7 Lipedema

W. Schmeller, I. Meier-Vollrath

Lipedema was first described by Allen and Hines in 1940 (1). Today, however, a large number of physicians remain unaware of the disease and its symptoms. Many lipedema patients are not correctly diagnosed or effectively treated until after they have endured decades of suffering (2).

Epidemiological statistics on the incidence of lipedema are not available. An investigation of patients treated at one lymphedema clinic from 1995 to 1996 revealed lipedema in approximately 15% of the patients treated on an inpatient basis (3). A similar number (8% to 17%) was reported in surveys carried out in 2003 in four lymphedema clinics in Germany (4).

In contrast to lymphedema, literature on lipedema is extremely sparse. Lipedema findings in the Anglo-American literature in particular often report only isolated case studies (5-8). Many more publications, including a monograph, are available in German literature (9). Table 7-1 lists synonyms for lipedema.

- **Lipalgia**
- **Adiposalgia/Adipoalgesia**
- **Adiposis dolorosa**
- **Lipomatosis dolorosa of the legs**
- **Lipohypertrophy dolorosa**
- **Painful column leg**
- **Painful lipedema syndrome**

Table 7-1)
Synonyms for lipedema

Figure 7-1)
Stage I lipedema: Disproportion of the upper and lower body

7.1 Definition

A chronically progressive, symmetrical accumulation of fat in the subcutaneous tissue with orthostatic edema occurring almost exclusively in women. Primarily the lower extremities are affected (Figure 7-1). It may occur in combination with the upper extremities. Lipedema is characterized by tenderness and easy bruising.

7.2 Pathogenesis

Lipedema occurs almost exclusively in women. The rare cases in which men are affected involve severe hormonal functional disorders such as testosterone deficiency. Figure 7-2 shows a patient with ethyl-toxic cirrhosis of the liver.

Examinations of 119 lipedema patients carried out in the 1940s showed initial manifestation of the disease distributed evenly across the various decades of life (10). In contrast, newer literature emphasizes that the disease generally manifests during puberty. However, our own experience

Figure 7-2)
Lipedema in a man with ethyl-toxic cirrhosis of the liver
(Courtesy of C. Schuchhardt, MD, St. Blasien-Menzenschwand)

shows that onset also frequently occurs after pregnancy. In 100 of the patients assessed in recent years, disease onset occurred between the ages of 30 and 40 (11).

In addition to hormonal impacts, a genetic predisposition is assumed since frequently several women in one family have the disease. However, related statistics vary widely, ranging from 16% to 64% of the cases (1, 10, 12). Damage to the autonomic nervous system has been reported in isolated cases (13); however, its role in pathogenesis is unclear. It is not certain whether observed changes in microcirculation (14, 15) actually trigger the disease or are secondary phenomena. Also, it is not clear whether the adipokines released by adipose cells in obese patients play a role in lipedema (16). In contrast to obesity, however, the fat cannot be "starved off." In addition, the typical complications of obesity do not occur with lipedema. Consequently, other receptors, and possibly even another type of fat cell, may be responsible.

- **Hyperplasia/Hypertrophy of fatty tissue**
- **Increased permeability of blood vessels**
- **Fluid and protein accumulation in subcutaneous fatty tissue**
- **Orthostasis**
- **Increased fragility of blood vessels**

Table 7-2)
Key factors in lipedema

7.3 Pathophysiology

Lipedema is caused by several factors (Table 7-2). The most striking visual symptom is the disproportion between the upper and lower body (Figure 7-1) due to the pathological fat deposits. It is unclear whether this is a hypertrophy of the fat cells, hyperplasia or a combination of the two. Another important factor is increased capillary permeability, which leads to increased accumulation of fluid and protein in the interstitium, causing orthostatic edema (14, 17). The extent of this accumulation, and not the absolute fat volume, is the major reason for sensitivity of the tissue to pressure and touch (18). Increased capillary fragility causes the frequently observed easy bruising. Earlier it was assumed that the increasing tissue pressure caused by fat and edema caused mechanical draining impairment of initial lymphatics, precollectors and collectors. The temporary edema of the thighs and lower legs occurring in the evening causes increased volume during the day which overtaxes the intact lymphatic system (dynamic or high volume insufficiency). However, decompensation does not occur until the increased lymphatic load has exceeded the available transport capacity for years or even decades (19) due to the exhaustion of the functional reserves of the lymphatic system (mechanical or low volume insufficiency). Secondary changes of the lymph collectors are major factors. Abacterial inflammation with consecutive lymphangiosclerosis and perilymphovascular fibrosis occurs due to persistent protein accumulation in the interstitial tissue, leading to a reduction of the transported volume. In this late phase of the disease, a combination of increased lymphatic waste and reduced transport

Figure 7-3)
Lipohypertrophy. Morphologically, lipohypertrophy can resemble lipedema

Figure 7-4)
Lipedema. In addition to the change in shape, other symptoms are associated with lipedema

occurs, giving rise to secondary lymphedema (lipolymphedema). Lipedema, per se, is not a primarily lymphological disease.

It is repeatedly reported that lipedema develops over a period of decades from lipohypertrophy (see "Differential Diagnosis") (20, 21). Lipedema and lipohypertrophy can have a very similar morphology (Figure 7-3 and Figure 7-4). This would mean that the painful clinical picture of lipedema is preceded by a pain-free stage. Lipedema can occur very early in puberty, while lipohypertrophy most frequently occurs late in life without major edema or pain. Whether one disease transitions to the other, and how this occurs, is unclear. It appears there are other relevant factors such as genetics, hormones, and possibly even circulatory disorders and neural changes, as well as age. They are responsible for the increase in capillary permeability and fragility which causes the disease and its characteristic symptoms.

