LIPEDEMA GUIDELINES IN THE NETHERLANDS 2014

INITIATIVE

Dutch Society for Dermatology and Venereology and the Dutch Academy of medical specialists (ORDE)

PARTICIPATING SOCIETIES/ORGANIZATIONS

- Dutch Society for Dermatology and Venereology (NVDV)
- Dutch society for Pycho-dermatology (NVPD)
- Dutch Society for Surgery (NVH)
- Dutch Society for Nuclear medicine (NVNG)
- Dutch Society for Physiotherapy within the lymphology (NVFL)
- Dutch society of skin therapists (NVH)
- Royal Dutch Association for Physiotherapy (KNGF)
- Lipedema patients circle (LIPV)
- Dutch Network for lymphedema and lipedema (NLNet)

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- Office of the Dutch Society for Dermatology and Venereology

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THIS DIRECTIVE IS AUTHORISED BY THE PARTICIPATING ORGANISATIONS

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INTRODUCTION

MOTIVATION

Lipedema is a chronic, incurable, often progressive affliction that occasionally causes significant morbidity. Initially, patients develop a disproportionate increase of body fat in the legs, buttocks and/or arms. Dieting and physical exercise have only limited effect on this disproportionate body fat distribution. The legs may be sensitive and are prone to bruising after only mild trauma. This can deteriorate into severe pain and reduced mobility, ultimately leading to a limitation of activity and social participation. As a result, lipedema patients may often be diagnosed with obesity. Dietary measures generally affect the obesity component but have little effect on the disproportionate body fat distribution. Because lipedema contributes to an increased BMI, even in non-obese patients, a connection between lipedema and excessive calorie-intake or obesity is often incorrectly assumed. In addition to physical problems, lipedema can also lead to psychosocial problems. These are often caused by the failure of consulting professionals to recognize or acknowledge the condition, or because (incorrect) recommendations for weight loss and physical exercise do not contribute to improvements in the complaints.

Therefore, it is important to recognize lipedema early so that its accompanying symptoms can be acknowledged at an early stage and be incorporated into an integrated treatment.

Little consistent information regarding the diagnosis or treatment of lipedema is found in the literature. Therefore, the goal of this directive is to attempt to establish a consensus for the diagnosis of lipedema and to discuss its symptoms and influencing factors, as well as its effects on activity and social limitations, as these negatively influence the life of the patient.

GOALS OF THIS DIRECTIVE

This directive will serve as an everyday practice guideline to improve the quality of care for lipedema patients. As lipedema is a chronic condition, a proposition will be made to rate the *chronic care model* [Nivel, 2012] – which is the current approach to chronic conditions – in terms of its applicability in patients with lipedema. In accordance with government policies, this program-based approach aims to incorporate care in all the phases of a chronic illness: early identification, prevention, self-management and good health care. Furthermore, it is beneficial to government and society to ensure that people lead longer, healthier lives and stimulate lasting social participation.

This approach has four defining points of attention:

- Better-attuned, multidisciplinary health care
- Interconnected organizations and health care
- Increased focus on patient autonomy and responsibility
- Better integration between preventive and curative treatment

The work group collectively expresses its hope that, based on this directive, all healthcare providers will be able to produce protocols or training material for their respective hospitals or clinics, in which steps for (physical) research, diagnostics, treatment and (after-)care are described.

PROBLEM DEFINITION AND TERMINOLOGY

This program-based vision of chronic care encompasses both the concepts of the *chronic care model* (CCM), as included by the American Edward Wagner (Wagner 1998) in the *International Classification of Functioning, Disability and Health* (ICF) model that is used by the WHO (World Health Organization (2001).

The CCM is a model for developing and improving health care for the chronically ill. The general principle is that people with chronic conditions will only change their behavior when they are granted a leading role in this process. Therefore, self-management is central to this model. Self-management means that people will be able to cope well with a chronic condition and its consequences. There are many initiatives to improve self-management, but not all of these are widespread. Supporting self-management requires caregivers to fulfill a role that is similar to that of a coach, which may differ from prior approaches. However, patients generally require support in assuming control themselves (NIVEL 2011). (See also figure 1, below.)

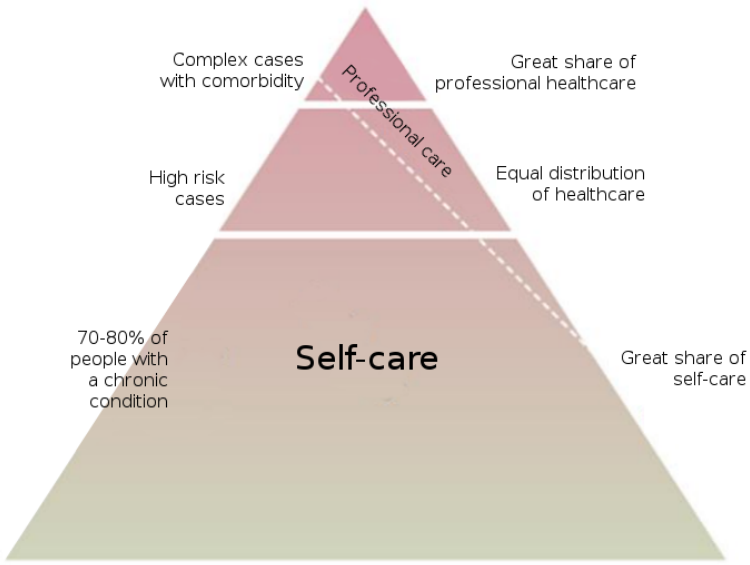

Figure 1: Distribution of self-care and professional care according to the complexity of the health situation among people with chronic illnesses. Source: NIVEL 2011

The International Classification of Functioning, Disability and Health (ICF) has been a globally accepted system of classification since 2001. The ICF is a classification model that distinguishes between different domains and describes the symptoms of an illness as well as the patient's functional capability (WHO 2001). The ICF defines human functioning from three perspectives: 1) the human organism (described under functions and anatomical characteristics); 2) human action (described under activities); and 3) human participation in social life (described under participation). Furthermore, the ICF includes personal and external factors that influence human functioning. Personal factors are characteristics such as age, education, personality and character, experiences and competence. External factors concern both the social and the physical environments in which people live, for instance the attitudes of other people, social standards, legislation, facilities, external resources, and working and living conditions (NIVEL 2011). (See figure 2, below.)

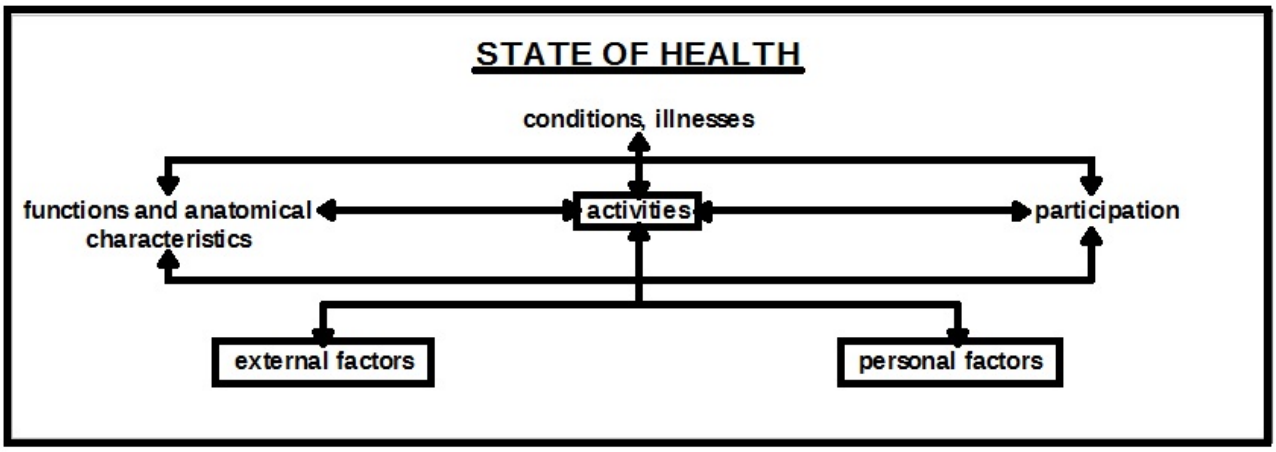

Figure 2: The interaction between the different domains of the state of health (functions and anatomical characteristics / activities / participations) and external and personal factors in the biomedical – psycho – social model of ICF.

