



The Dutch lymphedema guidelines based on the International Classification of Functioning, Disability, and Health and the chronic care model

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ABSTRACT

Lymphedema is a chronic and progressive condition due to an imbalance between lymphatic filtration and transport capacity. A relative overload of fluid is caused by lymphatic impairment (afterload impairment); an absolute overload is caused by increased filtration (preload raised). Lymphedema can result in considerable disability and loss of quality of life. The Dutch Society of Dermatology organized a task force to create guidelines using the International Classification of Functioning, Disability, and Health.

As lymphedema is a chronic condition, a new approach was chosen according to the chronic care model.

In the development of the guidelines, the following topics of lymphedema care were proposed: (1) lymphedema detection; (2) clinimetric instruments; and (3) rational treatment modalities. A systematic review of the literature published up to June 2013 was conducted. Statements were based on scientific evidence and experience.

The guidelines propose recommendations based on an interdisciplinary approach to lymphedema using a continuum of care from prevention to initial treatment phase, maintenance phase, and follow-up. An integrated treatment program can be conservative or surgical. A therapeutic program depends on stage and origin of lymphedema; International Classification of Functioning, Disability, and Health-based limitations; needs of the patient; ability to perform self-management; and ability to reduce patient-related risk factors, such as overweight and lack of exercise. (*J Vasc Surg: Venous and Lym Dis* 2017;5:756-65.)

Lymphedema is a chronic and progressive condition that is caused by improper functioning of the lymphatic system. Relatively little is known about this phenomenon in medical science. As all interstitial fluid is transported by lymphatics, the overload of fluid can be relative, based on lymphatic impairment and dysfunction of the lymphatic system (afterload impairment), or absolute, based on an increased filtration rate (preload raised). Lymphedema can be congenital or secondary to conditions associated with increased filtration, such as a phlebologic impediment, or it can develop as a complication after medical (oncologic) treatment. Primary lymphedemas represent a heterogeneous group including sporadic, hereditary, and syndromal forms, and more and more genes are being recognized in association with lymphedema.¹

Secondary lymphedema is most frequently associated with cancer treatments by damage to the lymphatics or infection. In these cases, early diagnosis and treatment are essential to prevent the progression of the disease and its complications; late-stage lymphedema may cause severe physical and psychological problems for patients as a result of chronic swelling, impaired physical function, recurrent infections, and disfiguring skin changes.

Disease- and treatment-related risk factors for the development of lymphedema² are not modifiable by a patient. Awareness and programs to reduce risk factors such as obesity and lack of exercise,^{3,4} enhancing secondary prevention,⁵ are complex.

Lymphedema is generally accompanied by severe morbidity and a reduction in quality of life for the patient. Nevertheless, a high degree of unfamiliarity with the condition persists with health care workers, as a result of which lymphedema is often not detected or is detected only at an advanced stage. Moreover, care is often fragmentary because it is distributed among many different disciplines. These factors affect the quality of patients' care negatively. Care providers should be accustomed to the demands of the patients as much as possible. Regulatory coordination ought to be the guiding principle. Standardization of diagnostics, treatment, effect measuring, and follow-ups would benefit both the patient and the therapist and health insurer. In addition, this standardization and clarification lead to better opportunities for scientific research. Treatment advice will be tailored to the patient. The protocol holder

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will be the spokesperson for the patient in most cases. In case of surgery for lymphedema, it is fully integrated in the program for the initial treatment phase.

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 lymphedema management were invited. The medical professions represented at the meeting included dermatology, oncology surgery, radiology, psychology, physical therapy, dietetics, and skin therapy. In addition, representatives of 17 Dutch organizations were invited, such as the Federation of Medical Specialties, the Dutch Society of General Practitioners, the Organization of Nurses and wound care specialists, several health insurance companies, the Ministry of Health Care, branch organizations, and patient representatives.

During this initial meeting, clinical procedures that capture the most significant issues in lymphedema care were proposed on the basis of consensus. These procedures involved (1) making the diagnosis; (2) defining measurements to ensure early detection and functional, holistic follow-up (clinimetrics); and (3) treating and supporting the patient. Subsequently, a systematic analysis of English and German literature published up to June 2013, retrieved from PubMed, MEDLINE, Cochrane, and Cumulative Index to Nursing and Allied Health Literature databases, was conducted. The method of evidence-based guideline development was employed according to the levels of scientific evidence (Appraisal of Guidelines for Research and Evaluation, <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 made available in April 2014, with a version for patients and a full professional version since 2015.

