					ISL Stage		
				Cases	II Stage		
Lipoedema	Lower limbs	Women	99	113	113		
		Men	14				
	Lower limbs duplicated upper limbs	Women	56	58	58		
		Men	2				
				171	ISL Stage		
				Cases	I Stage	II Stage	III Stage
Lymphedema	Primary	Lower limbs	Women	73	15	92	12
			Men	46			
		Upper limbs	Women	6	0	6	0
			Men	0			
	Secondary	Lower limbs	Women	123	0	171	31
			Men	79			
		Upper limbs	Women	82	0	69	20
			Men	7			
	Other			19			
				435			
Total cases				606			

day includes 1 hour of manual lymphatic drainage according to Vodder, 1 hour of LPG Endermologyâ, 2 hours of intermittent pneumatic compression multichamber therapy, and a multilayer bandaging with short-stretch bandages. For patients with facial lymphedema, the protocol included 1 hour of manual lymphatic drainage and the Linforollâ method for one more hour. Patients who also presented web axillar syndrome or lymphatic drainage disturbed by scars, we combined the lymph taping.

At the end of the CDT, the patient's measurements are taken again, so the clinical response can be evaluated. The specialist then prescribes the correct elastic stocking with the right compression class, which is supplied by our trusted technician based on accurate measurements, so one or more customized elastic garments are packaged. Stabilized patients continue with 1 weekly session for a further 3 months and subsequently 1 session a month for 6 months, in our center or with a Vodder physiotherapist near the patient's residence. Patients refractory after at least 2 intensive cycles of CDT may be cases for surgery indications. In our center, the following surgical procedures are performed based on the residual edema and its component (fibrotic *vs* adipose) and on the lymphoscintigraphic study: i) one site deep Lymphatic-Venous Anastomosis (LVA) In the inguinocrural region for the lower limb and in the middle third of the arm for the upper limb, using the blue patent violet and operating microscope; ii) supermicrosurgery, with the use of indocyanine green, with multiple lymphatic venous anastomoses; iii) Minimally Invasive Endoscopic Fasciotomy (MIF), for cases where is not possible to perform lymphatic-venous anastomosis, or for oncological criteria; iv) fibrolympholiposuction, in patients with a higher percentage of residual adipose tissue; v) resective surgery, in patients where exuberant tissue remains at the end of the decongestant therapy, or in patients presenting asymmetrical lipodystrophy, resective plastic surgery of the external genitalia, removal of cystic lymphangioma, lymphocele; vi) treatment of chylothorax and chyloperitoneum, in patients with abnormalities of the thoracic duct or of the chili cistern, who are preventively undergoing hospitalization and total parenteral therapy and subsequently laparoscopic or thoracoscopic video surgery; vii) implantation of monoclonal cells, in patients with lymphedema and ulcerations.

Results

In our center, we treated all patients with CDT according to our protocol, and in order to evaluate the response to the treatment, we decided to use the ICF Disability Scale. The ICF is outlined as a classification that aims to describe the state of people's health in relation to each field of life (social, family, work) in order to highlight the difficulties that can cause disability in the socio-cultural environment of the patient. The ICF Disability Scale is a framework of predefined tables each patient is asked to fill out at the beginning and the end of the cycle of treatments.

Thanks to this methodology, we have highlighted the physical and psychological difficulties of the patient during the various stages of the disease. The aim was to verify the patient's improvements by comparing the results obtained based on the subjective responses they made. The questionnaires collected data about the disability degree that they express personally, demonstrating how the proposed treatments were able to improve psycho-physical and social conditions of the patient suffering from lymphedema and lipedema (Tables 2 and 3). In order to get the Disability Average Score, the patient has to express each skill with a score between 0 and 4. Then, the professionals calculate the total amount by adding each single score and dividing it by the total number of items. The number obtained is related to an index that

Table 2. ICF Upper Limb Questionnaire.

Skill	0	1	2	3	4
Lift and carry object					
Fine movement of the hand					
Vehicule					
Self-cleaning					
Caring of single parts of the body					
Dressing					
Preparing meals					
Housework					
Caring of house things					
Simple interpersonal interactions					
Complex interpersonal interactions					
Interacting with new people					
Formal relations					
Informal relations					
Family relations					
Intimate relations					
Job					
Free time					
Religion and spirituality					
Political activity					

Table 3. ICF Lower Limb Questionnaire.