Clinimetrics (defined as the measuring of clinical phenomena) are used to map the health care needs of the patient from all perspectives of human functioning. Clinimetrics include diagnosis/quantitation and evaluation and should be used frequently in the initial treatment phase, which is more aimed at addressing functional physical characteristics. The measurement intervals increase during the maintenance phase, when individual monitoring plays a larger role; the focus of care also shifts to the domains of activity and participation.

STUMBLING-BLOCK ANALYSIS

Lipedema is characterized by a broad spectrum of disease burden. Not every patient with lipedema requires treatment. The evidence for both diagnosis and therapy is limited. Lipedema has not been included in the International Classification of Diseases (ICD), although the European Society of Lymphology has recently requested that it be included in the ICD. *(WHO 2000, International Statistical Classification of Diseases and Related Health Problems, 10th Revision).*

A major obstacle is the lack of scientific literature, where there is little consistency concerning the diagnosis of lipedema.

PRINCIPAL QUESTIONS

- How and when is lipedema defined?
- What elements are needed for adequate (early) diagnosis and follow-up of lipedema?
- What should the treatment of lipedema patients consist of?
- What is the role of patient organizations in lipedema aid?

TARGET AUDIENCE / DIRECTIVE USERS

The target audience consists of all healthcare providers who are active in the lipedema field or involved in the diagnosis and/or treatment of patients with this condition. This includes medical, allied health professionals, and nursing professions.

SCIENTIFIC SUBSTANTIATION

This directive is based on a recent, high-quality review (Forner et al. 2012) of the systematic research that has been done over the last sixteen years (1995-2011). R.J. Damstra's book (Diagnostic and therapeutic aspects of lymphoedema 2013, Rabe verlag Bonn, chapter 15) and a systematic review by Langendoen (2009) were used as supplements. Most of the literature cited in these articles overlaps.

PATIENT PARTICIPATION

Patient organizations will attempt to publish this directive in a special “patient edition”. The goal is to put this directive at the disposal of non-professionals in clear language, allowing patients to actively learn about and participate in the diagnosis and treatment of their condition.

IMPLEMENTATION

Much attention has been paid to practical applicability, effectiveness, purpose and necessity when drafting the directive. The project leaders have worked towards this orientation. Some of their recommendations therefore include norm indications such as: minimal norm, standard norm and target norm.

DISTRIBUTION

The directive has been made digitally available to everyone and will specifically be brought to the attention of all hospitals and scientific organizations. A summary of the directive will be offered for publication in the Dutch Magazine of Medicine (*Nederlands Tijdschrift voor Geneeskunde).* We also ultimately intended to produce this English publication.

JUDICIAL SIGNIFICANCE

Directives are not lawful regulations, but scientifically substantiated and profusely supported insights and recommendations that healthcare providers should follow to assure high quality care. Because the directive is based on the average patient, health care providers can deviate from the guidelines in individual cases when deemed necessary. Deviating from the directive can even be imperative when the patient’s situation demands it. However, when deliberate deviation occurs, it has to be justified, well documented and, where possible, done in consultation with the patient.

REVISION

Because this directive is modular in structure, sections can be easily revised.

SUMMARY OF RECOMMENDATION

CLINICAL PRESENTATION / DIAGNOSTIC CRITERIA

Due to a lack of unambiguous criteria when establishing the diagnosis of lipedema, the work group has assembled a list of criteria that is based on clinical experience and supported by the literature; the criteria are displayed below:

Diagnosis is certain when the following criteria are 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 additional criteria F13 or F14 also assures diagnosis.

		Medical history (A) (criteria of Wold et al.)
A	1	Incongruent fat distribution
	2	No / limited influence of weight loss on incongruence
	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	Incongruent 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 the vicinity
	11	Sudden termination at elbow
		Lower arm:
E	12	Thickened subcutaneous fat, accompanied by slender back of hand (cuff sign)
		Extra criteria
F	13	Pain with bimanual palpation
	14	Distal fat tissue tendrils at the knee

Diagnosing lipedema can be challenging. Subjective complaints often contribute to the difficulty in diagnosis. Therefore, it is important to thoroughly map the complaints of patients with (suspected) lipedema, paying special attention to objective, measurable parameters [see chapter 2, (early) diagnostics and effect measurement].

(EARLY) DIAGNOSTICS AND EFFECT MEASUREMENTS

The diagnosis of lipedema can generally be determined based on the medical history and physical examination. When taking the medical history and performing the physical examination, the criteria established by the work group should, at a minimum, be addressed (see recommendations in chapter 1; minimal norm).

Due to the wide variety of involved complaints, it is recommended that a general health profile (using suitable clinimetrics) be created to document overall functioning. This profile should be used unequivocally during diagnosis and when designing a therapeutic and follow-up regimen.

At a minimum, the information to be obtained in the diagnostic phase includes (repetitive) girth measurements of the involved extremities, BMI and abdominal circumference, and establishing the daily activity level. This protocol is also applicable to patients who display (as of yet) too few of the criteria to diagnose early-stage lipedema. Timely recognition and intervention can be particularly important in the context of functionality and cost reduction.

Recognition and potential treatment of psychosocial problems is important in both the diagnostic and treatment phases.

Additional diagnostics can be performed when a second condition is suspected.

For instance, dynamic lymphatic scanning can be considered when lymphatic involvement is suspected. Endocrine evaluations can be considered when a metabolic disorder is suspected. Venous pathology must be excluded.

TREATMENT

CONSERVATIVE TREATMENT

An active lifestyle in conformity with the Dutch Norm for Healthy Exercise *(Nederlandse Norm Gezond Bewegen or* NNGB), as well as maintenance of a healthy body weight, should be encouraged in lipedema patients who do not already meet these standards (Figure 3).

A patient with lipedema who also has an unhealthy lifestyle and/or functional limitations requires multidisciplinary treatment with suitable clinimetrics. Emphasis should be placed on stimulating vascular or lymphatic pumping, regulating caloric balance (food intake versus caloric burn), and a meticulous exercise program aimed at strength training and conditioning (using the graded activity method), if necessary in combination with a weight loss and exercise program.

The treatment and support of lipedema patients requires expertise in the areas of training, graded activity and cognitive behavioral principles, supported by the application of suitable clinimetrics.

Intensive coaching and proficiency in motivational interviewing is recommended to assure patient independence.

Medical providers must recognize psychosocial problems and provide basic psychological aid (and, if necessary, referral to a specialist) in patients with lipedema.

SURGICAL TREATMENT

Liposuction and/or reductive surgeries are a part of lipedema treatment and take place in specialized centers where these procedures are performed according to established protocols, using good clinimetrics and multidisciplinary cooperation.

PATIENT ORGANIZATIONS

The existence of organizations for lipedema patients should be noted, such as NLNet, Stichting Nederlandse Lipoedeemdag and the Lipoedeem Patiënten Vereniging in the Netherlands.

1. DETERMINING DEFINITION

INTRODUCTION

A major obstacle is the limited available scientific literature, along with inconsistency concerning the diagnosis of lipedema. The condition is characterized by symmetrical accumulation of fat tissue with typical clinical characteristics, generally in the extremities (more detail provided later). Lipedema is an unfortunate term, as it often evokes the idea of swelling due to fluid accumulation. However, it refers to swelling – in the sense of an increase in volume – due to increased fat tissue.

PRINCIPAL QUESTION

- How and when can lipedema be confirmed?

The clinical presentation, pathophysiology, stages and diagnosis of lipedema are discussed in this chapter.

SCIENTIFIC SUBSTANTIATION

1.1 PATHOPHYSIOLOGY

The etiology of lipedema is still unknown. The condition almost exclusively occurs in women, which is currently an unexplained phenomenon, although hormonal factors may contribute. Externally, one generally perceives a disproportionate increase of subcutaneous fat tissue in the extremities, buttocks and hips. The arms may also be affected. It is currently unclear if this is due to adipocytic hypertrophy, adipocytic hyperplasia or a combination thereof. The extent to which adipocytes possess other metabolic characteristics is also unknown. However, there are significant differences in the occurrence of (physiological) fat distribution in the world. For example, as all lipedema experts can confirm, the condition is practically absent among women of Asian origin.

Another component of lipedema is the development of edema. The characteristics of classical lymphedema – increased interstitial protein synthesis or classical fibrosis – are absent here. Edema that is similar to other forms of so-called dynamic lymphatic insufficiency may occur in protracted cases with a clinical picture consistent with lymphatic overload.

Some publications presume that there is microangiopathy of the lymphatic capillaries (located in the connective tissue septa, between the fat lobes) [Földi 2005], causing fragile vessel walls and increased permeability. The hypothesis is that this increased capillary permeability causes excessive lymphatic filtration in the interstitial cavity. This increased interstitial volume exceeds the natural lymphatic clearance capacity

and can eventually result in dilatation of the pre-lymphatic structures. Furthermore, increased fat tissue potentially leads to compression of the lymphatic system, resulting in a vicious cycle in which limited lymphatic drainage and increased fat tissue sustain each other, which in turn could enable the development of secondary lymphedema (lipo-lymphedema) [Damstra 2013; Langendoen 2009].