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 of Wagner⁶ and the International Classification of Functioning, Disability, and Health (ICF) of the World Health Organization⁷ were used; the World Health Organization and the Dutch government promote these models on the basis of their proven effectiveness and positive economic impact⁸ in the care of chronic conditions.^{9,10}

ICF

The ICF is a framework for measuring and 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

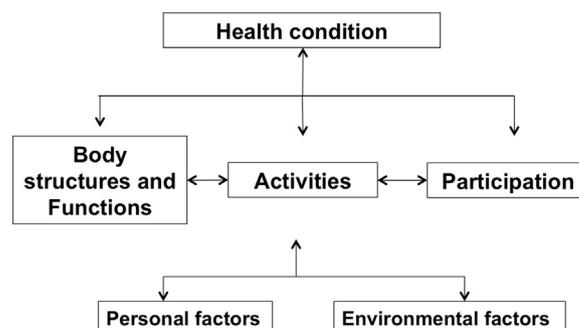


Fig 1. The interaction between the different aspects of state of health and external and personal factors. (International Classification of Functioning, Disability, and Health: ICF. Geneva: World Health Organization; 2001.)

concept of the ICF is based on a biopsychosocial model. The holistic ICF framework describes the interaction among body function, body structure, activity, and participation, which are subsequently related to environmental and individual factors (Fig 1). 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, that 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.¹¹ The full ICF described >1400 categories in the mentioned domains, which is not very applicable for daily practice. Therefore, a more comprehensive core set specific for lymphedema needs to be developed.¹²⁻¹⁴ Working with the ICF in a chronic condition will improve the functioning and quality of life of patients in association with and not merely as a consequence of their health condition.^{15,16}

CHRONIC CARE MODEL

The chronic care model centers on pre-existing or long-term illness, as opposed to acute care, which is concerned with short-term or severe illness of brief duration. The model was initially proposed by Wagner⁶ in 1988 in response to the acknowledgment by health plans and provider groups that the care of patients with chronic illness required improvement. Evidence has shown that "usual care" is not effective for management of a chronic condition; sizable numbers of chronically ill patients are not receiving effective therapy, have poor disease control, and are unhappy with their care. In addition, chronic medical care accounts for >75% of health care dollars spent in the United States, with approximately 125 million (45%) of the population faced with some type of chronic disease.¹⁷

The principle of the chronic care model involves active patient participation in treatment, patient empowerment and self-efficacy, and a more "hands-off" supportive

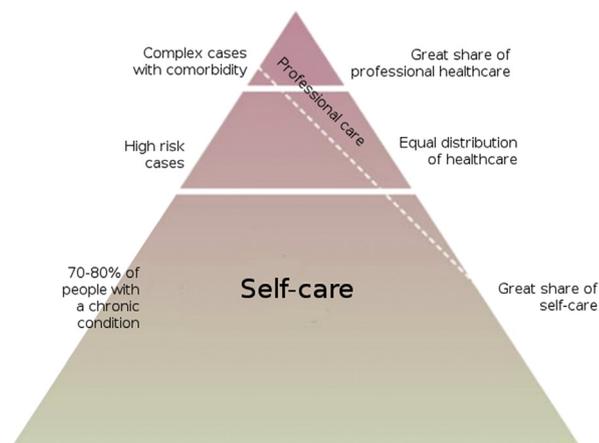


Fig 2. The Kaiser Permanente pyramid as the model for chronic care within a network of care.¹⁸

approach by health professionals working in a network of care to share data and using validated clinimetrics according to guidelines. The patient is the center of the care process. Care is organized according to a pyramid in a network of care: expert-centered and highly complex care is provided in small volume at the top, and when possible, care is provided by a caregiver at a more regional/home base location together with the patient.^{18,19} This approach has been shown to be effective in several other conditions, such as diabetes,²⁰ neurologic conditions,²¹ and atrial fibrillation²² (Fig 2).