7.4 Pathology

The histological changes occurring in lipedema are not sufficient diagnostic indicators. In addition to an increase in fat cells, some of which are hypertrophic, a high volume of capillary blood vessels is found in the interstitial tissue. Perivascularly there are macrophages, fibroblasts, mast cells and isolated necrotic adipose tissue. In the late stages of the disease, the fibrotic part increases (14). The descriptive term edematous fibrosclerotic panniculopathy (22) is now obsolete. The histology is generally identical to that of dermatoliposclerosis. This is apparently an unspecific inflammatory reaction of the tissue similar to that which occurs in chronic venous insufficiency (23).

7.5 Diagnostic Procedures

Lipedema can generally be diagnosed on the basis of clinical criteria. Typical symptoms are the time of onset, the symmetrical distribution of fat, edema, sensitivity to pressure and easy bruising. Generally, instrumental or invasive diagnostic procedures are not required. In the case of lipolymphedema, imaging procedures (indirect lymphangiography, lymphoscintigraphic function test) can be used to provide additional quantification of morphological and functional changes of the lymphatic system.

Stage I	Skin surface normal, nodular (small) fatty tissue structure
Stage II	Skin surface uneven (peau d'orange), nodular (big) fatty tissue structure
Stage III	Lobular deformation due to increased fatty tissue

Table 7-3)
Grading of lipedema

7.6 Results

Clinical results

Patient history: Onset of disease is usually during puberty, frequently following pregnancy or not until after menopause. In some cases, a simultaneous large weight gain is reported. However, patients with stage I lipedema often are not overweight. Women with lipedema report a spontaneously occurring feeling of tension or swelling as well as a noticeable hypersensitivity to touch and pressure in the thighs and lower legs. The complaints generally become worse as the day progresses, particularly after standing or sitting for long periods. The severity of the symptoms is not related to the amount of accumulated fat. In addition, edema, generally occurring in the evening and with higher frequency in warm weather, is reported. Patients also report that even minor bumps result in bruising. Small injuries that cause no macroscopic lesions in healthy people can result in hematoma when lipedematous extremities are affected. Generally, symptoms are less severe in the upper extremities than in the lower extremities. The patients' appearance causes them major distress. This problem is mentioned in the very earliest publications on the disease (1, 10). Many women describe their frustration as their condition deteriorates, often leading them to overeat and consequently gain weight. Thus it is not surprising that around half the patients end up suffering from obesity in addition to the lipedema (24). Secondary lymph flow impairment in the form of lipolymphedema occurs

Figure 7-5)
Stage 1 lipedema

more frequently among overweight women than in women with normal weight (25).

Inspection: Most patients present a marked discrepancy between their slim upper body and corpulent lower body. For approximately 97% of the women with lipedema, the lower extremities (thighs and hips, often the lower legs as well) are affected. The upper arms are affected in approximately 30% of lipedema patients; the forearms to the wrist can be affected as well (3).

Lipedema is chronic and usually progressive with various stages (Table 7-3). Figures 7-5 to 7-8 show various degrees of severity. In the early stage, the skin is smooth with a thickened subcutaneous layer with evenly distributed small nodules (stage I). Over time, the nodules become enlarged causing the skin surface to be uneven. It may resemble the peel of an orange (stage II), referred to by laypeople as "cellulite." In advanced forms, the subcutaneous tissue becomes increasingly indurated. Ultimately, bulging protrusions of fat occur on the thighs and knees that can interfere with normal gait (stage III). The affected tissue is tender, generally

Figure 7-6)
Stage II lipedema

Figure 7-7)
Stage III lipedema

Figure 7-8)
Severe stage III lipedema
(Courtesy of W. J. Brauer, MD, Emmendingen)

relative to the extent of the edema. The skin may be cool at certain places (13, 26). Sometimes bruising occurs without the patient recalling the trauma that may have caused it. In stage III, weeping erosions in areas affected by intertrigo below the groin or between the thighs may appear. Lipedema symptoms do not include edema of the dorsal foot or toes (Figure 7-9). If this occurs during later stages of lipedema as a result of secondary lymph flow obstruction, lipolymphedema has developed. The skin is thickened on the toes and cannot be lifted (Figure 7-10; positive Stemmer sign).

Palpation: The localized accumulation of fat is not pitting, which is only the case with an obvious secondary edema. In stage I, the subcutaneous tissue is soft and its structure is either even or is characterized by tiny nodules. In stage II, nodules can be felt underneath the skin. Symptoms are extreme in stage III. The increase in volume in the extremities may end abruptly above the ankle (Figure 7-6 and 7-9) or wrist with a fatty cuff. In German, this condition used to be referred to as "Turkish pants phenomenon". Similar descriptions of the condition, including "jodhpur-like riding breeches", continue to be used today. Localized fat de-

Figure 7-9)
Lipedema. No edema of the ankles, feet or toes

posits occur, sometimes drooping down over the knees like an apron. Generally there is pronounced hardening of the subcutaneous tissue, especially in patients with lipolymphedema. This is attributed to fibrosclerosis of the interlobular septa (14, 27).

