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First Dutch guidelines on lipedema using the international classification of functioning, disability and health

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Abstract

Introduction: Lipedema is a chronic, progressive condition that can result in considerable disability. In 2011, the Dutch Society of Dermatology and Venereology organized a task force to create guidelines on lipedema, using the International Classification of Functioning, Disability and Health of the World Health Organization.

Guideline development: Clinical questions on significant issues in lipedema care were proposed, involving (1) making the diagnosis of lipedema; (2) clinimetric measurements for early detection and adequate follow-up; and (3) treatment. A systematic review of literature published up to June 2013 was conducted. Based on available evidence and experience of the task force, answers were formed and recommendations were stated. The guidelines define criteria to make a medical diagnosis of lipedema, a minimum data set of (repeated) clinical measurements that should be used to ensure early detection and an individually outlined follow-up plan, pillars on which conservative treatment should be based and recommendations on surgical treatment options.

Conclusions: Little consistent information concerning either diagnostics or therapy can be found in the literature. It is likely that lipedema is frequently misdiagnosed or wrongly diagnosed as only an aesthetic problem and therefore under- or mis-treated. Treatment is divided into conservative and chirurgic treatment. The only available technique to correct the abnormal adipose tissue is surgery.

Recommendations: To ensure early detection and an individually outlined follow-up, the committee advises the use of a minimum data set of (repeated) measurements of waist circumference, circumference of involved limbs, body mass index and scoring of the level of daily practice and psychosocial distress. Promotion of a healthy lifestyle with individually adjusted weight control measures, graded activity training programs, edema reduction, and other supportive measures are pillars of conservative therapy. Tumescent liposuction is the treatment of choice for patients with a suitable health profile and/or inadequate response to conservative and supportive measures.

Keywords

Lipedema, guideline, obesity, international Classification of Functioning, disability and health, exercise, liposuction

Introduction

Lipedema is a chronic, progressive condition that can result in considerable disability, problems in daily functioning, and psycho-social difficulties especially if the condition is unrecognized and untreated.[1,2]

The diagnosis/definition of lipedema has not (yet) been registered in the International Classification of Diseases (ICD-10) of the World Health Organization (WHO). Given the lack of consistent diagnostic criteria, the prevalence of lipedema is difficult to establish. It is likely that lipedema is frequently misdiagnosed or wrongly diagnosed as only an aesthetic problem.

In this regard, in 2011, the Dutch Society of Dermatology and Venereology (NVDV) organized a task force to evaluate the current literature and propose evidence- and expert-based recommendations suitable for the national implementation of guidelines for the treatment of lipedema. The objective was to provide recommendations in the field of lipedema diagnostics,

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multidisciplinary treatment, and follow-up for professionals and patients.

The new guidelines embrace a functional, patient-centered approach, focusing on early diagnosis and a comprehensive follow-up with tailored treatment and support. Both the Chronic Care Model (CCM) of Wagner and the International Classification of Functioning, Disability and Health (ICF) of the WHO were used; the WHO and the Dutch government promote these models based on their proven effectiveness in the care of chronic conditions.

The ICF is a framework for documenting functioning and disability at individual and population levels. This framework provides a universal language to assess a person's functioning and disability and facilitates international comparisons of disability-related data. The basic concept of the ICF is based on a holistic framework of the interaction among body function, body structure, activity, and participation, which are subsequently related to environmental and individual factors (Figure 1).[3] The ICF method contains two parts. Part 1 describes functioning and disability and includes three components or domains: Body functions, body structures and activities and participation. Part 2 addresses contextual factors and includes two components: Environmental factors and personal factors, which influence the domains in part 1. Officially, "Personal Factors" is not described as a classification in the ICF given the significant social and cultural variety associated with these factors.[3]

The principle of the chronic care model (CCM) is active patient participation in treatment, patient empowerment and self-efficacy, and a more "hands-off," supportive approach by health professionals. Secondly, according the CCM, health professionals cooperate and form a so called network of care.[4]

Development of the guidelines

The development of guidelines is often a time consuming and challenging task. Initially, a preliminary meeting was held, and numerous individuals and organizations involved in both lymphedema and lipedema management were invited. The medical professions represented at the meeting included dermatology, surgery, radiology, psychology, physical therapy, dietetics, and skin therapy. In addition, representatives of several Dutch organizations were invited, such as the Federation of University Medical Centres, Dutch Society of General Practitioners (Huisartsen genootschap), Organisation of Nurses, Organisation of Nurses for Wound Care, several health insurance companies, the Ministry of Health Care, branch organizations, and patient representatives.