Skill	0	1	2	3	4
Change body positions					
Keep body position					
Move in space					
Walking					
Vehicule					
Driving					
Caring of single parts of the body					
Body needs					
Dressing					
Housework, Caring of house things					
Simple interpersonal interactions					
Complex interpersonal interactions					
Interacting with new people					
Formal relations					
Informal relations					
Family relations					
Intimate relations					
Job					
Free time					
Religion and spirituality					
Political activity					

describes the degree of disability (Table 4). All patients affected by lipoedema have been treated with CDT, and those with a Body Mass Index (BMI) higher than 25 had to follow a suitable diet combined with regular physical activity, getting benefits from both decongestant therapy and weight loss (*Supplementary Figure 1*). All patients reported a reduction in pain, volume, and sensation of tension. About the patients affected by lymphedema, of all 435 who underwent CDT, 285 had a good reduction of volume, in some cases up to the complete normalization of the edema and reduction of cellulite episodes. All patients wore elastic stockings in the maintenance phase and still continued to take sessions of Vodder Manual Lymphatic Drainage, depending on the residual clinical stage (*Supplementary Figures 2-8*). The examples below show patients affected by lymphedema of limbs, before and after CDT. The 135 patients refractory to conservative therapy after two intensive cycles, 6 months apart, underwent surgery. The indication was placed on the basis of lymphoscintigraphy, the stage of the edema and the component of the edema itself (fibrotic/adipose tissue), the localization of the edema or the congenital anomalies, *e.g.* anomalies of the thoracic duct or chili cistern (Table 5). The patients subjected to the different surgical techniques benefited from the result by stabilizing the residual edema and, in many cases, improving the measurements of volume. Some patients underwent also a combined technique (*Supplementary Figures 9-18*). The results collected demonstrate that when the combined complex decongestant therapy is conducted in specialized referral centers, with a qualified team of professionals in collaboration with an expert lymphologist, it is profoundly effective in controlling the lymphedema and lipoedema process in the majority of cases (less than 20 percent of patients need for surgical therapy). Based on the data obtained using the ICF Score, the subjects under examination were able to express themselves, demonstrating the improvement of the psycho-physical and social conditions of their lives, and feeling great satisfaction with the treatments received (Table 5, Figures 1 and 2). We integrated the reading of the results also with the radar charts, for better consultation and understanding, especially for the patients, so they can easily recognize the changes. In this way, it is clear to see in a unique image how the general situation before the treatment was located mostly in the area of moderate disability, then after the treatment moved to a new area between minor and absence of disability, thanks to the changes of the data.

Conclusions

Lymphedema is a disease which is very difficult to manage for patients, mostly because of the disability that the disease manifest in their life. At the most evident level there are the physical changes of the body, especially with limbs that became swollen,

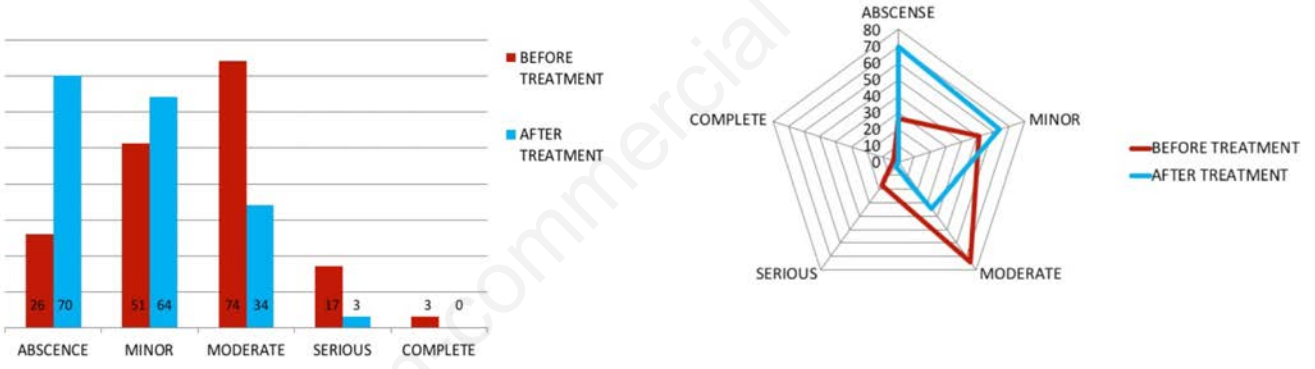

Figure 1. Results for ICF lipoedema.