7.7 Differential Diagnosis

Lipedema must be differentiated from **lipohypertrophy** (Figure 7-3), which in women is also characterized by a disproportionate body shape due to symmetrical fat deposits in the hips and legs and a slender trunk. The terms localized adiposity (fat pad) and gluteofemoral obesity are used to describe the most frequent form of lipohypertrophy; the upper extremities are rarely affected. While the morphological changes may be practically the same, in lipohypertrophy no edema is involved. Therefore, these women are without pain or hypersensitivity to touch, which means they do not require treatment. However, over time, lipohypertrophy may develop into lipedema (20, 22).

Lipedema is frequently mistaken for **lymphedema.** Table 7-4 shows the major differences between the two diseases. Primary lymphedema gen-

Figure 7-10)
Lipolymphedema, left side more pronounced than the right side. Swelling of the dorsal foot and thickened skin folds on the toes

erally occurs in women, usually in puberty, with unilateral or bilateral swelling of the lower extremities. When it is bilateral, it is always asymmetrical. In primary lymphedema, swelling typically begins at the toes and then reaches the thigh. With lipedema, the increase of volume usually begins in the thigh. In lymphedema patients, the Stemmer skin fold sign is positive, while in lipedema patients it is negative. Hypersensitivity of the tissue or easy bruising are not present. However, combinations of lymphedema and lipedema (lipolymphedema) are possible.

Phlebedema is a symptom of chronic venous insufficiency (CVI). It can occur in both men and women, unilaterally or bilaterally, and precedes the typical cutaneous and subcutaneous changes (stasis dermatitis, hyperpigmentation, panniculitis, dermatoliposclerosis, ulceration). Functional examinations such as ultrasound, light reflection rheography and duplex ultrasound yield pathological results. This is not the case for lipedema. Combination forms (phlebolipedema) exist here as well, however (21).

Dercum's disease (adiposis dolorosa) occurs almost exclusively in women and is characterized by painful, symmetrical fat deposits primarily in the legs. Feet are not affected. In contrast to lipedema, the disease

Lipedema	Lymphedema
Symmetric	Asymmetric
Painful	Painless
Hematoma	No hematoma
No erysipelas	Erysipelas frequent
No foot edema	Foot edema
Stemmer sign negative	Stemmer sign positiv

Table 7-4)
Differentiation between lipedema and lymphedema

does not manifest until the onset of menopause. Edema is not a symptom. Often, muscle weakness is reported. Alcohol abuse, emotional instability and depression are frequently present (28). It is unclear whether Dercum's disease is actually a separate entity or rather a special form of late-manifesting lipedema.

Benign symmetrical Launois-Bensaude lipomatosis (Madelung's disease) is characterized by a diffuse accumulation of fatty tissue in the neck and throat region (type I), shoulders and upper arms (type II) (Figure 7-11) or the pelvic region (type III) (Figure 7-12). This disease is reported to affect men more than women. Alcohol abuse and cirrhosis of the liver are nearly always present in the patient history (29).

Lipedema is often confused with **obesity**. However, in the case of obesity, generally, overweight is caused by a usually soft accumulation of fatty tissue either in the trunk (truncal obesity) or the entire body. Here the fatty tissue is not painful and the proportions between the trunk and the extremities can be normal (28). Both men and women are affected. Genetic factors (including the leptin gene and mutations in the melancortin-4 receptor gene) and overeating, in particular, are responsible (30). The proteins (adipokines) secreted from hypertrophic fat cells are considered to be responsible for the occurrence of the characteristic

Figure 7-11)
Madelung's disease manifested in the shoulders and upper arms of a 52-year-old woman (type II)

complications of insulin resistance, dyslipidemia and hypertension (16). Therapy emphasizes weight reduction through calorie restriction (dieting) or increased burning of calories (physical exercise).

The major clinical characteristics of the above-mentioned diseases are listed in Table 7-5.

Imaging methods

Computed tomography (CT) and magnetic resonance imaging (MRI) can be used to assess the extent and location of fat deposits in lipedema (27, 31, 32). Ultrasound can also be used to assess quantitative and qualitative aspects of the fatty tissue. While healthy subcutaneous tissue tends to be of low echogenicity, in lipedema patients it is homogenously thickened, with higher echogenicity in color doppler images. In addition, highly echogenic septa were viewed in this region (33). Later studies also showed poorly echogenic regions, most likely due to an accumulation of interstitial fluid. In compression ultrasound, the more

Figure 7-12)
Madelung's disease manifested in the pelvic area of a 67-year-old woman (type III)

severe the edema, the greater the sensitivity to touch is and the more difficult the subcutaneous fatty tissue is to compress.

Indirect lymphangiography

With lipedema, indirect lymphangiography demonstrates a curved course of lymph collectors (Figure 7-13) and feathery and flame-like contrast agent deposits. This is similar in obese patients, however (17). With lipolymphedema, depending on the extent of impairment of the lymphatic system, dilated initial lymph vessels and a winding course and even hypoplasia of lymph collectors are also shown in the lymphangiogram. In comparison, the sack-shaped or fusiform microaneurysms of the initial lymph vessels revealed by fluorescence microlymphography appear to be indicative of lipedema (34). Their role in pathogenesis is unclear. Table 7-6 presents the incidence of lymphangiographic symptoms in cases of lipedema and lipolymphedema.