During this initial meeting, clinical questions of investigation that capture the most significant issues in lipedema care were proposed based on consensus. These questions involve 1. making the diagnosis of lipedema; 2. defining clinimetric measurements to ensure early detection and functional, holistic follow-up; and 3. patient treatment and support. Subsequently, a systematic analysis of English and German literature published up to June 2013, retrieved from PubMed, MEDLINE, COCHRANE, and Cinahl databases, was conducted. The method of evidence-based guideline development was employed based on the levels of scientific evidence (AGREE, http://www.agreetrust.org). Based on available evidence and the experience of the members of the task force, answers to the clinical issues were formed, and recommendations were stated. An initial draft of the guidelines was presented in December 2013, and the guidelines were finalized and published in April 2014.[5]

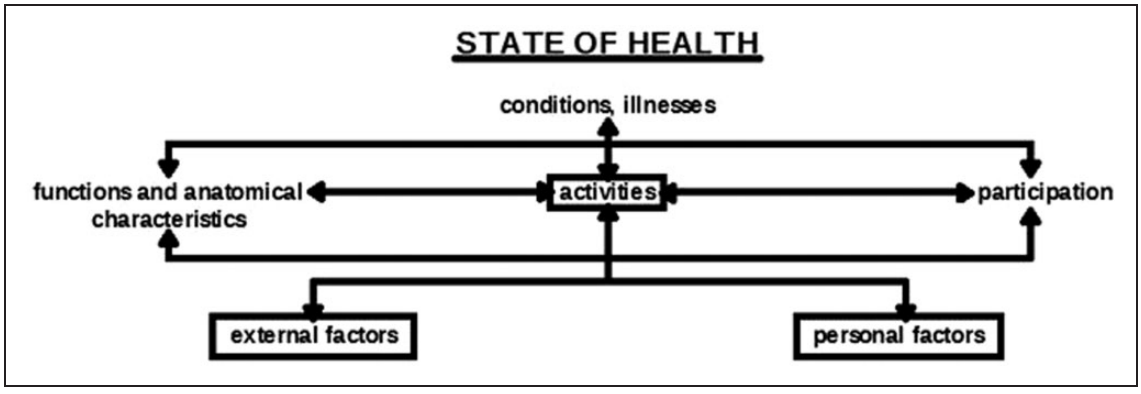

Figure 1. The interaction between the different aspects of state of health and external and personal factors.

Making the diagnosis

Given that lipedema is not an ICD10-recognized disease, increased confusion has been generated by wide use of the term "lipedema." The term lipedema, suggesting swelling caused by fluid, is somewhat confusing. Lipedema involves abnormal, disproportionate depositions of subcutaneous fat, leading to bilateral, disproportional enlargement of the legs and, in some cases, also the arms. The pathophysiological mechanisms involved in lipedema remain unclear.[6]

The diagnosis is made on clinical evidence and the exclusion of other differential diagnoses. Lipedema is frequently accompanied with (mild) edema, especially in more advanced cases and cases aggravated by obesity (generalized increase of subcutaneous fat).

