

Manuel E. Cornely and Matthias Gensior

Abstract

In many respects, lipoedema of the arms and legs is still an un-researched disease within the lymphatic spectrum. In terms of clinical aspects, symmetrical fat distribution in the arms and legs and pathognomonic tenderness are frequently evident in female family members. However, 75 years after the first descriptions by Allen and Hines, we still lack pathological evidence that goes beyond the theses of Marsch and Brauer. Neither do we know anything about the hormonal influence on hyperplastic fatty tissue nor the causes of obviously increased lymph formation in the fatty tissue of the lipohyperplasia dolorosa. We know more about the effects of complex decongestive therapies, which have been used since the 1960s. Since 1997, the clinical picture can be cured surgically. The success rate with long-term observation (15 years) is 97 %.

M.E. Cornely, M.D. (✉)
CG Lympa, Bachemer Str. 31,
50931 Köln, Germany

Private Practice, Kaiserswerther Str. 296,
Düsseldorf 40474, Germany
e-mail: post@cg-lympa.de;
post@hautarzt-duesseldorf.de

M. Gensior, M.D.
Department of Plastic and Esthetic surgery,
CG Lympa GmbH, Bachemerstraße 31,
50931 Köln, Germany
e-mail: m.b.gensior@online.de; post@cg-lympa.de

79.1 Introduction

Seventeen years after a paradigm shift in the treatment of lipoedema, the operative method (carried out as “lymphological liposculpture”) has gained increasingly more acceptance. An estimated incidence of the disease lipohyperplasia dolorosa [1] with 11 % of the female population would represent more than four million women affected by this disease in the legs and arms. Well over 80 % of patients are affected on all four extremities. Despite this significant frequency in the population, there is not enough knowledge about the lymphological disease pattern of lipoedema, including in specialist circles.

79.2 Clinical Picture

In contrast to previous publications, it must be noted that the frequency of the lipoedema on the arms is surely within the 80–90 % range (Fig. 79.1).

Examination of the arms is not usually performed, as patients are primarily examined for venous diseases due to pain in the leg and suspected venous disorders. As a result, they see a phlebologist and the arms are not examined. Lipoedema patients with suspected lipoedema of the legs must always be examined on the arms as well, in order to establish the full clinical picture (Fig. 79.2).

Although first described by Allen and Hines [2] as far back as the 1940s, the distribution of the fatty tissue is often not taken seriously as a clinical diagnosis – the cardinal symptoms are deliberately overlooked and patients are given the stigma of obesity. However, medical history regularly shows that experiments with diet and sport fail to reduce the disproportional enlargement of the fatty tissue, especially that of the extremities. This urges patients to make futile attempts that offer everything from ineffective diets to high-performance sports. They are still unsuccessful [2].

Since patients usually seek medical help because of their leg problems, the lipoedema should always be taken into consideration with a differential diagnostic whenever leg complaints occur symmetrically: the feet are slim and unaffected from the ankle joint onward, and there is no pitting or even a Stemmer's sign on the feet

or pitting of the ankles in the area of the aching legs. When this becomes clear to the physician, it is necessary to look at the patient's whole body (nearly unclothed) and pay special attention to the arms. As a rule, the symmetric distribution of the fatty tissue is also pronounced from the wrists to the shoulder, as well as from the ankle to the groin. The simple medical history question as to whether fat distribution on the trunk can be reduced via diet will normally be affirmed; a simple pressure test (performed as gentle, paradoxical pinching on the outer sides of the legs and arms) will be confirmed as painful. If the same pressure test is entirely normal during manual examination of the abdomen, the painfulness of the fat on the extremities compared to the painless fat on the trunk is clinically evident. Beyond these clinical examinations and a correctly performed history, further diagnostic methods are not helpful any more, even under medical guideline aspects [3]. A sonography of the fatty tissue may outline the difference between lymphoedema and lipoedema, but the decision regarding therapy will not be amended.

The pathophysiological moment of high-volume transport insufficiency, as researched by Brauer [4] and set out as a model in the Marsch Thesis [5], is the goal of conventional therapeutic approaches. Complex decongestive therapy is responsible for removing the imbalance between the production and removal rate caused by increased lymph formation, in totally normal lymph vessels in the arms and legs.



Fig. 79.1 Lipoedema of the arms



Fig. 79.2 (a) Lipoedema stage 1. (b) Lipoedema stage 2. (c) Lipoedema stage 3

In lipoedema, the lymph vessels are intact but fail in their transport capacity in the long run – this constitutes a dynamic deterioration of the clinical picture. It is important to interrupt this dynamic, which is why early therapies are indicated, whether with conventional or curative surgical methods [6].