Lymphoscintigraphic function tests can be used to diagnose potential lymph flow impairment. In cases of lipedema, often no or only slight lesions of the superficial lymph transport (12, 35) are observed. The pre-

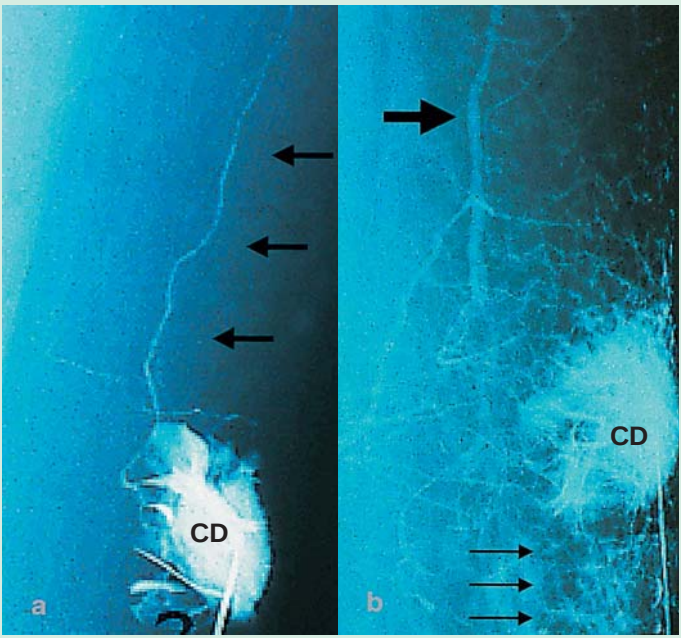

Figure 7-13)
Interstitial lymphangiography. a) Lipedema: contrast agent deposit (CD) with tipped contours, curved course of a superficial lymph collector (arrows). b) Lipolymphedema. Abnormal interstitial lymphangiogram. Reticular pattern caused by dilated initial lymph vessels. Dermal backflow (small arrows). Dilated lymph collector (thick arrows)

viously described hyperdynamic lymph transport in the early stages is attested to by elevated lymph node uptake levels (12, 19). Lymph transport decreases as the body ages (36). Especially in the case of the strain on the lymphatic system occurring for years manifested by lipolymphedema, the extent of lymph impairment can be measured by reduced transport times and decreased uptake levels in the regional lymph nodes (Figure 7-14). The use of these methods is especially important in stage I for early detection of potential secondary damage of the lymphatic system that is not clinically apparent.

	Sex	Onset	Localization	Symmetry	Fat increase	Pain	Edema	Feet involved	Responds to dieting	Other
Lipedema	female	mostly puberty	legs/arms	yes	yes	yes	yes	no	no	hematoma
Lipohypertrophy	female	mostly puberty	legs/arms	yes	yes	no	no	no	no	extensive distribution
Primary lymphedema	female male	frequently puberty	mostly legs	no	no	no	yes	yes	no	Stemmer+, abnormal lymphoscintigraphy
Phlebedema	female male	adulthood	legs	no	no	no	yes	no	no	abnormal venous function tests
Dercum's disease	mostly female	mostly menopause	legs	no	yes	yes	no	no	no	pain, muscle weakness, alcohol abuse, often depression
Madelung's disease	mostly male	adulthood	neck shoulder pelvis	yes	yes	yes	no	no	no	generally alcohol abuse, liver damage
Obesity	female male	all age groups	total body	yes	yes	no	no	no	yes	BMI >25

Table 7-5)
Differential diagnosis in lipedema

Indirect Lymphangiography		
	Lipedema (n: 54 extremities)	**Lipolymphedema** (n: 42 extremities)
Normal:	83.4%	33.3%
Abnormal:	16.6%	66.7%

Table 7-6)
Frequency of lymphangiographic symptoms in cases of lipedema or lipolymphedema

7.8 Course of Disease

In the majority of cases, symptoms worsen with age. However, the course cannot be predicted from case to case. The increase in the localized volume of fat, which sometimes occurs within a short period of time without any changes in eating habits (Figure 7-15), can be responsible. In addition, the long-term protein-rich edema can bring about increased fibrosis of the tissue with lipolymphedema, attested to by swelling of the forefoot. In addition, the Stemmer sign is positive (Figure 7-10). As already mentioned, this appears to occur more quickly and more frequently in obese patients. The reported incidence of lipolymphedema in specialized lymphedema hospitals varies widely, ranging from 4 % to 23% (4).

7.9 Therapy

The lack of awareness regarding lipedema continues to lead physicians to prescribe absurd and even counterproductive treatment, including dieting, training and toning the affected body parts and drugs. Because of the extreme symptoms, nearly all patients try some sort of dieting regimen. However, this only helps treat obesity and reduce the circumference of the trunk. Since the fat deposits specific to lipedema cannot be "starved off," dieting may lead to an even greater discrepancy between the trunk and the extremities. Burning calories through physical exercise also does not lead to the desired reduction of fat at the affected areas. Laxatives, to potentially reduce the absorption of nutrition, and diuretics, to alleviate edema symptoms, are not indicated.

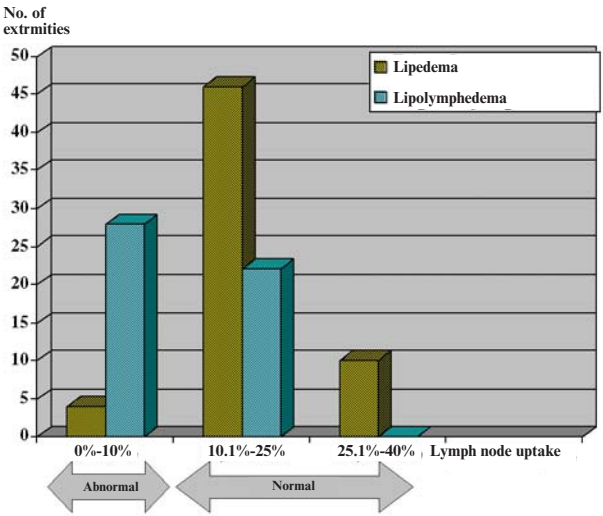

Figure 7-14)
Lymphoscintigraphic function test. Lymph node uptake, 120 minutes after injection of the radioactive tracer in patients with lipedema or lipolymphedema

Modern lipedema therapy involves two major components (37):
1. conservative treatment to eliminate edema and
2. surgical intervention to reduce fat.