Clinical characteristics

Lipedema generally affects women and often starts during puberty. There are only two male cases reported.[7,8] Substantial variability in disease severity and progression is noted. Some women develop minor lipedema, which stabilizes and does not progress over time. Other women exhibit a gradual progression of the lipedema, whereas a sudden exacerbation is occasionally provoked by a stressful situation, such as pregnancy or surgery, in other cases.[2] The most common complaints are sensations of heaviness and discomfort in the legs, with moderate to severe sensitivity to digital pressure. The swelling and pain worsens during warm weather and exercise and is not alleviated by limb elevation; however, elevation might improve the associated edema component slightly. Weight loss measures generally influence the obesity component but exhibit a minimal effect on the abnormal body fat distribution. A typical sign of lipedema tissue is a sharp separation between normal and abnormal tissues at the ankle (cuff sign of lipedema). Significant differences in prevalence and even existence worldwide are noted.

Differential diagnoses

The most relevant differential diagnoses of lipedema include obesity, lipohypertrophy, and lymphedema. In cases of more advanced edema, other causes of classical edema of the lower legs (chronic venous insufficiency, idiopathic cyclic edema, edema due to internal disease, and orthostatic edema) should be considered. It is difficult to distinguish among mild forms of lipedema, "normal" variations of fat, and lipohypertrophy. A more functional approach focusing on the limitations and disability of a patient is more rational.

Diagnostic criteria

The guideline committee advises the use of defined criteria to make a medical diagnosis of lipedema. The diagnosis can be made when the following criteria are met (Figure 2).

Clinimetrics

Given the variation of presentation and large role of subjective complaints in lipedema, attention to the diagnostic criteria alone is not enough to ensure early recognition and therapeutic advice, nor to outline a holistic, patient-centered follow-up program with tailored treatment. Repeated collection of objective clinimetric data on functioning and disability levels is necessary to scale disease severity and to detect specific individual needs. Therefore, the guideline committee wanted to define a minimum data set of (repeated) clinimetric measurements that should be performed in patients who are diagnosed with lipedema and in patients who are suspected to be at risk for developing lipedema (but may not (yet) completely achieve the diagnostic criteria).

In creating this data set, the committee embraced the approach of the "ICF model" of the WHO. Based on this model, disease-specific "core-sets" can be defined, specifying the clinimetrics that should be measured to identify disease-related disability and problems in daily functioning via a consistent and internationally comparable manner. Currently, no ICF core-set is available for lipedema, but suggested measurements that could be of value are depicted in Figure 3.

Currently, for the assessment of clinical signs in all lipedema patients (including individuals with a risk of lipedema), the guideline committee advises the use of a minimum data set of (repeated) circumference measurements of the involved limbs, body mass index (BMI), waist circumference, and the level of daily practice score. Given the disproportionate enlargement of the lower extremities, the body mass index and total body weight are suboptimal obesity parameters in lipedema patients. The guideline committee advises the use of waist circumference measurements to identify healthy weights. When the medical diagnosis is confirmed, the addition of the Distress Thermometer (DT), which is the English translated version of the Dutch "LAST meter" to the minimum data set is recommended to define the potential psychosocial distress of the patient, which often accompanies lipedema.[9]

Treatment

Given that the cause of lipedema remains unknown, lipedema cannot be cured. Treatment is primarily focused on reducing disability and subjective

DIAGNOSIS LIPEDEMA, THE WORK GROUP HAS ASSEMBLED A LIST OF CRITERIA, BASED ON CLINICAL EXPERIENCE AND SUPPORTED BY THE LITERATURE		
Diagnosis is certain when present: A1+2+3+4+5 PLUS ((B6+B7) or (C8+C9) or (D10+D11) or E12). In the absence of at most 2 of these five criteria (A to E), the presence of the extra criteria F13 or F14 also assures the diagnosis.		
		Anamnesis (A) (criteria of wold et al.)
A	1	Disproportionate fat distribution
	2	No / limited influence of weight loss on disproportionate fat distribution
	3	Easily in pain / bruised
	4	Sensitivity to touch / fatigue in extremities
	5	No reduction of pain when raising extremities
		Physical examination (B,C,D,E)
		Upper leg:
B	6	Disproportionate fat distribution
	7	Circularly thickened cutaneous fat layer
		Lower leg:
C	8	Proximal thickening of subcutaneous fat layer
	9	Distal thickened of subcutaneous fat, accompanied by slender instep (cuff-sign)
		Upper arm:
D	10	Significantly thickened subcutaneous fat layer in comparison with vicinity
	11	Sudden stop at elbow
		Lower arm:
E	12	Thickened subcutaneous fat, accompanied by slender back of hand (cuff-sign)
		Extra criteria
F	13	Pain when applying bi-manual palpation
	14	Distal fat tissue tendrils of the knee (popliteus)