Another characteristic includes capillary fragility, which may explain the ease of bruising and be responsible for the increased tendency to the development of edema.

The sensitivity and pain – sometimes to light touch – could perhaps best be thought of as the metaphorical straw that breaks the camel's back, resulting in an excessive increase in pain. This explanation is supported by the fact that even a relatively small reduction of fat tissue using liposuction results in reduced pain.

Finally, an adipose component often is present, which involves a general increase in body fat and a generalized, mostly centripetal distribution of fat (in the abdominal area), along with increased fat in typical locations. Moreover, it is well known from the lymphological literature that obesity and inactivity negatively influence lymphoedema and lymphatic drainage; these factors may also play a role in lipedema with edema.

CONCLUSION

No level	The pathophysiology of lipedema is unknown. Hypertrophy and / or hyperplasia of adipocytes, excessive filtration of fluid and capillary fragility may be causal mechanisms. Additionally, obesity and physical inactivity often exacerbates and complicates lipedema (especially in later phases).

1.2 CLASSIFICATION AND STAGES

The classification of lipedema uses a clinical description without objective quantification. Strößenreuther, like Meier-Vollrath and Schmeller, established a classification based on skin changes [Fife 2010; Strößenreuther 2001; Meier-Vollrath 2004]. This classification includes three stages: in the first stage, the skin appears smooth and even, yet because of thickening of the subcutaneous fat tissue the skin will feel like 'Styrofoam balls in a plastic bag' on palpation; in the second stage, 'walnut to apple sized' subcutaneous indurations develop, along with an irregular skin exterior resembling a "mattress"; and in the third stage, the indurations will increase in size and prominence, and deformed fat deposits will become visible.

A different classification was established by Schrader, based on the anatomic localization of fat deposits [Schrader]. Type one describes fat deposits on both buttocks without spread to the rest of the legs; type two describes fat deposits located between the buttocks and knees; type three describes the fat deposits

located between the buttocks and malleoli; type four describes fat that is predominantly deposited in the arms, without affecting the lower extremities; and type five describes symmetrical deposits between the knees and malleoli, without affecting the feet.

Both classifications are insufficient in practice, as many patients cannot be grouped accurately based on these classifications.

Because lipedema is a chronic condition that significantly affects everyday functioning, decreases social participation, and has considerable effects on the quality of life, more information is needed than just a description of location and stages. Assessment of patients using the ICF method (see further), which is used for many other chronic conditions and is being researched for lymphedema, has not been attempted for lipedema yet may be a suitable method to describe relevant aspects of the condition, including its quantitative aspects by the use of validated clinimetrics.

CONCLUSION

No level	There is no adequate classification or staging system for lipedema. The existing classification is insufficient. Assessment based on the ICF method may contribute to more accurate classification and dedicated care

1.3 CLINICAL PRESENTATION / DIAGNOSTIC CRITERIA

Lipedema almost exclusively occurs in women. In the literature, there are only two described cases of lipedema in men [Wold 1951; Chen 2004]. Lipedema generally develops during or after puberty, but can also develop during pregnancy or even during menopause [Fife 2010]. Lipedema is not included in the International Classification of Diseases (ICD); however, the European Society of Lymphology has recently requested that lipedema be included in the ICD (WHO 2010, International Statistical Classification of Diseases and Related Health Problems, 10th Revision.). The precise incidence is therefore unknown.

The first signs are often cosmetic in nature and relate to disproportionately heavy legs. Attempted weight loss is ineffective and in fact leads to disappearance of fat tissue in unaffected areas, resulting in an "unhealthy", disproportionate figure (for instance, concave cheeks or smaller breasts). Even with healthy nourishment and exercise patterns, the body parts affected by lipedema tend to increase in volume, worsening the disproportion. The increased fat tissues can cause mechanical difficulties. The significantly increased volume of fat on the insides of the knees and upper legs can force a straddling position (resulting in *genu valgum* or "knock-knees") and damage the skin due to friction.

When lipedema is not recognized, the patient's increased but failed exertions often lead to frustration, social and societal isolation and abandonment of dietary regimens due to hopelessness. An (accelerated) obesity component can develop at this point. Mechanical difficulties also result in reduced mobility, which in turn contributes to obesity.

The great variety of pain complaints is noteworthy. Pain can vary from mild to extreme, and even light touch can cause distress; for example, compression can become intolerable (when not well-placed). Pain is an important consideration in the diagnosis of lipedema. Fatigue in the extremities is often present as well, which reduces mobility. Furthermore, the majority of patients will develop distinctive 'cellulite' (a fundamentally physiological phenomenon in women due to the anatomy of the subcutaneous tissue). Many patients report easy bruising after slight trauma.

Lipedema can easily be recognized during physical examination although scientific consensus on the terminology and diagnosis is lacking. Typical symptoms are the sudden termination of the increased fat tissues: this can occur at different levels: at knee height on the medial side, right below the knee, or right above the ankles, leading to a 'cuff sign' at the elbows or wrists. The lipedematous tissue is more sensitive to compression than unaffected and / or obese tissue. This phenomenon can also be used to determine the upper limit of involvement for patients in whom inspection is difficult (e.g., when obesity is present in the torso). The subcutaneous fat layer of the upper leg is affected circumferentially along its entire length, giving rise to 'wobbly legs'. This is unlike lipohypertrophy, where the fat accumulations are more localized. The upper and, to a lesser degree, lower arms ultimately also are involved in this process in some patients.

Special attention is required based on the degree of lipedema. This is unpredictable per individual. On one hand, lipedema exists in a very mild form where there are no subjective complaints and a minor increase of subcutaneous fat is the only symptom. When the lipedema is non-progressive it can remain life-lastingly mild and need not result in distress. On the other hand, lipedema can rapidly develop progressively. To date, it has not been possible to predict the future developments of lipedema from the initial stage. Distinguishing between mild forms of lipedema, disproportionate fat distribution, racial- and postmenopausal variations of fat distribution in women and lipohypertrophy prove difficult (also due to lack of adequate differentiating diagnostics).

An obesity component often plays a role in more extensive forms of lipedema and there may also be mixed image of adiposity and lipedema. It is estimated that of all women with an increase of fat deposits in the lower extremities 60% of these are caused by obesity, 20% by lipedema and the remaining 20% by a combination of the two [Herpetz 1995].

CONCLUSIONS

Level 2	Lipedema occurs almost exclusively in women and generally develops during or short after puberty. There is a great range of both mechanical and functional distress as well as pain and fatigue complaints in lipedema. *Fife 2010.*

Level 4	Lipedema has not (yet) been included in the International Classification of Diseases (ICD). Distinguishing between mild forms of lipedema, disproportionate fat distribution and lipohypertrophy can prove difficult because of this (as well as due to lack of adequate differentiating diagnostics). *Work group's point of view*

ADDITIONAL CONSIDERATIONS

considering the frequency of disproportional fat distribution or mild forms of lipedema, it should be prevented that diagnosing patients with lipedema leads to excessive perception of illness and possibly unnecessary medicalization. On the other hand, recognition and diagnosis at an early stage is important because the course is difficult to predict and could have significant negative consequences for patient functioning.

RECOMMENDATION

Due to a lack of unambiguous criteria for establishing the diagnosis of lipedema, the working group has assembled a list of criteria that are based on clinical experience and supported by the literature. These criteria are listed in the table below:

Diagnosis is certain when the following criteria are 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 confirms the diagnosis.		
		Medical history (A) (criteria of Wold et al.)
A	1	Incongruent / disproportinal fat distribution
	2	No / limited influence of weight loss on incongruence
	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	Incongruent / disproportional fat distribution
	7	Circumferentially 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 the surrounding area
	11	Sudden termination at elbow
		Lower arm:
E	12	Thickened subcutaneous fat, accompanied by slender back of hand (cuff sign)
		Extra criteria
F	13	Pain with bimanual palpation
	14	Distal fat tissue tendrils at the knee

Diagnosing lipedema can be challenging. Subjective complaints often contribute to the difficulty in diagnosis. Therefore, it is important to thoroughly evaluate the complaints of patients with (suspected) lipedema, paying special attention to objective, measurable parameters [see chapter 2, which discusses (early) diagnostics and effect measurement].