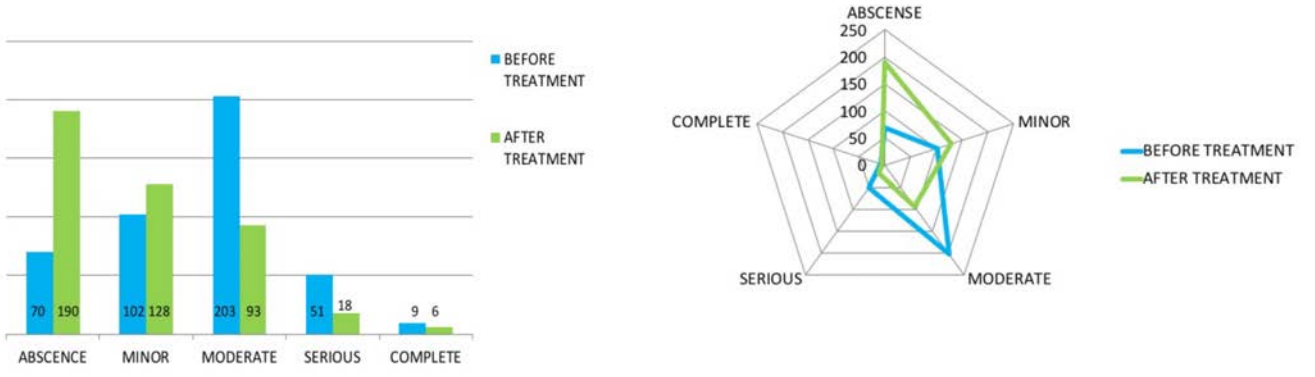

Figure 2. Results for ICF lymphedema.

Table 4. Disability Average Score.

0	ABSENCE OF DISABILITY 0-0,5	Patients do their activity with no restrictions in choosing, or problems in any function, even with orthoses
1	MINOR DISABILITY 0,6-1,5	Patients have minor disability in at least one activity
2	MODERATE DISABILITY 1,6-2,5	Patients have moderate disability in at least one activity
3	SERIOUS DISABILITY 2,6-3,5	Patients have serious disability in at least one activity
4	COMPLETE DISABILITY 3,6-4	Patients have complete disability in at least one activity

Table 5. Characteristics of patients.

N° patients	Type of surgery	Pathology
89	LVA one site	Lymphedema
9	LVA multiple	Lymphedema
2	VLT	Chylothorax
13	Resective surgery	Lymphocele post surgery
1	VLE	Chyloperitoneum
5	Plastic surgery of genitalia	Genitalia Lymphedema
20	Mini-invasive Fasciotomy	Lymphedema
10	Fibrolymphosuction	Lymphedema with fibrotic-adipose tissue
1	Monoclonal cell implantation	Lymphedema with ulcer
Tot. 150		

stiff, painful and easy to get infections and complications due to the immunological impairment of a decreased lymph flow. But beyond the physical manifestations there is a huge impact on the psychological level, in private and social life. Our study has finally demonstrated that a well conducted strategy of therapies, known as Complex Decongestive Therapy (CDT) performed by specialized professionals can lead to astounding results for the patients, in terms of both data and disabilities. A proper Decongestive Therapy is made first of all by the Manual Lymphatic Drainage that only a Vodder professional can execute in a precise way, thanks to the deep knowledge and sensitivity that is reached by the therapist in order to adapt each time to the different tissue that the hand finds. This can make a great difference. Vodder Manual Lymphatic Drainage associated with the right compression of bandages and stocking in the following stages is the real CDT that a patient has to find in a serious center. The real CDT has a fantastic number of demonstrations in the literature about the effectiveness and tangible improvement of the quality of life found by people affected by lymphedema or lipoedema disease.

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Online supplementary material:
Supplementary Figure 1. Patients affected by lipoedema at the lower limb in different steps while making Complex Decongestive Therapy (CDT).
Supplementary Figure 2. Breast cancer.
Supplementary Figure 3. Hysterectomy.
Supplementary Figure 4. Prostatectomy.
Supplementary Figure 5. Hysterectomy.
Supplementary Figure 6. Primary lymphedema.
Supplementary Figure 7. Primary lymphedema.
Supplementary Figure 8. Stabilization of lymphedema with elastic stocking.
Supplementary Figure 9. Lymphatic Venous Vein Anastomosis (LVA) one site with the microscope.
Supplementary Figure 10. Secondary lymphedema results one site Lymphatic Venous Vein Anastomosis (LVA) + Complex Decongestive Therapy (CDT).
Supplementary Figure 11. Mini-Invasive Fasciotomy (MIF).
Supplementary Figure 12. Secondary lymphedema results from multiple LVA + CDT.
Supplementary Figure 13. Primary lymphedema Mini-Invasive Fasciotomy (MIF) result.
Supplementary Figure 14. Plastic surgery of genitalia.
Supplementary Figure 15. Resective surgery.
Supplementary Figure 16. Fibrolymphosuction technique with PDE.
Supplementary Figure 17. Fibrolymphosuction with Complex Decongestive Therapy (CDT).
Supplementary Figure 18. Monoclonal cell implantation in primary lymphedema with ulcer, with PDE.