Figure 2. Due to a lack of unambiguous criteria when establishing the diagnosis lipedema, the work group has assembled a list of criteria, based on clinical experience and supported by the literature.

complaints and the prevention of progression. This treatment goal incorporates target components that could influence these factors, such as edema, unhealthy lifestyle, obesity, lack of physical activity, lack of knowledge about the disease, and psychosocial distress.

Treatment is divided in conservative and surgical treatment. The promotion of a healthy lifestyle with individually adjusted weight control measures, graded activity training programs, vascular/lymphatic pump optimization, edema reduction, and other supportive measures, such as psychosocial therapy and orthopedic counseling, are important pillars of conservative therapy.[2,10,11] The only available technique to correct abnormal adipose tissue is surgery.[12–14]

Conservative Treatment

Stimulation of a healthy lifestyle: Weight normalization, exercise, and other supportive measures. Although lipedema patients are not generally obese, more than half of patients are overweight.[2] Diets and changes in nutritional behavior cannot reduce the disproportional lipedema tissue that is already present. However, obesity prevention is important because extra weight gain at affected body areas does not respond well to dieting and exercise.

Recommended weight control measures include the creation of an adapted diet plan based on individual caloric balance goals. If possible, this diet regimen should be combined with a weight loss exercise schedule.

In the Netherlands, a graded activity program is a widely accepted method to improve muscle strength and conditioning for various additional chronic medical conditions. Exercise is not only essential for weight control and general condition improvement but is also important to reduce the edema component by improving the calf muscle pump by increasing lymphatic drainage.

For some patients who experience the burden of lipedema on psychosocial functioning, psychological support measures should be offered.

Figure 4 reflects conservative treatment recommendations adjusted to patient-specific disease profiles, based on the clinimetric data set.

Disorders of functioning and anatomical characteristics / limitations in activity and participation	Measurement tool
Circumference	Measuring tape (+ mathematical formula) Perometer
Mobility	Goniometer
Strenght	Medical Research Scale (MRC)[&] Hand-held dynamometer
Pain	Visual Analogue Scale (VAS)[#]
Fatigue	Multi Dimensional Fatique index (MVI)[#]
Weight	BMI (Body Mass Index)* Abdominal circumference / clothing size and difference between upper and lower body
Activity	Dutch Standard of healthy daily activities (NNGB)[#] Short Questionnaire to Asses Health Enhancing Physical Activity (Squash)[@] LASA Physical Activity Questionnaire (LAPAQ)[$] Pedometer (also in the context of self-monitoring) Patient Specific Complaints List (Patiënt Specifieke Klachtenlijst or PSK)[$]
Condition / walking capacity	6 minute walking test[$] Submaximal exertion test Derived VO2 max test, i.e. Steep Ramp test
Quality of life	SF 36 health condition questionnaire[#]

& Access and explanation on: http://www.ucsdbglab.org/tools/MuscleScales.htm
**The often used term BMI often causes confusion in lipedema. The lipedema contributes to body weight therefore to increased BMI, which is used as a measurement for adiposity and obesity. Lipedema patients will therefore often have a raised BMI, without this having to be the cause of excess weight in the traditional sense. A strong discrepancy in clothing size between the upper and lower body is characteristic for lipedema patients.*
 # Access and explanation on: http://www.oncoline.nl/cancer-rehabilitation
$ Access and explanation on: http://www.fysionetevidencebased.nl/images/pdfs/guidelines_in_english/parkinsons_disease_practice_guidelines_2004.pdf
@ Access and explanation on: http://bmcmusculoskeletdisord.biomedcentral.com/articles/10.1186/1471-2474-9-141

Figure 3. Summary of clinimetric instruments suggested by the work group to be used when composing a health profile of patients with (suspected) lipedema.