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2. (EARLY) DIAGNOSTICS AND EFFECT MEASUREMENT

INTRODUCTION

It is important to recognize and monitor lipedema in the early stages of disease because of its frequent occurrence, the diversity of symptoms involved and the condition's chronic, progressive nature. Because lipedema requires specific treatment, it is important to avoid confusing it with other conditions.

PRINCIPAL QUESTION

- Which aspects need to be determined for adequate (early) diagnosis and follow-up of lipedema?

SCIENTIFIC SUBSTANTIATION

2.1 ANAMNESIS AND PHYSICAL EXAMINATION

It is generally assumed that anamnesis and physical examination suffice for diagnosis [Forner 2012]. Patients with lipedema may complain of discomfort, heavy legs and mild to severe sensitivity to touch with very easy bruising. Exercise and heat cause increased pain and swelling of the lipedematous areas, which do not improve when the legs are elevated.

Lipedema patients often have a history of various diets and exercise regimens that do not lead to reduction in the volume of the extremities or improvement in the accompanying complaints.

Complaints concerning posture and movement are common. Many patients describe fatigue – both in the legs and generally – and reduced conditioning, weakness in the leg muscles (manifested in difficulty climbing stairs, getting up from a deep chair, etc.). Orthopedic problems are often found in lipedema patients, mostly occurring in the knee and less frequently in the ankles. Many patients have stance deformities, for instance increased valgus stance of the knee, and flat and / or spread feet. It is importance to distinguish between pain caused by these stance deformities and pain caused by the sensitive lipedematous tissue surrounding the knees. The latter complaints are often wrongly attributed to problems in the knee joint. Gait deviation caused by pain and mechanical restrictions due to increased fat deposits are likewise often observed [Langendoen 2009].

The entire spectrum of complications can lead to a reduction of everyday functioning and societal participation (in work, hobbies and sports), causing the patient to end up in a downward spiral. Generally, a reduced physical activity level is observable in a large part of the patient population.

The symmetric, bilateral fat deposits on the extremities develop slowly and gradually but are disproportionate in comparison to the torso, face and neck. Lipedema can be hard to recognize in its early stages. The only indicator is often the disappearance of the concave sulcus on both sides of the Achilles tendon [Fife 2010]. It becomes easier to make the diagnosis when lipedema increases. A characteristic feature of the condition is that, when the lower legs are affected, the fat deposits end abruptly above the malleoli, leaving the feet and ankles unaffected and resulting in a contrasting edge (the so-called 'cuff sign'). A similar pattern can be seen when the upper extremities are affected; the fat deposits continue to the wrists, below which slender hands will be visible [Forner 2012].

The upper leg is affected circumferentially along its entire length, unlike other types of increased fat deposits in which the fat deposits are more defined. The thickened fat layer runs distally to the medial side of the knee or may continue as a defined tendril distal to the knee. Proximally, it stops either at the level of the trochanter or continues to the hip area. The buttocks can be involved in the process. An irregular surface ('cellulite') can be present in the initial stages, but generally develops during the course of the process.

A significant sign of lipedema is that mild compression of the subcutaneous tissue results in pain, which can vary per region.

2.2 CLINIMETRICS

Lipedema is a chronic, non-curable condition that can coincide with pain, loss of strength and conditioning. In more serious cases, lipedema can decrease the daily activity level, negatively influencing quality of life.

Along with making a medical diagnosis, one should compose a health profile with suitable clinimetrics.

Human functioning should be at the center of a health profile composed using the ICF method; functioning should be considered in reference to the medical condition, as well as personal and external factors (see figure 2).

Anamnesis should be directed at all facets of the ICF (see figure 2), and suitable clinimetrics should be chosen. In this way, the patient's ' subjective' story can be expressed quantitatively and will form the basis for the initiation, titration and evaluation of interventions.

In the diagnostic phase, the minimum dataset should include circumferential measurements of the affected extremities, BMI and abdominal circumference, as well as establishing the daily activity level. A first impression of the daily activity level can be obtained by following the Dutch Norm for Healthy Exercise (Nederlandse Norm Gezond Bewegen or NNGB) [Annex 3]. These same tools can be used during follow-up. Choices regarding therapy can be based on the following summary (see table 1). In the treatment phase, the array of measurement tools is more extensive.

Both in the diagnostic and the treatment phases, recognition and possibly treatment of physical and psychosocial issues are important. The use of measurement tools is also recommended here, but a description of these tools is beyond the scope of this directive.

The choice of clinimetric instruments is not based on the literature, but rather on the available knowledge and perceptions of the working group. Validated measuring tools are not available for patients with lipedema. For a summary of lipedema diagnostics, see [Annex 1: flowchart diagnostics]. Depending on the anamnesis (medical history of patient complaints), the clinimetrics will be more or less extensive.

TABLE 1. SUMMARY OF CLINIMETRIC INSTRUMENTS SUGGESTED BY THE WORKING GROUP FOR USE WHEN COMPOSING A HEALTH PROFILE OF PATIENTS WITH (SUSPECTED) LIPEDEMA.

Disorders of functioning and anatomical characteristics / limitations in activity and participation	Measurement tool
Circumference	Measuring tape (+ mathematical formula) Perometer
Mobility	Goniometer
Strength	Medical Research Scale (MRC) Hand-held dynamometer
Pain	Visual Analogue Scale (VAS)
Fatigue	Multi-Dimensional Fatigue 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 assess 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

**The commonly used term BMI often causes confusion in lipedema. Lipedema contributes to body weight and therefore to increased BMI, which is used as a measurement for adiposity and obesity. Lipedema patients will therefore often have an increased BMI, without this being the cause of excess weight in the traditional sense. A marked discrepancy in clothing size between the upper and lower body is characteristic of lipedema patients.*

2.3 ADDITIONAL DIAGNOSTIC TESTS

Additional diagnostic tests are not advised routinely and should be performed only in the case of specific indications. Additional tests can be valuable to evaluate other conditions that are within the differential diagnosis and to determine the severity of the lipedema.

LABORATORY TESTS

When there are doubts about the diagnosis, additional laboratory tests can be considered, a number of which are mentioned below.

BLOOD TESTING

There is no difference in blood test results in patients with lipedema. Some general blood tests (i.e., routine chemistry, hematology, thyroid function, blood glucose can be useful because some patients do have underlying medical conditions that are responsible for the disproportionate fat distribution [Forner 2012]. Although hormonal factors may play a role, there are no indications for performing endocrinologic evaluations.

STREETEN TEST

The diagnosis of lipedema can be suspected based on results of the Streeten test along with clinical characteristics [Forner 2012]. Cardiac, renal and venous insufficiency must be excluded before the Streeten test can commence. The patient drinks 20 ml of water per kg of body weight and stays in an upright position for four hours. During this observational period, urine samples are collected. The volume of the leg is measured before and after the test. Healthy individuals will secrete more than 60% of the ingested water, and the volume of the leg should not increase more than 350 ml per leg. Pathological results suggest the existence of increased capillary permeability.

Due to the logistic complexity of this test, its use is limited and reserved for situations in which there are no other means available to confirm the diagnosis.

(DUPLEX) ECHOGRAPHY

Duplex examination (Doppler color ultrasound) of lipedematous areas reveals thickened subcutaneous tissue with high echogenicity (displayed as white areas), whereas lymphedema typically reveals echodense (displayed as black, fluid), non-compressible areas. Cutaneous echography reveals that in lipedema patients, the dermal thickness and echogenicity are normal, whereas an increase in dermal thickness and lower echogenicity were characteristic of lymphedema patients [Forner 2012].

CT

A CT scan has a sensitivity of 95% and a specificity of 100% for diagnosing lipedema, according to Monnin-Delhom et al. Thickening of the skin can occur in lipedema, but subcutaneous fluid accumulation, the honeycomb pattern and muscle thickening are absent in lipedema, though these features are characteristic of lymphedema [Forner 2012].

MRI

MRI and its modified form, MR lymphangiography, can be helpful when evaluating lymphatic circulation, should there be a need to exclude lymphatic involvement [Forner 2012].

LYMPHOSCINTIGRAPHY

Lymphoscintigraphy can be useful when establishing a differential diagnosis of edema, as it can rule out lymphatic dysfunction. There are no initial lymphatic drainage disorders in lipedema. Lymphatic dysfunction can be observed in patients with protracted lipedema without the presence of morphological abnormalities that are characteristic of lymphedema [Forner 2012].

MEASURING DIELECTRIC CONSTANT

A hand-held unit emitting 300-MHz electromagnetic waves that reflect against tissue (to a maximum depth of 5 mm) can be used to evaluate local fluid levels. This allows untreated lymphedema to be distinguished from lipedema (or healthy tissue), as well as from lymphedema that has been treated with compression and lacks residual edema. This is a simple and non-invasive diagnostic method. Birkballe et al. have demonstrated that normal tissue fluid levels differ between patients with lymphedema on the one hand, and healthy individuals and lipedema patients on the other hand. Confirmatory research is still needed [Birkballe 2014].