Combining these two methods has proven very effective (38, 39, 40).

However, since these treatments do not have an effect on the increased capillary permeability and in turn on the tendency toward edema, lipedema generally requires long-term treatment, although to a lesser extent if surgery has been performed.

Conservative therapy

Conservative therapy aims to reduce the volume of interstitial fluid. In very early stages, orthostatic edema can be prevented by wearing compression garments. Combined decongestive therapy (CDT) is used for edema that is no longer spontaneously reversible.

The central component of CDT is manual lymph drainage (MLD). The varying degrees of pressure applied by massaging the superficial tis-

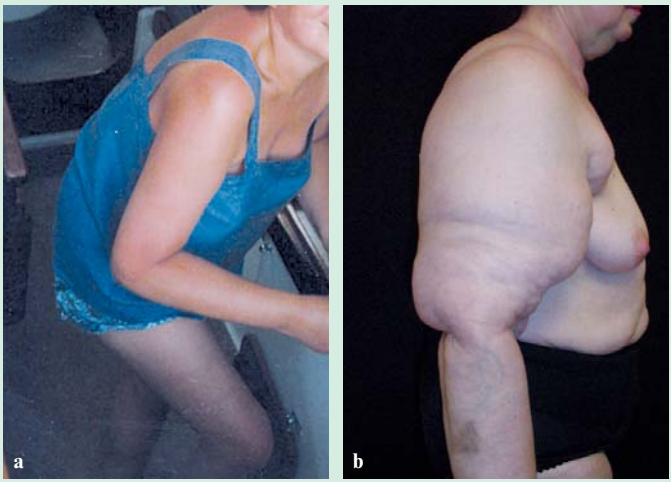

Figure 7-15)
a) Patient at age 51, no symptoms
b) Same patient at age 61, lipedema of the upper arms

sue increase the transport volume of the lymph collectors. Circular, pumping and scooping movements shift edematous fluid in a central direction toward the heart. The therapist starts MLD at the non-edematous trunk to create a suction effect and then continues to the edematous areas of the extremities (41). Compression with short-stretch bandages combined with physical therapy serves to reduce and prevent swelling.

During the edema reduction phase, therapy is performed daily. At the conclusion of this phase, flat knit pantyhose or compression stockings grade II or even grade III are fitted. In the subsequent preservation phase, Generally, MLD is required only once or twice a week. At home, intermittent pneumatic compression (IPC) with pneumatic multiple-chamber devices (12 chambers are ideal) may supplement this regimen (3, 18, 42). However, this method is not without its critics (43, 44). Therapy results must be regularly monitored. CDT is contraindicated with decompensated heart failure, acute bacterial infection and recent thromboses.

Figure 7-16)
37-year-old patient. a) Before liposuction, thighs; b) after removing 3,750 mL of fat from the hips and outer thighs

For lipedema patients, conservative therapy can bring about a reduction in circumference of approximately 10% and a decrease in volume of up to three liters per leg (45, 46), in turn alleviating the patient's hypersensitivity to pressure. If CDT is discontinued, edema will return. The procedure is basically the same for lipolymphedema patients. In severe cases, it may be advisable to begin treatment on an inpatient basis.

Surgical intervention

Surgical treatment for lipedema has become more common in recent years. Until the early 1990s, large-scale lipectomy or liposuction for lipedema was performed under general anesthesia with thick and sometime sharp suction cannulas without first infiltrating the subcutaneous tissue with liquid (dry technique). In some cases, postsurgical complications such as bleeding and lymph vessel damage of various degrees with persistent swelling occurred. For this reason, the procedure was generally not prescribed by lymphologists (13, 47).

The development of new anesthetic and surgical techniques has led to a turnaround in this attitude in recent years (48, 18, 21, 49). **Tumescent**

c) After removing 3,000 mL of fat from inner thighs and knees; d) after removing 2,800 mL of fat from the anterior thighs;

e) before liposuction, calves; f) after removing 3,050 mL of fat from the calves. A total of 12,600 mL of fat was removed within 14 months

local anesthesia (TLA) has eliminated the need for general anesthesia, along with the associated risks and disadvantages (50). With TLA, several liters of a 0.037% anesthetic solution are infiltrated into the subcutaneous tissue with a mixture of lidocaine and prilocaine (wet technique). During suctioning, a thin mixture of fat and solution is removed (51). The major advantages of this local anesthesia include the high level of safety for the patient, hydrodissection (detaching the fat lobes from the stabilized fibrous tissue, firming up of the tissue by reducing the shearing forces), hemostasis (vasoconstriction through increased tissue pressure and the proportion of adrenaline in the solution), long-term analgesic effects following surgery (due to the lipophilia of the local anesthetic), low rate of infection (due to the antibacterial effect and the washing-out effect of the TLA solution which continues to leak out of the incision sites after surgery.