Decongestive lymphatic therapy (edema reduction). Given that the combination of compression therapy and manual lymphatic drainage therapy (decongestive lymphatic therapy) is widely accepted as standard therapy for lymphedema, it has curiously also been generally accepted as standard therapy for lipedema. However, compression therapy and lymph drainage do not reduce adipose tissue. The guideline committee advises the use of a (flat knit) compression stocking only when an edema component is present; stockings can prevent additional edema formation and thus reduce resulting complaints of heaviness and pain.[2,11]

Manual lymphatic drainage therapy is not recommended in the treatment of lipedema because lymphological impairment is not observed in lipedema (in contrast with lymphedema), and the effectiveness of this technique is not proven.[2,11]

Surgery

Surgery is the only available technique to correct abnormal adipose tissue. There are two surgical treatments which have a place in the treatment of lipedema. The most performed treatment is liposuction.

Liposuction. Preceding studies have shown that liposuction under general anesthesia and/or without or relatively little subcutaneous infiltration is contra-indicated for lipedema, because of the substantial risk of causing damage to the lymphatic system.[1]

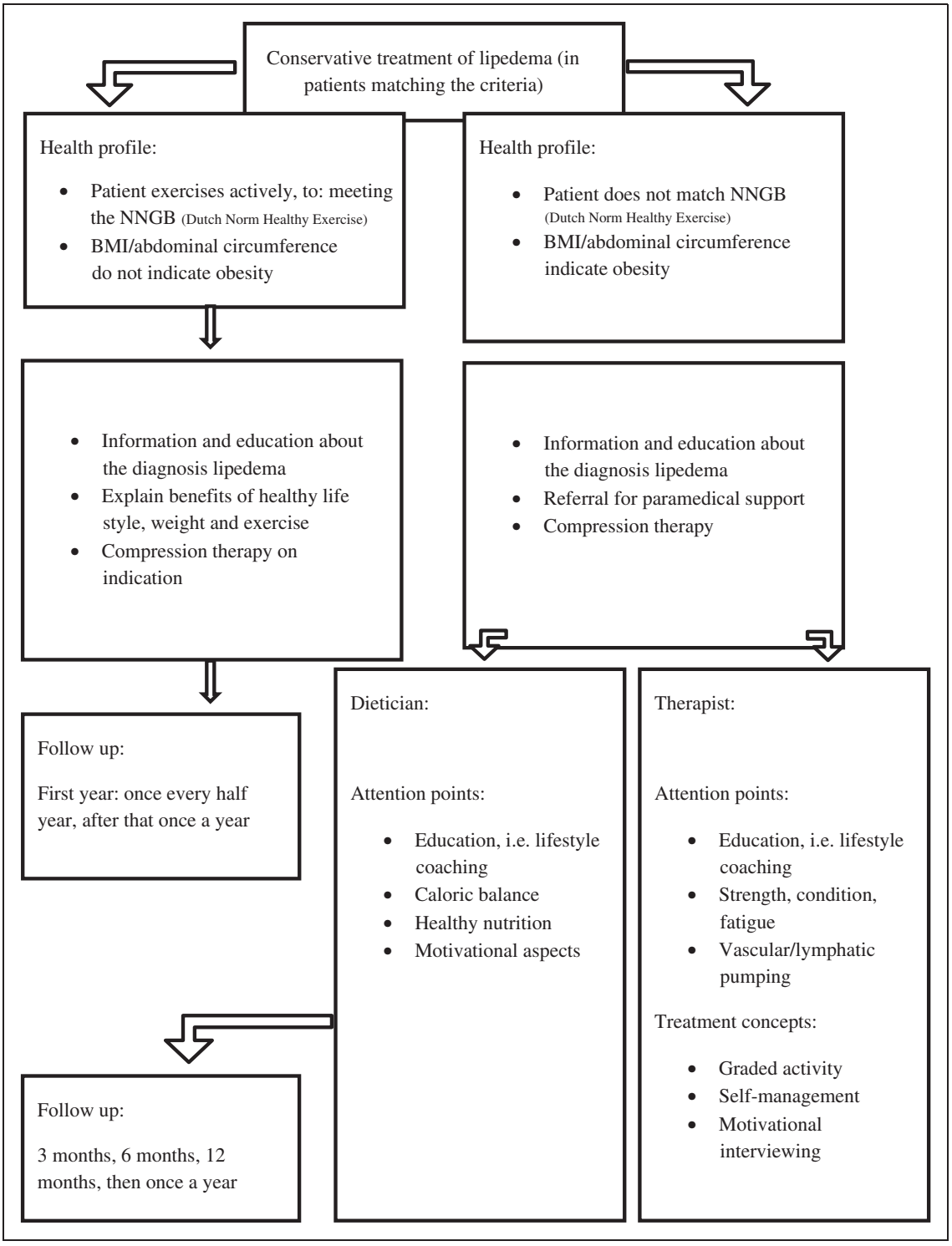

Figure 4. Flowchart conservative lipedema treatment.

With the introduction of tumescent local anesthesia (TLA) and subsequently supertumescent local anesthesia (STLA), the risk of damaging the lymphatic system during liposuction has dramatically decreased.

Tumescent liposuction (TLA) is a specialized technique, whereby local anesthesia is infiltrated in the subcutaneous tissues with large amounts of fluid (containing saline, lidocaine, sodium bicarbonate, and

adrenaline). The fluids are subsequently aspirated using vibrating microcannulas.

This technique has been proven to be a highly effective treatment for lipedema with positive morphological and functional long-term results, including the reduction of complaints, such as pain and bruising, and enhanced overall quality of life.[2,12–15]

Consistent criteria to determine the ideal time or patient characteristics for liposuction are not available. Some studies report better outcomes in early stages compared with advanced cases, but recent data demonstrate a more distinct change of complaints in more advanced cases.[12] Ideally, TLA is performed at a relatively early stage to prevent progression.[2,11]