2.4 DIFFERENTIAL DIAGNOSES

Lipedema has a typical clinical presentation, but is sometimes difficult to differentiate from obesity, lipohypertrophy and (lymph)edema. Although a hormonal component is assumed because lipedema most commonly develops or progresses during puberty, pregnancy or menopause, there are no known hormonal abnormalities in lipedema, and there is no related research.

In obesity, the increased subcutaneous fat tissue is distributed in a more generalized fashion rather than disproportionately, as is observed in lipedema. The feet are generally also affected in cases of obesity, so the 'cuff sign' is absent. Furthermore, the subcutaneous tissue is not sensitive. Some consider the fat distribution that is typical of lipedema to be a part of gynecoid fat distribution.

The cuff sign is present in lymphedema. Additionally, Stemmer's sign will be negative in lipedema, unlike in lymphedema (Stemmer's sign is positive when it is impossible to pinch and pull the skin on the back of the foot at the phalanx, between the second and third toe). [Wold 1951; Fife 2010; Damstra 2013; Langendoen 2009; Child 2010]. When there is a combination of lipedema and lymphedema, the cuff sign can progress and continue past the ankle. Other forms of edema as static edema, dependent edema, chronic venous insufficiency, CVI and edema as a result of internal trauma need to be considered and, if necessary, ruled out. In more rare occasions Morbus Dercum (painful lipoma, and Madelung's deformity (painless lipomatosis) are also included in the differential diagnosis [Forner 2012]. It should also be noted that a disproportionate fat distribution can also occur in the absence of lipedema, for instance as a racial characteristic. Cellulite is one of the characteristics of lipedema but is also very comment in women. Therefore, people with cellulite can have similarities with lipedema patients, even when the diagnosis is not present. [Terranova 2006]

CONCLUSIONS

Level 2	Anamnesis and physical examination are the most important diagnostic elements. *Forner 212*
Level 2	Laboratory tests can be considered to evaluate underlying medical conditions that may influence disease progression. *Forner 2012*
Level 2	Lipedema has no characteristic abnormalities in blood values. General blood tests are useful, however, because some patients do have underlying medical conditions that may be responsible for some of the complaints. *Forner 2012*
Level 3	CT can be useful for diagnosing lipedema, should alternative diagnostic methods yield no satisfying conclusion. *Forner 2012*
Level 3	When lymphogenic involvement is suspected, quantitative lymphoscintigraphy can be useful to include or exclude involvement of the lymphatic system. *Forner 2012*
Level 4	The working group shares the opinion that only quantitative scans, performed in a standard fashion, can be useful. *Opinion of the working group*
Level 2	The most important differential diagnoses are lipohypertrophy, disproportional fat distribution, edema of any nature, obesity *Forner 2012, Terranova 2006*

OTHER CONSIDERATIONS

Even though lipedema is generally easily recognizable based on anamnesis and physical examination, if there is an excess of midline fat, it may be difficult to determine where lipedema transitions to or combines with lipedema-free adipose tissue.

Lipedema has not yet been included in the International classification of diseases (ICD). There is little controversy about the classic pattern (diagnosis with pain) and concomitant functional impairments, but doubt can exist in borderline cases. To determine the appropriate questions to ask during history taking, the ICF proves a good starting point for a systematic approach.

Early recognition and intervention will definitely lead to cost reduction because this prevents incorrect diagnosis and unnecessary diagnostic tests.

Experience shows that some lipedema patients lose peripheral muscle strength. In many cases, this coincides with increased body weight, where one would normally expect an increase of strength.

This same pattern can be found in patients with sarcopenic obesity. Future research in this direction is needed.

RECOMMENDATIONS

The diagnosis of lipedema can essentially be determined based on the medical history and physical examination. When taking a medical history and performing a physical examination, the criteria established by the working group should at least be addressed [see recommendations in chapter 1]. (minimal norm)

Due to the wide variety of involved complaints, it is recommended that a general health profile (using suitable clinimetrics) be created to document overall functioning. This profile should be used unequivocally during diagnosis and when designing a therapeutic and follow-up regimen.

At a minimum, information to be obtained in the diagnostic phase includes (repetitive) girth measurements of the involved extremities, BMI and abdominal circumference, and establishing the daily activity level. This is also applicable to patients who display (as of yet) too few of the criteria to diagnose early-stage lipedema. Timely recognition and intervention can be regarded as particularly important in the context of functionality and cost reduction.

Recognition and potential treatment of psychosocial problems is important in both the diagnostic and treatment phases.

Additional diagnostics can be performed when a second condition is suspected.

For instance, dynamic lymphatic scanning can be considered when lymphatic involvement is suspected. Endocrine evaluations can be considered when a metabolic disorder is suspected. Venous pathology must be excluded.

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3. TREATMENT

INTRODUCTION

Education is essential in the treatment of lipedema patients. It is often a relief for patients to hear that their problem is recognized by name and cause; however, the lack of an easy solution may lead to disappointment. Recognizing these problems is of great importance, and psychological support may be recommended for some patients. [Langendoen 2009]

PRINCIPLE QUESTION

- What does the treatment and/or care for lipedema patients include?

SCIENTIFIC SUBSTANTIATION

The goals of lipedema treatment are various and include decreasing subjective complaints and functional limitations and preventing progression of the lipedema. The treatment should furthermore target factors that negatively influence lipedema (such as obesity, the presence of lymphatic or venous edema, incorrect insight into the condition and decreased physical activity).

A causal treatment is currently unavailable. Treatment should be based on several of the following pillars:

- When necessary, (re)developing a healthy lifestyle;
- Stimulating the vascular / lymphatic pump by means of compression of the calf muscles;
- Combating the edema component when present;
- Reducing pain complaints, where possible;
- Improving muscle strength and stamina;
- Alternatives (dietary precautions, psychological support);
- Reduction of the lipedema (liposuction by means of TLA and PC).

3.1 CONSERVATIVE TREATMENT

The standard conservative therapy for lipedema is significantly different from that of lymphedema. Some components such as compression therapy, (re)developing an active lifestyle and weight loss programs are mentioned, however. [Forner 2012] For a summary of conservative treatment, see [Annex 2: Flowchart conservative treatment].

(RE)DEVELOPING A HEALTHY LIFE STYLE (EXERCISE, NUTRITION)

As lipedema is a chronic, incurable condition, it can be accompanied by pain, loss of strength and deconditioning; in more severe cases, lipedema can lead to deterioration in daily activity levels. A chronic shortage of physical exertion is a risk factor for further deterioration of lipedema, especially in combination with an increase in body weight.

Interventions aimed at (re)gaining and maintaining a sufficiently active lifestyle are therapy-based and are aimed at building muscle strength and general fitness in the initial phase. These interventions generally take place under allied health professionals. The ultimate goal of the therapeutic intervention is improving strength and fitness to enable an active lifestyle, after which further guidance becomes unnecessary. Obesi-

ty is also an important modifiable factor, for which multidisciplinary cooperation with a dietician is necessary. Exercise aimed at reducing body fat is part of the therapeutic approach in this case.

It is a given that lipedema patients without accompanying complaints often already have a sufficiently active lifestyle, due to their efforts to keep their "heavy legs" under control.

Although dietary changes cannot prevent the disproportionate fat distribution in patients with lipedema, dietary modifications can improve prognosis and general well being, as well as reduce obesity. [Langendoen 2009; Reich-Schupke 2013]

COMBATING THE EDEMA COMPONENT (WHEN PRESENT)

Before beginning treatment, it must be determined whether there is a relevant edema component that exists separately from the lipedema. For this purpose, compression therapy is useful. Compression therapy and therapeutic compression hosiery do not result in a decrease of fat deposits but can help to prevent further edema formation, stimulation of arterial, venous and lymphatic flow and thereby reducing coinciding issues. Patients receiving compression therapy are often more mobile than those who do not. From the perspective of stimulating circulation (venous, arterial or lymphatic), wearing light (class two) compression hosiery can be advantageous. However, one should keep in mind that wearing round knitted hosiery that causes ligation of the ankles has the opposite effect. It is therefore preferred to provide flat knitted therapeutic compression hosiery; however, in a few exceptional cases, round knitted hosiery does suffice, in contrast to statements in the current guidelines. [Meier-Volrath 2005; Wienert 2006].