The application of blunt **microcannulas** from 2 to 4 mm in diameter and the introduction of **power-assisted liposuction** have enabled surgeons to avoid damaging important structures, resulting in markedly reduced tissue trauma. The cannulas, which vibrate at high frequency (4,000 Hz) aspirate only the fat that is loosely in the connective tissue structure, sparing the surrounding structures such as nerves and vessels, bringing about better cosmetic results and accelerated healing (51).

Liposuction is now a standard surgical procedure. If performed under the conditions described above, it is very effective and of low risk (52). Complications described in the literature, including deaths, are generally the result of failure to observe internationally established guidelines and are sometimes due to the lack of basic medical skills on the part of the surgeon (53, 54). If possible, not more than 4 liters of pure fat should be removed per procedure. Thus, depending on the extent of the disease, between one and five operations at intervals of several months may be required (Figure 7-16a-f). In our own patients, for example, in a sequence of four procedures, up to 16 liters of fat could be aspirated and a reduction in the circumference of the thigh of up to 15 cm and in the lower leg of up to 9 cm could be achieved. On the one hand, liposuction can lead to a greatly improved physical appearance of the patient with harmonious body proportions. On the other hand, edema and sensitivity to pain of the tissue can be eliminated or at least considerably reduced (2, 55, 26, 56-58). The patients describe a previously unknown increase in quality of life (39); (see also Chapter 13.4). Postoperatively, there is generally an increased tendency for swelling; thus CDT should be initiated or continued within a few days of the procedure.

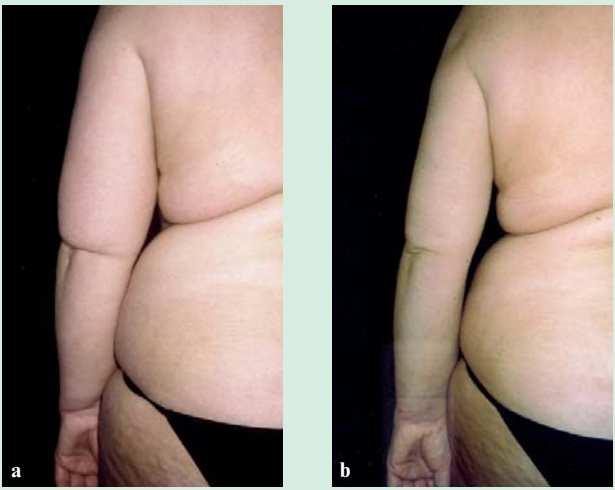

Figure 7-17)
56-year-old woman, a) before liposuction; b) four weeks after removing 1,550 mL of fat from the upper arms and forearms

The risk of lymph vessel damage caused by liposuction, with subsequent lymphedema described earlier in the literature, was not observed when using the new methods, either in experiments or clinically. Macroscopic anatomical studies after suction along the length of the extremities did not reveal any damage of the superficial lymph vessels (59, 60). Follow-up examinations of 19 patients over a period of eight years – now ten years (S. Rapprich, personal communication) – did not reveal increased swelling or a progression of the disease (56). Our own results with a follow-up period of up to three years confirmed the striking improvement in appearance and alleviation of symptoms described above. Currently, liposuction for lipedema is considered to be contraindicated in patients with accompanying lymphedema; whether this will change in the future remains open. In Germany, in contrast to conservative therapy, the costs for surgical treatment are in most cases not reimbursed by the public health insurers since liposuction is not part of their benefits' package.

Significance of therapeutic measures

In the past 10 years, advances in surgical technique have greatly impacted the way lipedema is treated. By combining surgical and conservative methods, today optimal results can be achieved. Liposuction alone does not suffice to eliminate lipedema. The severity of the tendency toward swelling, which is independent of the existing volume of fat, is decisive. While liposuction can reduce or normalize the amount of fat, it does not have any effect on the existing vessel permeability. The reduced tendency toward swelling experienced by patients after surgery is most likely the result of the diminished superfascial space. In some cases, liposuction leads to complete elimination of symptoms - regardless of the stage of the disease. For many patients, however, a tendency toward edema persists, thus requiring the continuation of basic treatment with CDT, albeit much less frequently. In some cases, compression can temporarily be dispensed with completely, while in other cases, patients can switch to garments with a lower grade of compression. Close cooperation between the liposuction surgeon who is familiar with lipedema and trained lymphedema therapists is essential.

►Teaching points

- **Lipedema is characterized by symmetrical fat accumulation primarily in the lower half of the body with orthostatic edema, tenderness and hypersensitivity to pressure.**
- **Lipedema is frequently confused with primary lymphedema. It can be diagnosed early on the basis of clinical criteria. In later stages, secondary lymphedema also may develop. This occurs more frequently in obese patients.**
- **Therapy includes conservative and surgical treatment. Liposuction with tumescent local anesthesia (TLA) should be performed in the early stages of the disease. In some cases, liposuction can bring about complete healing. With pronounced edema and lipolymphedema, combined decongestive therapy (CDT) should be carried out for life. The combination of liposuction and CDT can lead to considerable improvement in quality of life.**

7.10 References

1. Allen EV, Hines EA. Lipedema of the legs: A syndrome characterized by fat legs and orthostatic edema. Proc Staff Mayo Clin 1940; 15: 184-187.

2. Schmeller W, Meier-Vollrath I. Erfolgreiche operative Therapie des Lipödems mittels Liposuktion. Phlebologie 2004; 33: 23-29.

3. Herpertz U. Krankheitsspektrum des Lipödems an einer Lymphologischen Fachklinik - Erscheinungsformen, Mischbilder und Behandlungsmöglichkeiten. vasomed 1997; 5: 301-307.