DIAGNOSIS

To establish the diagnosis procedure, three reviews²³⁻²⁵ and two guidelines^{26,27} were used. In every patient with swelling of an extremity, the face, or the external genitals, the diagnosis of lymphedema should be considered. History and physical examination are the cornerstones of the diagnosis. In early-stage lymphedema, there is just a reversible, pitting edema. Later, the characteristic features of accumulation of macromolecules, fibrosis, and interstitial inflammation and fat deposition will occur. Preliminary to drafting a treatment plan for lymphedema, the causes of lymphedema need to be clearly stated.

MEDICAL HISTORY

In taking a medical history, the following aspects should be mentioned:

- The manner of initial development
- The type and nature of the condition: symptoms, such as transitional swelling, a propelling sensation, pain, tingling sensations, and a sense of heavy extremities
- Gravitational effects on the edema
- Association with exertion and localization
- Association with pregnancy; problems with sexuality or intimacy
- Family history of the ascertained affliction

- Case history and comorbidity
- Previous (edema) therapy
- Medication
- The patient's knowledge and discernment with regard to lymphedema and coping style
- The patient's disability of activity or participation, distress

PHYSICAL EXAMINATION

In performing a physical examination, the following aspects should be assessed:

- Presence of swelling or edema
- Presence of scar tissue from previous surgery or radiation therapy
- Suspicion of heart disease, kidney or liver problems
- Suspicion of (recurring) cancer
- Signs of venous or arterial insufficiency
- Nature of the swelling: "pitting" vs "nonpitting," unilateral vs bilateral, proximal vs distal
- Accompanying phenomena, such as redness, heat, pain on palpation (generally absent in lymphedema), hyperpigmentation
- Skin thickening and surface elevation by papillomatosis
- Extent of skin fibrosis
- Stemmer sign; when present, it is impossible to pinch and lift the skin fold at the base of the second toe because of an increase in skin thickness
- Nail abnormality, for instance, onychodystrophy, reduced growth rate, "ski-jump nails"
- Palpable resistance in the lymph drainage area
- Indications of recurring tumor growth or skin disorders

SUPPLEMENTAL DIAGNOSTICS

Supplemental diagnostics to the cause of the swelling is indicated in the following instances:

- Inexplicable edema at a young age (<35 years)
- Edema and swelling with an unknown cause
- Edema occurring during observation of a patient belonging to a risk group
- Quick, progressive development of edema with venous or neurologic symptoms after an oncologic treatment of the regional lymph nodes; it is important to discern between malignant and benign lymphedema
- Unilaterally persistent edema after having endured erysipelas, after adequate treatment with therapeutic compression hosiery
- Suspicion of hybrid conditions, for instance, edema resulting from venous insufficiency and lipohypertrophy or lipedema
- Discrepancy between trauma and persistent swelling, for instance, after insect bites, strained ankles, or knee surgery
- Discrepancy between the patient's subjective complaints and the amount of objectively ascertained measurement of swelling in the extremities

CUTOFF VALUES OF SWELLING BEFORE TREATMENT

During the early diagnostic phase, cutoff points for starting treatment (in other words, for making the diagnosis of “clinically relevant” lymphedema) were defined on the basis of the literature.

- When there is a volume difference or change of >5% to 10% of the arm^{28,29} or 10% of the leg³⁰ in comparison with the nonaffected side or the preoperative volumetric values, treatment should commence.
- A suitable, validated means of measurement should be used, one that warrants consistent and reliable results, such as reversed water volumetry, Perometer (Pero-System Messgeraete GmbH, Wuppertal, Germany), or stringent measurement with a measuring tape, converted into volume (see Clinimetrics).
- Volumetric difference of <5% in the arm or 10% in the leg compared with the nonaffected side or preoperative volumetric values does not exclude the presence of lymphedema. Lymphedema can be present on the trunk, breast, or genitals as well.
- When swelling occurs in an area where volumetric measurement is not possible, such as midline lymphedema (genital, head and neck, chest, and thorax lymphedema), alternative instruments for determining lymphedema should be used. Documentation with photography is suggested.