79.3 Pathophysiology

The fat on lipoedema patients' extremities is of a particular quality, as it is a type of tissue that does not increase in volume, but rather in the number of cells. Due to the pain of the shift, the lipoedema is to be called lipohyperplasia dolorosa and not lipohypertrophy [7]. The diagnosis "lipohyperplasia dolorosa" requires obligatory proof of:

1. The disruption to symmetrical fat distribution on the extremities in relation to the torso in the case of asymptomatic hands and feet
2. The dietary and exercise-related resistance of fat due to genetic disproportion
3. Painfulness
4. Increased vascular fragility

In almost no case will the physician find pitting in the feet area or a positive Kaposi-Stemmer's sign in the toe region. The absence of this clinical symptom is an undoubted indication that a lipo-lymphoedema does not exist. It is amazing, however, to see how often this

referral-diagnosis is put forward by patients. The aim of these therapeutic options is to treat the high-volume transport insufficiency of the lipohyperplasia dolorosa, as this form of insufficiency in the lymphatic vessels of the extremities leads to a backflow of lymph between the fat cells, and therefore to increased pressure in the tissue of completely sound but disproportional lymphatic vessels that transport well (Fig. 79.3).

79.4 Extragenital Impact of Sex Hormones

Estrogen promotes proliferation, dilates the vessels, and increases extracellular fluid. The formation of edemas is encouraged by cycle-dependent hormone shifts and during stages of estrogen dominance (puberty, climax). Progesterone (corpus luteum hormone) promotes tissue differentiation and increases energy metabolism (in order to supply energy in pregnancy).

Together, these two hormones encourage the development of the typical gynoid fat distribution (particularly buttocks and thighs): for the first time in puberty and particularly during pregnancy [8].

The physiological effects of female sex hormones thus indicate causes for the clinical observation of the first occurrence of lipoedema in puberty, a "maturation" of preclinical lipoedema in pregnancy and also increased health problems during the premenopause phase (estrogen dominance).

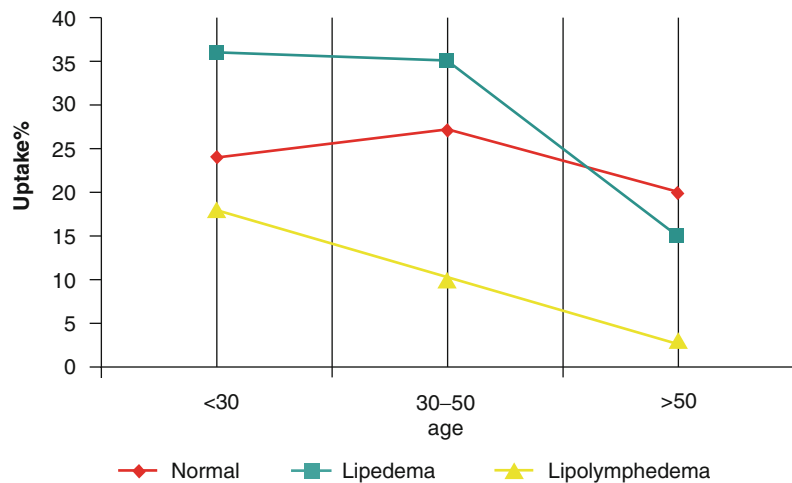


Fig. 79.3 Age-adapted uptake rate by Brauer

These indications of pathophysiological links were brilliantly worked on at the end of last century, resulting in the development of therapeutic options beyond complex decongestive therapy, i.e., a surgical treatment that prioritizes the irreversible removal of fatty tissue from a medical indication [9–11].

79.5 Therapeutic Options

The task was to normalize the imbalance ratio of lymph production and removal, to interrupt and halt the dynamic deterioration of the disease, and to achieve a healing procedure (namely surgical). Although it is not fully understood how lymph formation is disturbed, it is undoubtedly clinically evident that healing can be achieved by means of surgery – operations which have been successfully performed for 17 years.

Following the introduction of tumescence local anesthesia by Klein in the 1980s and liposuction in Germany by Sattler in the early 1990s for cosmetic indications, a secure set of tools became available [12]. Using such tools, the first tentative attempts in applying medically indicated liposuction on lipohyperplasia dolorosa began in Düsseldorf in 1997.

79.6 Lymphological Liposculpture

Despite clear clinical findings, the “pioneer patients” were all subjected to preoperative

indirect lymphangiography and dynamic functional lymphoscintigraphy on the arms and legs. This was to gain a clear preoperative picture of lymph flow and to demonstrate that the medically indicated removal of fatty tissue, i.e., lymphological liposculpture on lipohyperplasia dolorosa would have no adverse side effects on the lymphatic vessels. None of these patients developed a lymphoedema resulting from the treatment. Long-term studies were completed after 7 years in 2004 (Fig. 79.4).