It has not been proven that manual lymphatic drainage or MLD has a role in the treatment of lipedema [Reich-Schupke 2013, Langendoen 2009]. If lipedema is combined with lymphedema, one should refer to the lymphedema directive. Apart from that, the edema component in lipedema is often caused by dynamic insufficiency, leaving the lymphatic system unaffected. [Földi 2005]

The sensitivity of the skin to palpation in lipedema patients is difficult to explain. It is unknown if hypersensitivity is caused by nociceptive pain (due to stimulation of the nerve endings by nociceptors), neuropathic pain (due to damaged nerve fibers), or central sensitization (due to an increased response of the central nervous system to sensory information from the periphery), making the treatment of pain difficult. Pain-relieving interventions may have a place in the initial treatment phase, as pain relief may increase the patient's capability to initiate a healthier and more active lifestyle. Additionally, the perception of illness can be addressed in an educational program for patients with chronic pain. Changing a non-desirable perception of illness can contribute to treatment of the sequelae of chronic pain. [Work group opinion]

ALTERNATIVES

In case of abnormalities in the posture or movement of a patient, correction by means of orthotics or orthopedic referral is indicated. The stance of the foot and knee is of particular importance, due to the prevalent pes planus and valgus stances. In case of blemishes, skin care may be indicated.

Recognizing these problems is crucial, and psychosocial support can be required for some patients [Langendoen 2009]. When psychosocial problems are perceived, referral to suitable support is indicated.

CONCLUSIONS

Level 2	Exercise, weight loss programs and reconditioning according to a graded activity program and, when indicated, physiotherapy form important pillars of conservative treatment. *Damstra 2013, Reich-Schupke 2013*
Level 2	Dieting and altering eating patterns can improve the prognosis and general well being in patients with obesity and excessive caloric intake, when these interventions are combined with increased exercise. *Langendoen 2009; Reich-Schupke 2013*
Level 2	Compression therapy and therapeutic compression hosiery can reduce edema and its accompanying distress. *Reich-Schupke 2013; Meier-Vollrath 2005; Wienert 2006*
Level 3	It has not been proven that manual lymphatic drainage (MLD) plays a role in the treatment of lipedema. If lipedema is combined with lymphedema, one can refer to the lymphedema directive. *Langendoen 2009; Reich-Schupke 2013;*
Level 2	Recognizing the problem is of high importance, and some patients require psychosocial support. [*Langendoen 2009]*

OTHER CONSIDERATIONS

Lipedema often requires persistent lifestyle changes that are focused on achieving and maintaining a sufficiently active lifestyle and a healthy body weight. The patient needs motivation and stimulation to continue the "new life style" in which the patient needs to have an active role (active coping style).

Promising strategies for musculoskeletal pain in adults include supervised (personalized) remedial therapy, repetition sessions and education and are based on graded activity. They furthermore include self-management programs and cognitive behavioral therapy [Jordan et al.]. Research regarding the efficacy of these treatment strategies has not been conducted on the lipedema population, yet the work group deems it likely that these conclusions are equally applicable to lipedema patients.

RECOMMENDATIONS

The lipedema patient must, should he not already do so himself, be encouraged to maintain an active lifestyle according to the Dutch Norm for Healthy Exercise (Nederlandse Norm Gezond Bewegen or NNGB) and a healthy body weight [Annex 3].

A patient with lipedema combined with an unhealthy lifestyle and/or functional limitations requires multidisciplinary treatment with suitable clinimetrics. Emphasis should be placed on stimulating the vascular and lymphatic pumps, determining caloric balance (food intake vs. caloric burn) and engaging in a meticulous exercise program that is based on developing muscular strength and fitness (using the graded activity method), possibly in combination with a weight loss and exercise program.

Treatment and support of lipedema patients requires expertise in the areas of training, graded activity and cognitive behavioral principles, supported by the use of suitable clinimetrics. Intensive coaching and proficiency in motivational interviewing are recommended to assure patient independence.

Medical providers should recognize psychosocial problems and provide basic psychological aid (and if necessary referral to a specialist) in patients with lipedema.

3.2 SURGICAL TREATMENT

Two surgical treatments are used to treat lipedema. The most commonly performed treatment is liposuction. Preceding studies have shown that liposuction under general anesthesia and/or that is performed without subcutaneous infiltration (or with relatively little subcutaneous infiltration) is contraindicated for lipedema because of the substantial risk of causing damage to the lymphatic system. [Forner 2012]

With the introduction of tumescent local anesthesia (TLA) and the subsequent use of supertumescent local anesthesia (STLA), the risk of damaging the lymphatic system during liposuction has dramatically decreased. Liposuction using STLA and vibrating cannulas is an effective treatment for lipedema [Habbema 2009; Fife 2010; Forner 2012; Rapprich 2011; Schmeller 2012].

Although these treatments are non-curative, they can improve functionality, pain, swelling, physical appearance and the quality of life [Forner 2012; Rapprich 2011; Schmeller 2012].

Multiple sessions are necessary to adequately treat the extremities circumferentially and along the entire length. To prevent local recurrence, as much of the subcutaneous fat tissue should be removed as possible; the boundaries of safety and cosmetic consequences should obviously be considered. It is presumed that as a result no new fat cells will be formed, or that at worst, the few remaining overlooked cells will only cause minor visible growth of the tissue. Recent reports show that over a longer period of time, volume reduction occurs, and conservative treatment can be terminated or reduced in over 65% of the patients treated. In the remaining patients, quality of life, sensitivity and mobility are notably improved, yet conservative treatment remains necessary. [Schmeller 2007; Schmeller 2012]

It should be obvious that, even after liposuction, a healthy lifestyle should be maintained and attention should be paid to caloric intake. A reduction in pain, as well as improved mobility and increased general well-being should facilitate this for the patient after treatment.

If there is a lymphedema component prior to treatment, one should keep in mind that this component will still be present after treatment, and the appropriate therapy needs to continue. Experience shows that in some patients, the lymphedema component also improves or resolves after liposuction.

A second treatment method involves excision. Large localized deposits of lipedematous tissue ("lumps") can form in lipedematous areas on the legs and cause serious mechanical difficulties, valgus deformity of the knees or even the total inability to walk. In such cases, surgery can be considered, although minimally invasive action using liposuction is preferred. However, sometimes longitudinal excision (lumpectomy) is

the only treatment option for severe mechanical limitations (for instance, severe ptosis). If varicose veins are present, minimally invasive procedures can be considered.

CONCLUSIONS

Level 3	Liposuction with tumescent local anesthesia (TLA) and vibrating cannulas is an effective method for the treatment of lipedema. It restores patient function, reduces (hyper) sensitivity and swelling, and improves the physical appearance as well as the quality of life. *Langendoen 2009; Habbema 2009; Fife 2010; Schmeller 2012*
Level 4	Lipedema can cause large localized deposits of lipedematous tissue ("lumps") to form in the legs. Sometimes longitudinal excision (lumpectomy) is the only option to remove severe mechanical limitations. *Work group opinion*

OTHER CONSIDERATIONS

Liposuction under STLA with powered cannulas (PC) leads to a direct improvement in nearly all lipedema patients. Because the necessity for conservative treatment is likewise reduced after liposuction, the health care service expenditures may also decrease. Calculations regarding these issues have not been made.

Liposuction under TLA with PC is a highly specialized treatment. Therefore, treatment will only be available in a limited number of specialized centers and for a select group of patients. A rational approach would be to first offer conservative treatment and offering surgical alternatives to highly selected patients who do not sufficiently benefit from conservative treatment.

Liposuction under TLA with PC should be performed according to the applicable field norms.

RECOMMENDATION

Liposuction and/or reductive surgery are a part of the treatment for lipedema and should take place in specialized centers, where these procedures should be performed by a multidisciplinary team in accordance with standard protocols and with the backing of good clinimetrics.

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4. PATIENT ORGANISATIONS

INTRODUCTION

A patient organization is a best interest organization aimed at, for instance, facilitating interpatient contact, representing patient interests and/or providing information and education for affiliated patients or third parties. In the Netherlands, the Dutch Network for Lymphedema and Lipedema (*Nederlands Netwerk voor lymphoedeem en lipoedeem* or NLNet), the Dutch Lipedema Day Foundation (*Stichting Nederlandse Lipoedeemdag)* and the Lipedema Patient Association (*Lipoedeem Patiënten Vereniging* or LIPV) are active on the area of lipedema.

PRINCIPLE QUESTION

- What is the role of patient organizations in the care of lipedema patients?