CLINIMETRICS

Given the large role of subjective complaints in lymphedema, attention to the edema volume alone is not enough to ensure early recognition and therapeutic advice or to outline a holistic, patient-centered follow-up program with tailored treatment and support.

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 lymphedema and in patients who are thought to be at risk for development of lymphedema (but who do not yet meet the cutoff value).

A summary of the clinimetric instruments used to evaluate the functional and anatomic problems and the limitations of activity and participation (ICF domains) among patients with a high risk of lymphedema can be found in [Table I](#). All methods and instruments are validated and published.

EDEMA MEASUREMENT

Different measurement instruments can be used to determine volume of swelling and edema: a water tank/“inverse water volumetry apparatus”; an optoelectrical device; and a measuring tape, using a mathematical

formula. There are also methods that can show the presence of edema without taking volume measurement, such as the tissue dielectric constant, three-dimensional scanning, and bioimpedance spectrometry. All these techniques are proven effective and valid but are not interchangeable.³¹

- Measurements at the extremities bilaterally are taken at several set time points: before surgery; before starting and during lymphedema treatment; and after that, periodically at follow-up.
- To get a good impression of changes in volume deviation, it is advised to take measurements every 10 cm from a fixed point (processus styloideus radii/ulnae) on the arm. On the leg, measurements are generally taken from the lower edge of the calcaneus.
- Bioimpedance spectrometry (spectroscopy) and the tissue dielectric constant measurement (MoistureMeter; Delphin Technologies, Stamford, Conn) are inadequate means of determining the general degree of lymphedema and the effects of treatment in everyday practice.
- An isolated volume measurement does not contribute to the diagnosis of lymphedema. Routine taking of measurements of the extremities is recommended, combined with a periodic follow-up program to measure changes in time.

QUESTIONNAIRES

- For measuring the quality of life of patients, both the Lymphedema Functioning, Disability, and Health questionnaire and the Upper Limb Lymphedema 27 can be used for the arms, and the Lymphedema Functioning, Disability, and Health questionnaire for lower limb lymphedema can be used for the legs.³²⁻³⁴
- For measuring psychosocial and physical distress, the Distress Thermometer, which is the English translated version of the Dutch LAST meter, can be used.^{35,36}

SUPPLEMENTARY INVESTIGATIONS

As weight and mobility are the key risk factors for development or deterioration of lymphedema, routine measurement of weight, body mass index (BMI), level of activity, and screening mobility is essential.^{29,37-40}

PREVENTION AND TREATMENT

The treatment of lymphedema comes in two phases, an initial treatment phase and a maintenance phase. There is also a transitional phase between these two phases, in which the initial phase transitions to the maintenance phase during the course of several weeks. Nonoperative treatment is often called decongestive lymphatic therapy. Each phase involves more specific treatment options ([Table II](#)).

Table I. Clinimetric data set

ICF domain and test	Monitoring patient's risk (assessment/diagnostics of secondary prevention)	Initial phase (intensive treatment: mainly therapy evaluation)	Maintenance phase (after treatment and at follow-up assessments: tertiary prevention)	Frequency of measurements
Clinimetric instruments concerning the ICF domains of functions and anatomic characteristics				
Volume measurement (water tank, Perometer, centimeter)	X (during oncologic examination)	X	X	At each examination
Pitting test	X	X	X	At each examination
Body weight, BMI	X	X	X	At each examination
Stemmer sign (for legs)	X	X	X	At each examination
Pain, VAS ^a		X	X	Whenever relevant
Range of motion, goniometer		X	X	At each examination
Fatigue, VAS ^b (also, tiredness of limbs)		On indication	On indication	On indication
Power grip strength, Medical Research Council Scale ^b Hand-held dynamometer		X	On indication	On indication
Clinimetric instruments concerning the ICF domains of activity and participation				
Disabilities of the Arm, Shoulder, and Hand questionnaire ^c		On indication	On indication	On indication
Gait analysis		On indication	On indication	On indication
Dutch Standard for Healthy Physical Activity ^a		X	X	Every examination
Clinimetric instruments concerning the ICF domain of general human functioning: general questionnaire				
Distress Thermometer ^d	X (as a part of oncologic examination)	X (especially after oncologic treatment; in other instances, consider freely)		At least once, during intake
Arm: ULL27 ³⁴ or Lymph-ICF arm ³³ Leg: Lymph-ICF leg ³²		X	X	Several times; a minimum of two
<i>BMI</i> , Body mass index; <i>ICF</i> , International Classification of Functioning, Disability, and Health; <i>Lymph-ICF</i> , Lymphedema Functioning, Disability, and Health questionnaire; <i>ULL27</i> , Upper Limb Lymphedema 27; <i>VAS</i> , visual analog scale. ^a Access and explanation at: http://www.oncoline.nl/cancer-rehabilitation . ^b Access and explanation at: http://www.ucsdglab.org/tools/MuscleScales.htm . ^c Access and explanation at: http://www.fysiovrageenlijst.nl/docs/pdf/DASH%20-%20Disabilities%20of%20the%20arm,%20shoulder%20and%20hand.pdf . ^d Access and explanation at: http://www.oncoline.nl/uploaded/docs/Detecteren%20behoefte%20Psychosociale%20zorg/Lastmeter%20engels.pdf .				