Once again, the operated patients had to undergo a clinical examination in which no lymphoedemas were detected. In addition, they also underwent a dynamic functional lymphoscintigraphy and an indirect lymphangiography [13]. The clinical aspect was proven: none of the operated patients had developed a lymphoedema caused by the operation. These findings were histologically supported in studies by Rapprich [14] and Schmeller [15] as well as negative evidence of lymphocytes in the extracted tissue.

The efficiency of the method was also substantiated numerous times by other surgeons [6, 14, 15]. From a clinical point of view, it is absolutely clear that lymphological liposculpture procedures – if applied on lipohyperplasia dolorosa and if the fatty tissue of the extremities is fully removed (and this is *conditio sine qua non*) – will result in healing of the clinical picture. Moreover, the surgical procedure requires mandatory postoperative management in order to achieve the excellent results. “Lymphological liposculpture” is not a plastic surgery procedure for removing fatty tissue;

Lipödem | Studie 2004

- No manual lymphatic drainage approx.: 95 %
- No compression, no skin-protection
- No more pain approx.: 100 %
After 1–2 weeks post surgery

Lipödem | Studie 2004

Diagnosis Lipedema & combination forms	762	100 %
Conservative	622	81.6 %
Lymphological liposculpture	140	18.3 %

Fig. 79.4 Lipoedema study 2004

it means consistent operative, conventional treatment of a lymphological disease [10, 16].

In 2004, i.e., 7 years after the paradigm shift, the quality of the procedure was researched for the first time in a large clinical monocentric study using 140 patients [17]. None of the patients had suffered from a postoperative lymphoedema. Five percent of the operated patients occasionally underwent complex decongestive therapy. The healing rate was therefore 95 %.

79.7 Cologne Lipoedema Study 2012

Fifteen years after the introduction of the lymphological liposculpture method, the authors reviewed the data on a significantly greater number of patients in order to establish the quality of the method and its effect on healing. Throughout the entire Cologne Lipoedema Study in 2012, 3,140 patients suffering from a lipoedema were examined at the surgery. Nineteen percent of these patients were operated on. The indication for surgery had been set as follow: all patients who did not exhibit internistic (clinical) contraindications and weighed a maximum of 125 kg. Since the surgical treatment is private and must be paid for by the patient, a further limitation arose, namely, those willing and able to undergo this operation. In addition, a number of patients were familiar with conventional therapy and did not want to go without it.

Of the total number of lipoedema patients, 19 % were operated on, i.e., 592 patients. These patients were then asked to answer a questionnaire. The questions were as follows (Fig. 79.5):

1. How long had you been suffering from symptoms until the correct diagnosis was made?
2. For how long did you experience postoperative discomfort?
3. Do you still have pain?
4. Do you still undergo complex decongestive therapy?
5. What do you think of the cosmetic result?
6. How would you evaluate your quality of life?
7. Would you have the surgery again?
8. Would you recommend the operation?

The response rate was 57 % with 292 patients.

Since all patients had to undergo several operations due to the amount of fat, among other things, the total number of operations from 1997 until the beginning of the study in May 2012 for the indication lipohyperplasia dolorosa amounted to 1,654 procedures (Fig. 79.6). From medical records, we determined regular accumulation of stage II at an average age of 23–45 – the youngest patient was 15 and the oldest 83 years of age.

The amount of removed fat varied between 1,500 and 8,000 mL per operation; in the most complex case, we removed 26.5 l of fat in several steps. This magnitude underlines the fact that the indication for this operation was by no means cosmetic [6, 10]. For nearly a decade, preoperative preparation has been done not only by a surgeon but also by an anesthesiologist. The surgery itself always takes place in a team with the anesthesiologist: tumescence local anesthesia and accompanying analgesation. After removing such huge amounts of tissue, it is mandatory to monitor patients for 24 h in inpatient conditions. The cutting-suture time takes up to 320 min as a rule.

The question of the duration of symptoms until correct diagnosis yielded a spread of 5 months to 15 years (Fig. 79.7). Postoperative symptoms were indicated on average between 2 and 6 months (Fig. 79.8). On the question of whether complaints still existed at the time of the study, 83 % of the patients responded that they no longer had symptoms (Fig. 79.9).

The authors were particularly interested in the continuation of complex decongestive therapy. Findings showed that 3 % of patients, i.e., nine of the operated patients, still underwent decongestive therapy from time to time. Ninety-seven percent of patients reported requiring no further complex decongestive therapy.