SCIENTIFIC SUBSTANTIATION / DISCUSSION

Literature that specifically applies to the principal question was not identified. The Dutch Network for Lymphedema and Lipedema (*Nederlands Netwerk voor lymphoedeem en lipoedeem* or NLNet; www.lymfoedeem.nl) has been active since 2006 in the Netherlands. The website is HON-certified (Health on the net foundation). This patient organization was originally aimed at patients with lymphedema or who were at risk of developing lymphedema (see Lymphedema Directive 2014) and at lipedema patients with a lymphatic component. They have increasingly targeted lipedema in recent years as well, however. NLNet has ANBI status (Allowed by the government to accept inherence) and receives PGO subsidies (governmental grant for patient organizations) to support its efforts. There were more than 1400 benefactors in 2013.

NLNet is embedded in the Dutch subscription bases and provides Dutch patient interest representation through *IEDER(in)*, the umbrella organization for people with physical and mental handicaps or chronic illnesses (IEDER(*in*); www.iederin.nl), the Dutch Patient Consumer Federation (*Nederlandse Patiënten Consumenten Federatie* or NPCF; www.npcf.nl) and Skin Patients Netherlands (*Huidpatiënten Nederland* or HPN; www.huidpatienten-nederland.nl). NLNet has an informative website, organizes a biennial congress and publishes a biannual magazine (Lymphologica). Support groups are organized twice yearly, and certified instructors in every region teach support in self-management courses for patients. Furthermore, patients are kept fully informed through social media (Facebook and Twitter) about the newest developments concerning lymphedema and lipedema.

The Lipedema Patient Association (*Lipedema Patiënten Vereniging* or LIPV; www.lipv.nl) has been active since April 2009 in the Netherlands. It was established because, at the time, no active patient organization existed specifically for lipedema patients. Apart from providing information and initiating inter-patient contact, the LIPV issues a minimum of six newsletters each year. They sponsor lipedema symposia and organize national meetings as well as occasional regional meetings.

The Dutch Lipedema Day Foundation (*Stichting Nederlandse Lipoedeemdag;* http://lipoedeemdag.nl) organizes a biennial congress on lipedema.

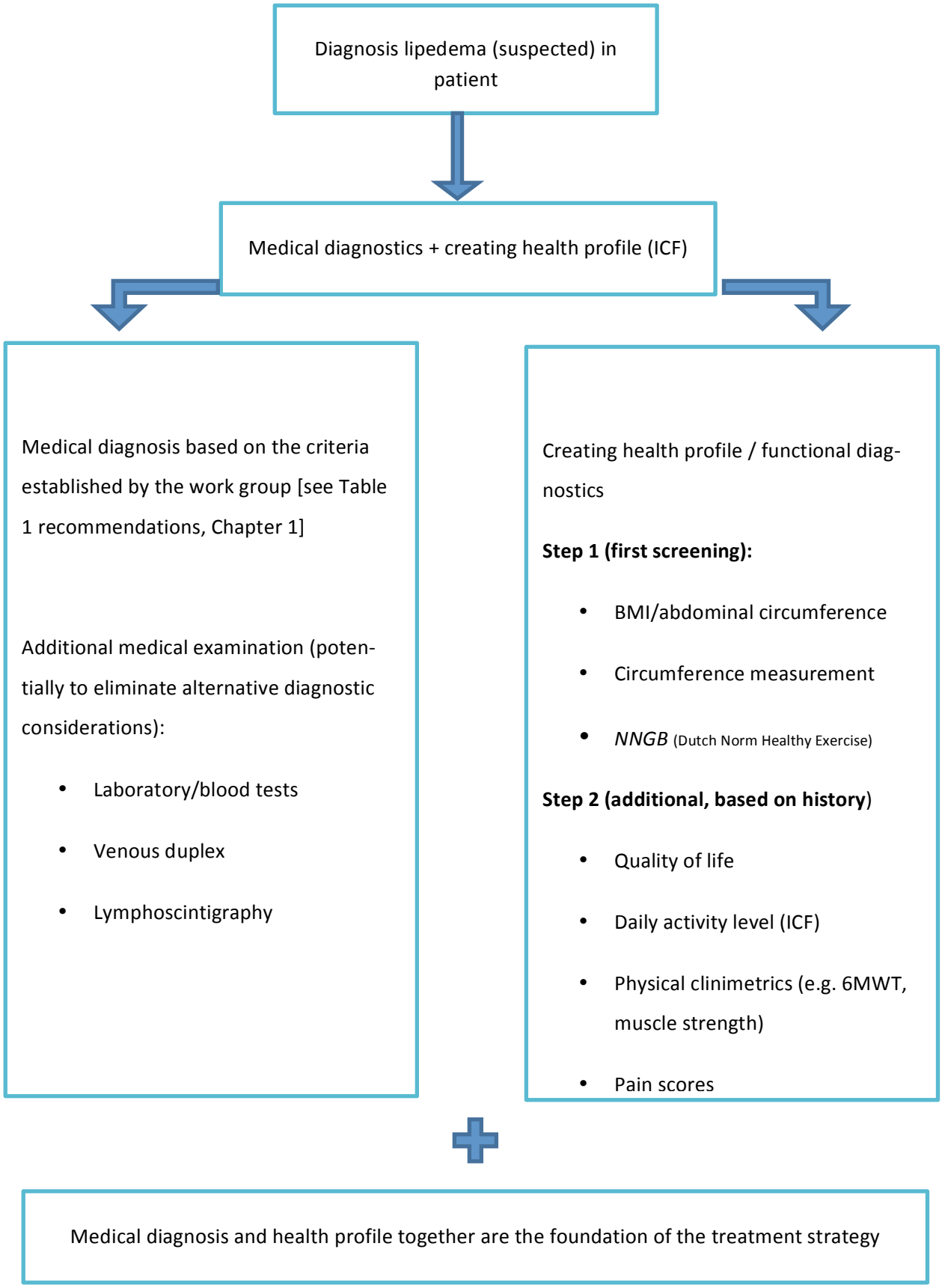
CONCLUSION

No level	Patient organizations represent the best interests of patients. These organizations are an experienced source of information, provide opportunities for inter-patient contact and contribute to the quality of care for lipedema patients. Group interests are furthermore supported.