In the initial treatment phase, the main focus is on edema reduction (ie, compression therapy, remedial therapy, exercise, lymphatic drainage), improvement of functioning and well-being, and provision of self-management skills. In the maintenance phase, compression therapy with garments is provided, and patient self-management is further encouraged (ie, skin care, more remedial therapy and exercise, weight management). A professional's involvement should be minimized in this phase, and there is no indication for manual lymph drainage from an efficacy and cost-effectiveness point of view.^{41,42}

The therapeutic interventions used in the different phases of disease and during follow-up are summarized in [Table II](#).

Education and self-management support

Education on lymphedema and self-management occupies an important position in the care process.⁴³ Information, instruction, psychosocial support,⁴⁴ and stimulation of self-management are in fact key elements. Timing of education is crucial.⁴⁵

Self-management domains include (1) activities focused on health improvement and buildup of physical resistance; (2) coping with health care providers and adherence to treatment; (3) ability to self-monitor health, to draw conclusions from signs and symptoms, and to translate this to decisions (eg, going to see a physician or starting self-bandaging); self-efficacy is part of self-monitoring; and (4) coping with the consequences of a chronic disease and trying to get control by

Table II. Summary recommendation of conservative therapy

Therapeutic intervention	Secondary prevention	Initial treatment phase	Maintenance phase
Manual lymphatic drainage	—	X (only obstructive)	Self-massage ^a
Bandaging	—	X	Self-bandaging
Compression hosiery	—	Volumetric difference 5%-10%	X
Pressotherapy, intermittent pneumatic compression	—	In combination with other modalities	Absence of mobility only
Strength training and exercise	X	X	Self-exercise
Mobilizing tissue, fascia release technique	—	X	Self-massage
Weight management	X	X	X
Skin care	X	X (instruction)	Self-application skin care
Awareness	X (course)	X (instruction)	Self-application knowledge
Reduction surgery		X (in clinic)	

^aVoluntary.

self-regulation of personal, behavioral, and environmental factors. Examination of weight and BMI should occur systematically among both patients at risk and patients with lymphedema. Informing them of the healthy norms and standards (such as the Dutch Standard of Healthy Physical Activity in The Netherlands) is important. Instructions on skin care are also an important part of all phases of treatment. Patients need to be able to notice skin changes and to know when to consult a physician. In the maintenance phase, self-application of skin care is a significant factor.

Weight management

Active awareness of obesity or gain of weight (BMI >25-29) and interventions aimed at the reduction thereof are important. This can be achieved by the intervention of a dietitian to set up a calorie balance for the patient to monitor calorie intake and expenditure and to provide dietary advice, weight loss programs, and exercise programs⁴⁶ in cooperation with a physiotherapist.⁴⁰

Compression technology

During both the initial treatment and maintenance phases, effective selection of compression technology is essential in edema treatment. Compression technology consists of the use of several types of bandages, adjustable compression devices (Velcro wraps), pressure point devices, and intermittent pneumatic compression.

In case no further edema reduction can be achieved in the initial phase, compression hosiery should be used to continue compression therapy.