4. Meier-Vollrath I, Schneider W, Schmeller W. Lipödem: Verbesserte Lebensqualität durch Therapiekombination. Dtsch Ärzteblatt 2005; 102: A1061-1067.

5. Beninson J. Lipedema - The non-lymphatic masquerader. Angiology 1984; 35: 506-510.

6. Witte MH, Witte CL. Massive obesity simulating lymphedema. N Engl J Med 1992; 327: 1927.

7. Rudkin GH, Miller TA. Lipedema: A clinical entity distinct from lymphedema. Plast Reconstr Surg 1994; 94: 841-847.

8. Koss T, Lanatra N, Stiller MJ Grossmann ME. An unusual combination: Lipedema with Myiasis. J Am Acad Dermatol 2004; 50: 969-972.

9. Strößenreuther RHK (ed.). Lipödem und andere Erkrankungen des Fettgewebes. Viavital, Cologne 2001.

10. Wold LE, Hines EA, Allen EV. Lipedema of the legs: A syndrome characterized by fat legs and edema. Ann Intern Med 1949; 34: 1243-1250.

11. Herpertz U. Entstehungszeitpunkt von Lipödemen. LymphForsch 2004; 8 (2): 79-81.

12. Harwood CA, Bull RH, Evans J, Mortimer PS. Lymphatic and venous function in lipedema. Br J Dermatol 1996; 134: 1-6.

13. Földi E, Földi M. Das Lipödem. In: Földi M, Földi E, Kubik S (eds.) Lehrbuch der Lymphologie für Mediziner, Masseure und Physiotherapeuten. 6th edition, Elsevier, Urban&Fischer, Munich 2005, 443-453.

14. Kaiserling E. Morphologische Befunde beim Lymphödem. Lipödem, Lipolymphödem. In: Földi M, Földi E, Kubik S (eds.) Lehrbuch der

Lymphologie für Mediziner, Masseure und Physiotherapeuten. 6th edition, Elsevier, Urban&Fischer, Munich 2005, 374-378.

15. Strößenreuther RHK. Laser-Doppler-Flowmetrie. In: Strößenreuther RHK (ed.). Lipödem und andere Erkrankungen des Fettgewebes. Viavital, Cologne 2001, 79-86.

16. Fasshauer M, Klein J, Blüher M, Paschke R. Adipokine: Mögliches Bindeglied zwischen Insulinresistenz und Adipositas. Dtsch Ärzteblatt 2004; 101: A 3491-3495.

17. Weissleder H, Brauer WJ. Radiologische Diagnostik beim Lipödem-Syndrom. LymphForsch 1997; 1 (1): 26-30.

18. Marshall M, Breu FX. Das Lipödem - ein wenig beachtetes Krankheitsbild. vasomed 2002; 6: 254-257.

19. Brauer WJ, Weissleder H. Methodik und Ergebnisse der Funktionslymphszintigraphie: Erfahrungen bei 924 Patienten. Phlebologie 2002; 31: 118-125.

20. Marsch WC. Ist das Lipödem ein lymphologisches Kranheitsbild? J Lymphologie 2001; 1: 22-24.

21. Herpertz U. Ödeme und Lymphdrainage. Diagnose und Therapie von Ödemkrankheiten. 2nd edition. Schattauer, Stuttgart New York 2004, 168-181.

22. Curri SB. Zellulitis oder Pannikulopathia oedemato-fibrosclerotica (Liposklerose): Ein nosologisches Problem. In: Ges. ML Vodder, ed. Schriftenreihe "Manuelle Lymphdrainage nach Dr. Vodder" - Referate 1984, K.F. Haug, Heidelberg: 1984: 9-42.

23. Tronnier M, Schmeller W, Wolff HH. Morphological changes in lipodermatosclerosis and venous ulcers: Light microscopy, immunohistochemistry and electron microscopy. Phlebology 1994; 9: 48-54.

24. Greer KE. Lipedema of the legs. Cutis 1974; 14: 98-100.

25. Schuchhardt C. Das "Lipödem-Syndrom" - neue Antworten auf alte Fragen? LymphForsch 2001; 5: 68-70.

26. Meier-Vollrath I, Schmeller W. Lipödem - aktueller Stand, neue Perspektiven. JDDG 2004; 2: 181-186.

27. Monnin-Delhom ED, Gallix BP, Achard C, Bruel JM, Janbon C. High resolution unenhanced computed tomography in patients with swollen legs. Lymphology 2002; 35: 121-128.

28. DeFranzo AJ, Hall JH, Herring SM. Adiposis dolorosa (Dercum´s disease): Liposuction is an effective form of treatment. Plast Reconstr Surg 1990; 85: 289-292.

29. Ruzicka T, Vieluf D, Landthaler M, Braun-Falco O. Benign symmetric lipomatosis Launois-Bensaude. Report of ten cases and review of the literature. J Am Acad Dermatol 1987; 17: 663-674.

30. Hebebrand J, Dabrock P, Lingenfelder M, Mand E, Rief W, Voit W. Ist Adipositas eine Krankheit? Dtsch Ärzteblatt 2004; 101: A 2468-2474.

31. Dimakakos PB, Stefanopoulos T, Antoniades P, Antoniou A, Gouliamos A, Rizos D. MRI and ultrasonographic findings in the investigation of lymphoedema and lipedema. Int Surg 1997; 82: 411-416.