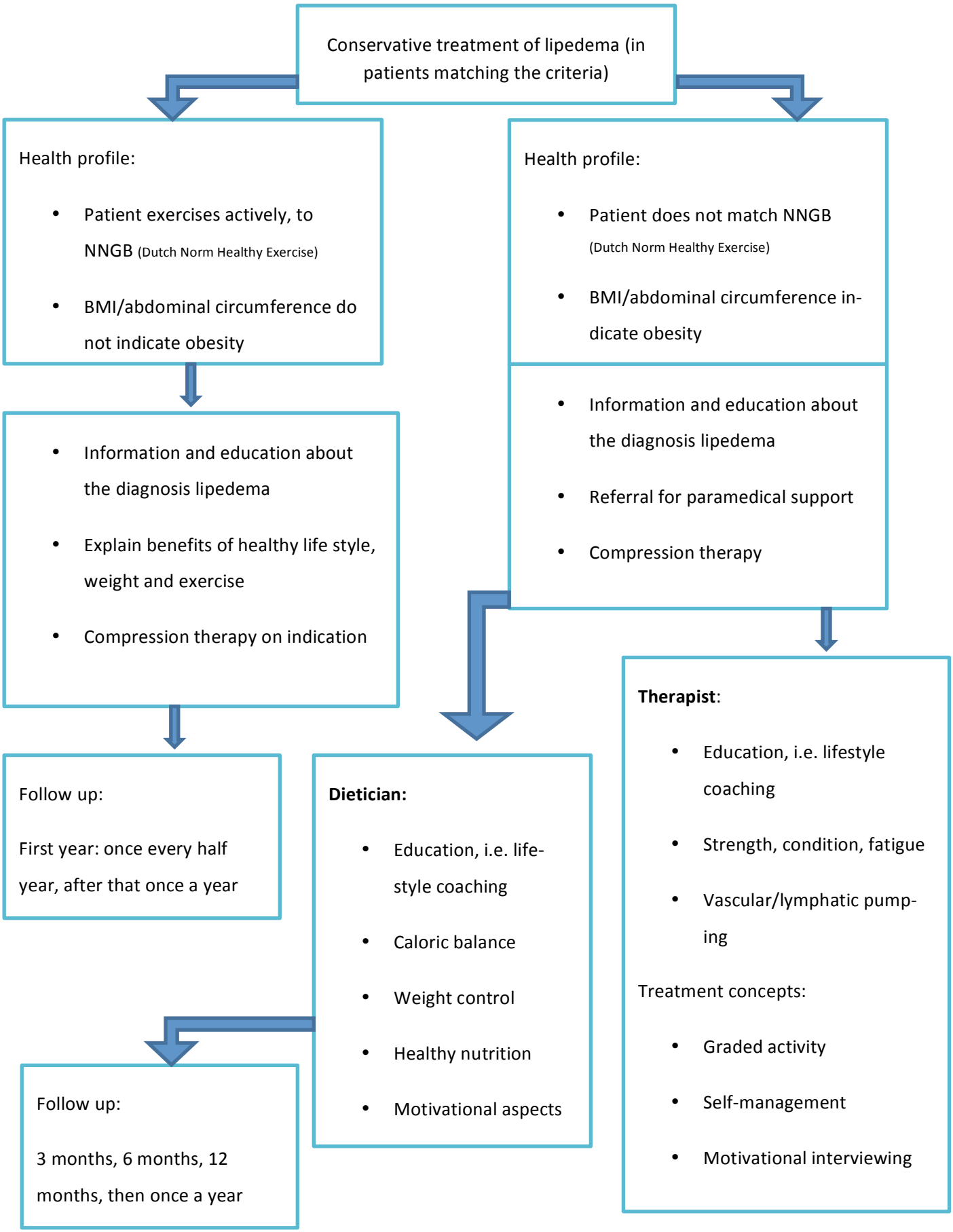
RECOMMENDATION

It is appropriate to note the existence of patient organizations and their important role in awareness and advocacy of lipedema in general. In the Netherlands the *NLNet and* the *Lipoedeem Patiënten Vereniging* (LIPV) cooperated in the process of development of the guideline 2014

APPENDIX 1: FLOWCHART FOR DIAGNOSING LIPEDEMA



APPENDIX 2: FLOWCHART FOR CONSERVATIVE LIPEDEMA TREATMENT



APPENDIX 3: DUTCH NORM FOR HEALTHY EXERCISE

NNGB

De Nederlandse Norm Gezond Bewegen (Dutch Norm for Healthy Exercise or NNGB)

Do you exercise enough? There are norms to assure you exercise enough. In the Netherlands, the following norms have been developed:

- De Nederlandse Norm Gezond Bewegen (NNGB)
- Fitnorm

There is an additional norm based on 10,000 steps a day. You will find information on the NNGB and chronic conditions on this page.

DUTCH NORM FOR HEALTHY EXERCISE (NNGB)

The Dutch Norm for Healthy Exercise gives general advice concerning exercise and well-being. Following this advice will contribute to a reduction in the risk of cardiovascular disease. The NNGB was established in 1998 and derived from international guidelines. The norm differs by age group and is based on a minimum of thirty minutes of exercise of average intensity, a minimum of five days a week. The advice aims to increase walking, cycling, gardening, swimming and the like. The norm mostly targets the heart and muscles.

THE NNGB PER AGE GROUP

1. Youth (under 18): one hour of moderately intense physical exercise daily, in which the activities are aimed at improving physical fitness (strength, agility and coordination) at least two times a week.
2. Adults (18-55): half an hour of moderately intense physical exercise on at least five days per week, but preferably daily.
3. 55-plus: half an hour of moderately intense physical exercise on at least five days per week, but preferably daily.
4. For inactive people, with or without physical limitations, any extra amount of physical exercise is a bonus.

The above stated norms can be seen as a recommendation regarding the minimum level of exercise. Up to a certain level, extra exercise will yield additional health benefits.

WHAT DOES “MODERATELY INTENSE PHYSICAL EXERCISE” MEAN?

This vague description becomes clearer when thought of in terms of our energy levels when idle. When idle, for instance when sitting or lying, we also require energy. The amount of energy required depends on our weight. The total idle energy usage is, as a result, different for everyone. The total idle energy usage for each individual is known as MET, which is short for metabolic equivalent and is a measurement of metabolic processes. Our MET-value when idle is 1. A MET-value of 2 therefore means double the amount of energy required than when idle. Once the idle value is known, "moderately intense" can be described. For adults, this means a MET-value between 4 and 6.5. Activities corresponding to this level are: walking 5 km/h = 4 MET, cycling 16 km/h = 6.5 MET. For youths, MET-values of 5 (cycling) to 8 (running) can be used. For adults 55 years of age and older, this means MET-values of 3 (walking 4 km/h) to 5 (cycling 10-12 km/h). For a summary of activities with corresponding MET-values, please consult the table on the next page.

Activity	MET-value
Idle (lying, sitting, standing at ease, eating, speaking)	1.0
Driving, playing the piano, computing, typing	2.0
Walking 4 km / hour	3.0
Walking 5 km / hour	4.0
Cycling 10-12 km / hour	5.0
Cycling 16 km / hour	6.5
Swimming (crawl) 1 km / hour	5.0
Swimming (crawl) 3 km / hour	20.0
Running / jogging	8.0

THE RELATION BETWEEN THE AMOUNT OF STEPS PER DAY AND THE DUTCH NORM FOR HEALTHY EXERCISE

Walking is possibly the safest and most suitable form of physical exercise for much of the population, including people who are physically inactive, obese, or have chronic conditions. People who walk less than 5,000 steps a day can be considered physically inactive. Those who take 10,000 steps a day can be considered physically active. People who take more than 12,500 steps per day are considered very active. The concept of "ten thousand steps a day" originated in Japan, where it has been common knowledge for decades. Walking 10,000 steps a day corresponds with an energy usage of approximately 400 kcal a day. The NNGB for adults corresponds with an energy usage of approximately 200 kcal a day. To measure the steps one can use a pedometer.

The difference between the "ten thousand steps a day" concept and the NNGB can be explained by the fact that the NNGB assumes moderately intense physical exercise, whereas the original concept involves all forms of activity, without limiting the conditions thereof. For instance, walking 30 minutes while shopping does not count for the NNGB, yet it does for the original "ten thousand steps a day" concept. Research shows that as a result of activities in daily life, people tend to walk approximately 6,000 steps but at a lower intensity. It has also been calculated that a healthy adult will roughly walk 4,000 steps during a half-hour walk. When all steps are added together, the goal of 10,000 steps can be reached.

HEALTHY OR FIT?

There is an important distinction between improving your health and increasing your fitness. Health benefits are already achieved with low- to average-intensity activities. Walking, cycling, climbing stairs, gardening and the like already provide major health benefits. However, to become fit, more is required. This is because fitness is defined differently. An internationally used definition for physical fitness is 'the capacity to perform physical labor to satisfaction'. Fitness improvement cannot be realized with moderately intense movement.

The NNGB provides general advice concerning exercise and well being. Following this advice will generally result in health benefits and a reduction in cardiovascular problems. Fitness improvements should not be expected of the norm.

People who are already "in shape" should continue this level of norm-activity. Fitness protects against cardiovascular problems as well as many other prevalent lifestyle-related diseases.

FITNORM

The Fitnorm for adults assumes at least 20 minutes of intensive physical exercise, at least three times a week. This involves activities that significantly increase heart rate (110-130 beats/minute), breathing rate and sweat production, such as sports or plowing a garden. This activity level leads also to the burn of adipose tissue.

Distinctions are made between:

- Non-fit: physically intensive exercise only a few times a year or not at all
- Semi-fit: regular physically intensive exercise, but less than three times a week
- Normfit: at least 20 minutes of intensive physical exercise, three times a week or more

Exercise to improve fitness requires greater effort than exercise to improve your general health.

THE NNGB AND CHRONIC CONDITIONS

The Dutch Norm for Healthy Exercise provides general advice concerning exercise and well-being. Following this advice will primarily contribute to a reduction in the risk of cardiovascular disease. However, the norm only has limited applicability to people with physical impediments and/or chronic conditions and may exclude people with a potentially higher risk of cardiovascular problems. Fitness improvements should not be expected of the norm either. People who are already "in shape" should not be encouraged to reduce their activity levels. Fitness not only protects against cardiovascular problems but many other prevalent lifestyle-related diseases. The IBV will readdress these issues in the future.

So, what good is the NNGB if you have preexisting heart and/or muscle problems? Can you follow this advice blindly? The Institute of Exercise Questions (Instituut voor BewegingsVraagstukken or IBV) maintains that you cannot. Yet, how (much) should you exercise if you have a different chronic condition? Does the NNGB apply, for instance, to people with osteoporosis? What quantity of exercise should be advised for obese people?

As we have previously stated in the NNGB article, fitness improvements should not be expected from following the norm. Again, people who are already "in shape" should not be encouraged to reduce their activity levels. The Institute of Exercise Questions (IBV) has suggestions and recommendations for adaptations of the norms for specific target audiences. Recommendations have been made for people suffering from osteoporosis or obesity. These recommendations can be found on the IBV website. (http://www.bewegingsvraagstukken.nl)