Qualified experts must prescribe compression hosiery. Flat-knitted compression stockings with a high stiffness level (elasticity coefficient) are preferable. Compression class III or IV garments are preferable for lymphedema of the legs; compression class II or III garments are preferable when the arms are affected. The effectiveness of the garments should be checked at least two or three times a year, and they should be replaced if necessary.

Skin care

As lymphatic impairment leads to reduced immunologic response, wounds or discontinuity of the skin should be prevented. In case of a skin defect, antiseptic treatment is advised. When a patient has a risk of skin damage or infection, precautions should be taken by protecting the skin with gloves or protective creams.

When a medical intervention, such as an injection or blood pressure measurement, is performed, the guidelines advise using the healthy side if possible. Otherwise, the ipsilateral side can be used when strict antiseptic precautions are taken in case of an injection or blood sample collection.

Surgery for lymphedema

Surgery for lymphedema is divided into reconstructive and reduction procedures. In general, surgery cannot be a stand-alone treatment modality. It needs to be fully integrated into a more holistic program using the same clinimetrics, ICF method, and chronic care program as for patients treated conservatively for primary and secondary lymphedema.

Reconstructive surgery. Microsurgical procedures have been proposed as a means of improving or restoring lymphatic flow in regions in which lymphatic pathways have been damaged. Many procedures, such as various types of lymphovenous reconstruction, lymph node transfers, and other microsurgical techniques, are researched. Most indications are for early-stage lymphedema without irreversible changes (stage 1-2). Although these reconstructions have been performed for decades with various procedures, there is no robust evidence of efficacy.⁴⁷⁻⁴⁹ In the Dutch guideline, reconstructive surgery is still not considered common care. More comparative efficacy studies are needed. In early stages, nonoperative treatment for lymphedema is effective.

Reductive surgery. In end-stage, irreversible lymphedema (stage 3), the tissue is changed into adipose tissue formation because of long-lasting lymphatic