The guideline task force recommends tumescent liposuction as part of the therapeutic armamentarium in the management of lipedema. However, tumescent liposuction is only the treatment of choice for patients with a suitable health profile and/or inadequate response to conservative and supportive measures. Before using TLA, associated deteriorating components, such as edema, obesity, unhealthy lifestyle, lack of physical activity, lack of knowledge about the disease, and psychosocial distress, should be addressed. Moreover, even after liposuction, women generally require conservative therapy, and weight normalization should remain a goal.

TLA requires specialized skills of the healthcare deliverer and should only be performed at a specialised center. Multiple sessions are often necessary to remove the extensive amount of adipose tissue, thus making the procedure time-consuming. This therapy is typically not supported by health insurance companies, making it an expensive treatment for the patient. Cost-effectiveness studies have not been performed. However, given that the need for conservative treatment is reduced after liposuction, health care costs could potentially decrease.

Excision. Large localized deposits of lipedematous tissue (''lumps'') can form in lipedema on the legs, with serious mechanical difficulties, valgus deformity of the knees or even inability to walk as a result. Sometimes longitudinal excision (lumpectomy) is the only option to remove the severe mechanical limitations.

Conclusions

Lipedema is a very challenging disease that is often misdiagnosed as lymphedema or obesity. Despite the lack of objective criteria for diagnoses and the understanding of pathophysiological mechanisms, lipedema is considered as an entity in its original form without the co-morbidity of obesity. Early recognition is crucial to prevent complications in the field of functionality, psychosocial morbidity, and the development of concomitant problems, such as obesity and lymphatic overload. The Dutch guidelines are a first step towards the improvement of diagnosis and a more rational treatment of lipedema as a chronic condition. The introduction of the systematic use of the ICF and CCM allows the patient to be treated on a more individual level based on limitations and to act more independently of health professionals.

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