32. Werner GT, Rodiek SO. Value of nuclear magnetic resonance tomography in leg edema of unknown origin. Lymphology 1993; 17: 2-5.

33. Breu FX, Marshall M. Neue Ergebnisse der duplexsonographischen Diagnostik des Lip- und Lymphödems. Kompressionssonographie mit einer neuen 13-MHz-Linearsonde. Phlebologie 2000; 29: 124-128.

34. Amman-Vesti BR, Fanzeck UK, Bollinger A. Microlymphatic aneurysms in patients with lipedema. Lymphology 2001; 34: 170-175.

35. van Geest AJ, Esten SCAM, Cambier JPRA, Gielen EGJ, Kessels A, Neuman HAM, van Kroonenburgh MJPG. Lymphatic disturbances in lipoedema. Phlebologie 2003; 32: 138-142.

36. Brauer WJ, Brauer VS. Altersabhängigkeit des Lymphtransportes beim Lipödem und Lipolymphödem. LymphForsch 2005; 9: 6-9.

37. Wienert V, Földi E, Schmeller W, Rabe E. Leitlinie: Lipödem der Beine. Phlebologie 2005; 34: 38-40.

38. Schmeller W, Meier-Vollrath I. Moderne Therapie des Lipödems: Kombination von konservativen und operativen Maßnahmen. LymphForsch 2004; 8 (1): 22-26.

39. Schmeller W, Meier-Vollrath I. Lipödem: Ein Update. LymphForsch 2005; 9 (1): 10-20.

40. Schmeller W, Meier-Vollrath I. Tumescent liposuction: A new and successful therapy for lipedema. J Cutan Med Surg 2006; 10:7-10.

41. Schneider W, Herpertz U. Indikation und Kontraindikation der physikalischen Ödemtherapie. Orthopädie-Technik 1996; 3: 185-191.

42. Rabe E (ed.). Apparative intermittierende Kompressionstherapie (AIK). Viavital, Cologne 2003.

43. Gültig O. Erfolg und Misserfolg bei Einsatz der Apparativen Intermittierenden Kompressionstherapie (AIK) – Ergebnisse einer breit angelegten Fragebogenaktion bei Patienten mit chronischem Lymphödem der Extremitäten. LymphForsch 2004; 8 (2): 91-92.

44. Weissleder H. AIK und Lymphödem - Glaube und Wirklichkeit. LymphForsch 2004; 8 (2): 93-95.

45. Deri G, Weissleder H. Vergleichende prä- und posttherapeutische Volumenmessungen in Beinsegmenten beim Lipödem. LymphForsch 1997; 1 (1): 35-37.

46. Brenke R, Siems W, Obendorfer H. Klinische Erfolge der Komplexen Physikalischen Entstauungstherapie beim Lipödem. In: Strößenreuther RHK. Lipödem und andere Erkrankungen des Fettgewebes. Viavital, Cologne 2001, 215-217.

47. Földi M. Lymphödem, Lipödem, chronisch venöse Insuffizienz und Kombinationsformen. Phlebol Proktol 1990; 19: 1-9.

48. Wienert V. Diagnose und Therapie des Lipödems. Der Deutsche Dermatologe 2001; 9: 614-616.

49. Diehm C. Lipödem und Lymphödem. Vernachlässigte Differentialdiagnose des dicken Beines. Cardiovasc 2002; 2: 1.

50. Klein JA. The tumescent technique. Anesthesia and modified liposuction technique. Dermatol Clin 1990; 8: 425-437.

51. Schmeller W, Meier-Vollrath I. Zum aktuellen Stand der Liposuktion. Der Deutsche Dermatologe 2002; 9: 590-594.

52. Sattler G, Sommer B, Hanke CW. Leitlinien zur Liposuktion. In: Sattler G, Sommer B, Hanke CW (eds.). Lehrbuch der Liposuktion. Thieme, Stuttgart 2003, 217-222.

53. Lehnert M, Homann HH, Druecke D, Palka P, Steinau HU. Liposuktion - kein Problem? Majorkomplikationen und Todesfälle im deutschen Raum zwischen 1998 und 2002. LymphForsch 2004; 8 (2): 74-78.

54. Schmeller W, Meier-Vollrath I. Kommentar zum Artikel: Komplikationen nach Liposuktion von Berroth R, Speichermann N, Liebau G. Intensivmed 2004; 41: 64-66.

55. Chen S, Hsu SD, Chen TM, Wang HJ. Painful fat syndrome in a male patient. Br J Plast Surg 2004; 57: 282-286

56. Rapprich S, Loehnert M, Hagedorn M. Therapy of lipoedema syndrome by liposuction under tumescent local anaesthesia. Ann Dermatol Venereol 2002; 129: 1S711.

57. Sattler G. Liposuction in lipoedema. Ann Dermatol Venereol 2002; 129: 1S103.

58. Sattler G, Hasche E, Rapprich S. Neue operative Behandlungsmöglichkeiten bei benignen Fettgewebserkrankungen. Zeitschr Hautkrh 1997; 72: 579-582.

59. Frick A, Hoffmann JN, Baumeister RGH, Putz R. Liposuction technique and lymphatic lesions in lower legs: Anatomic study to reduce risks. Plast Reconstr Surg 1999; 103: 1868-1873.

60. Hoffmann JN, Fertmann JP, Baumeister RG, Putz R, Frick A. Tumescent and dry liposuction of lower extremities: Differences in lymph vessel injury. Plast Reconstr Surg 2004; 113: 718-724; discussion 725-726.