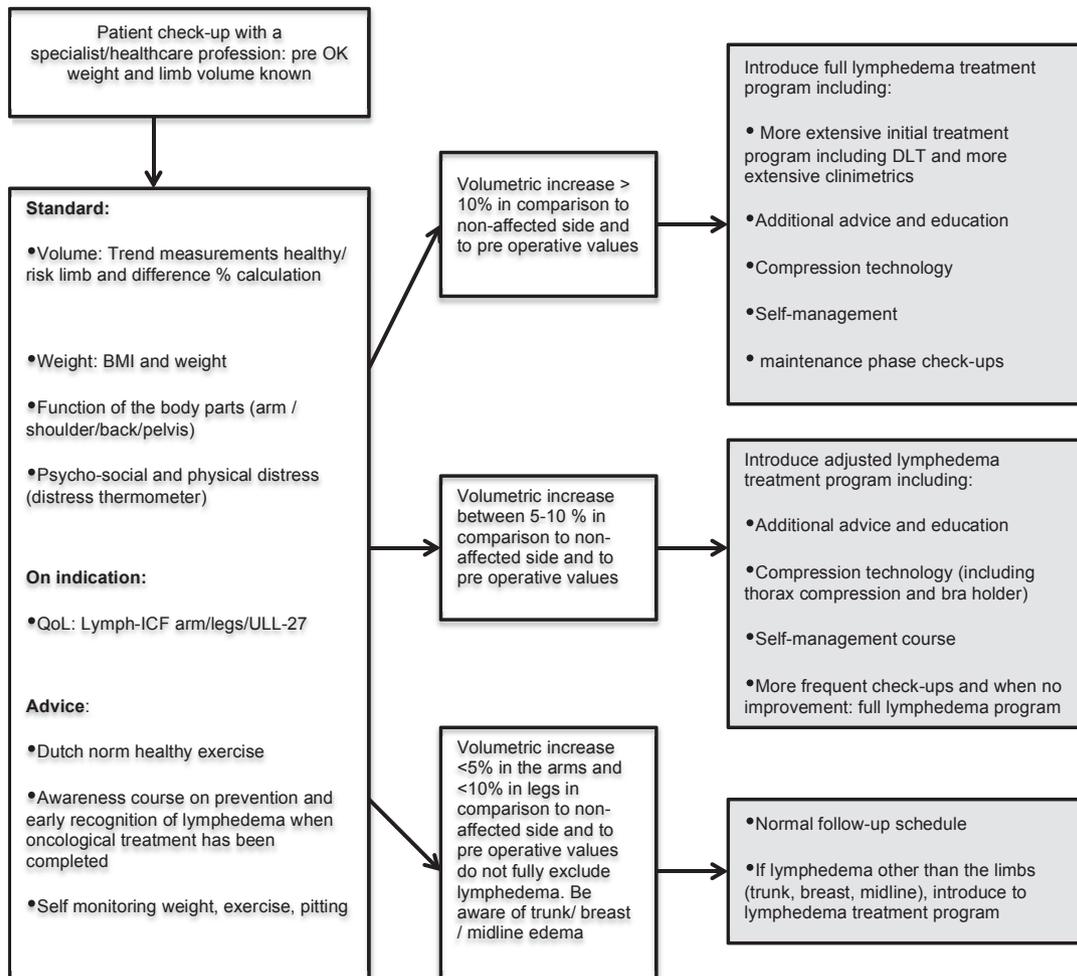


Fig 3. Recommended lymphedema screening of patients who have been treated for cancer with a risk of lymphedema (supplementing standard aftercare and revalidation screening). The recommendations are based on the conclusions from literature and the consensus of the working group. *BMI*, Body mass index; *DLT*, decongestive lymphatic therapy; *Lymph-ICF*, Lymphedema Functioning, Disability, and Health questionnaire; *QoL*, quality of life; *ULL-27*, Upper Limb Lymphedema 27.

impairment. Circumferential suction-assisted lipectomy, initially promoted by Brorson,⁵⁰ is effective in obtaining a long-lasting 100% volume reduction in arms and legs. The use of garments is mandatory. Many studies confirm these results,^{51,52} and the technique has recently proved to be effective for primary lymphedema as well.⁵³

In the Dutch guideline, circumferential suction-assisted lipectomy is considered common care when it is performed in a dedicated expert center and integrated in an ICF-based protocol according to chronic care principles.

FOLLOW-UP

Any type of lymphedema therapy (initial and maintenance phase as well) should be accompanied and followed by objective effect measurement in the several ICF domains. Systematic trend measurement is desirable

for each individual patient at high risk for lymphedema, especially after oncologic procedures (Figs 3 and 4).

CENTER OF EXPERIENCE/EXPERT CENTER

A center of experience/expert center is recognized by specialized health care providers working on an interdisciplinary team. Such a center is accepted as such by the colleagues and authorities in a country. These centers work in a network of care with other professionals in the field of lymphedema (Fig 2). A center provides a concentration of knowledge and more highly specialized health care in the field of diagnosis and nonoperative treatment of lymphedema. Furthermore, there is a concentration of resources for education and research.

Since 2016 in the European community, expert centers have joined forces in the field of rare diseases in the European Reference Network on Rare Multisystemic

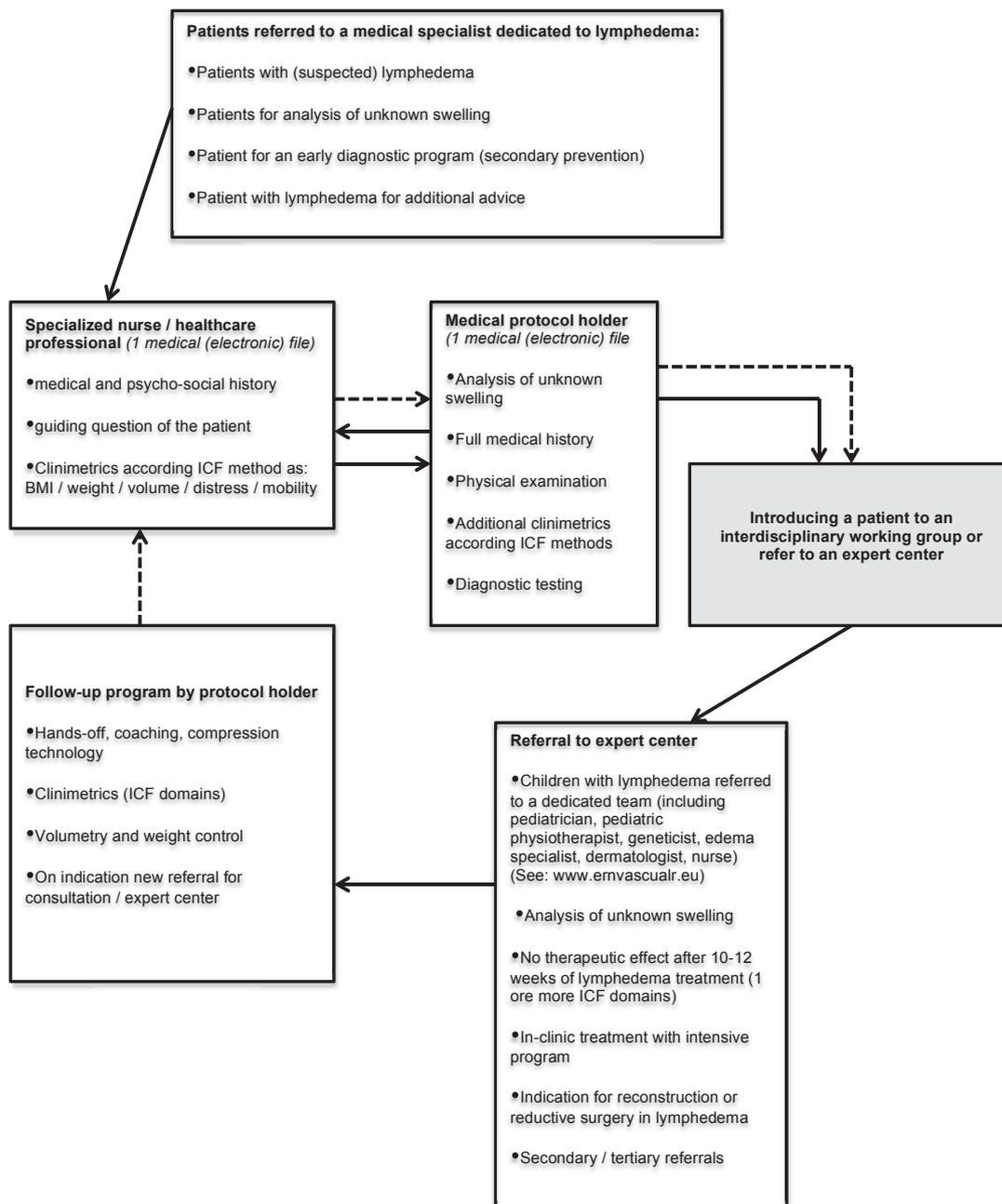


Fig 4. Recommended routine screening for patients with presumed lymphedema. The recommendations are based on the conclusions from the literature and the consensus of the working group. *BMI*, Body mass index; *ICF*, International Classification of Functioning, Disability, and Health.

Vascular Diseases (VASCERN; www.ernvascular.eu). There is a dedicated working group on pediatric and primary lymphedema.

RECOMMENDATIONS

Recommendations for screening for lymphedema based on conclusions from the literature and the consensus of the working group are presented in [Figs 3 and 4](#).

CONCLUSIONS

The new guidelines for lymphedema are based on new concepts of chronic care according to the chronic care management methodology and offer an integrated, more holistic functional approach by using the ICF model. This approach focuses on awareness, early diagnostics, and risk stratification. Early recognition and treatment ensure a reduced morbidity for the patient and decreased risk of complications and deterioration of the lymphedema.

In addition, a more systemic approach is proposed for treatment: choosing dedicated treatment options for the initial and maintenance phases, including focusing on an active participation of the patients in their treatment by learning more self-management. These guidelines provide transparency for health care professionals and insurance organizations for an effective and efficient approach for diagnosis, screening for risk of lymphedema, and nonoperative treatment.

AUTHOR CONTRIBUTIONS

Conception and design: RD, AH

Analysis and interpretation: Not applicable

Data collection: RD, AH

Writing the article: RD, AH

Critical revision of the article: RD, AH

Final approval of the article: RD, AH

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RD and AH contributed equally to this article and share co-first authorship.

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