While satisfaction with the cosmetic result was 89 %, considering stage III lipohyperplasia dolorosa, 95 % of patients stated that their quality of life had improved considerably; the recommendation rate for the operation was also 95 % (Figs. 79.2 and 79.10).

In addition to the exact operation procedure, the lymphological liposculpture method also includes a crucial postoperative setting. This consists of a

FRAGEBOGEN

zur Lebensqualität nach operativer Therapie des Lipödems

Sehr geehrte Patientin,

aufgrund Ihrer Diagnose Lipohyperplasia dolorosa (Lipödem) wurde bei Ihnen als operative Therapie eine Lymphologische Liposkulptur durchgeführt. Wir würden uns freuen, wenn Sie sich ein paar Minuten Zeit nehmen, um uns von Ihren Erfahrungen zu berichten.

▶ Wie lange hatten Sie bereits Beschwerden bis die richtige Diagnose gestellt wurde? 1 Jahr 2-5 Jahre > 5 Jahre

▶ Wie lange hatten Sie noch OP-bedingte Beschwerden? 1 Monat 2-6 Monate > 6 Monate

▶ Haben Sie heute noch Beschwerden? Ja Nein

▶ Wenn ja, welche Beschwerden haben Sie? Druckschmerzen Hämatomneigung (Blaue Flecken)

▶ Führen Sie noch komplexe Entstauungstherapie durch? Ja Nein

▶ Wenn ja, welche Maßnahmen? Manuelle Lymphdrainage
Anzahl pro Woche Kompressionsbestrumpfung

▶ Wie empfinden Sie das kosmetische Ergebnis? 😊 😐 ☹️

▶ Beurteilung der Verbesserung der Lebensqualität 😊 😐 ☹️
deutliche Verbesserung Verbesserung keine Verbesserung

▶ Würden Sie die Operation heute wieder durchführen lassen? Ja Nein

▶ Würden Sie die Operation weiter empfehlen? Ja Nein

▶ Sonstige Bemerkungen zur Veränderung Ihrer Lebensqualität nach der Operation:

Dr. med. **Manuel E. Cornely**
Prof. Honorario (Univ. Puebla)

Kaiserswerther Str. 296 | 40474 Düsseldorf | Fax: 0211. 35 76 01 | Tel: 0211.17 111 71
www.hautarzt-duesseldorf.de | post@hautarzt-duesseldorf.de

Fig. 79.5 Cologne Lipoedema Study 2012: questionnaire

Fig. 79.6 Cologne Lipoedema Study 2012: response rate

- 592 patients were contacted
- 299 patients responded

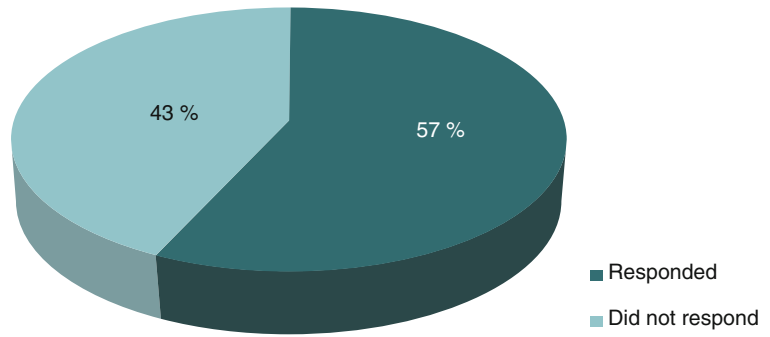


Fig. 79.7 Cologne Lipoedema Study 2012: suffering from symptoms before diagnosis

- How long had patients been suffering from symptoms before diagnosis?

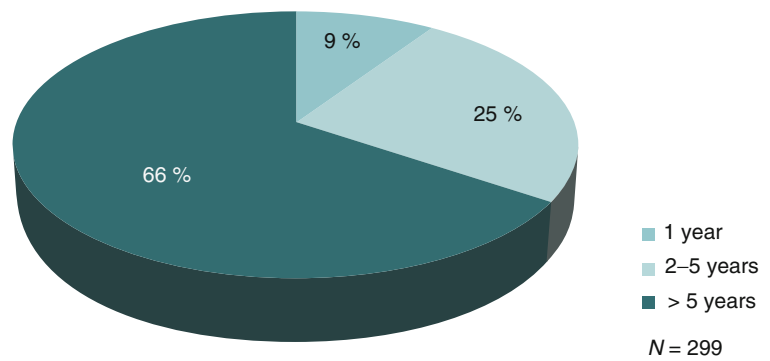
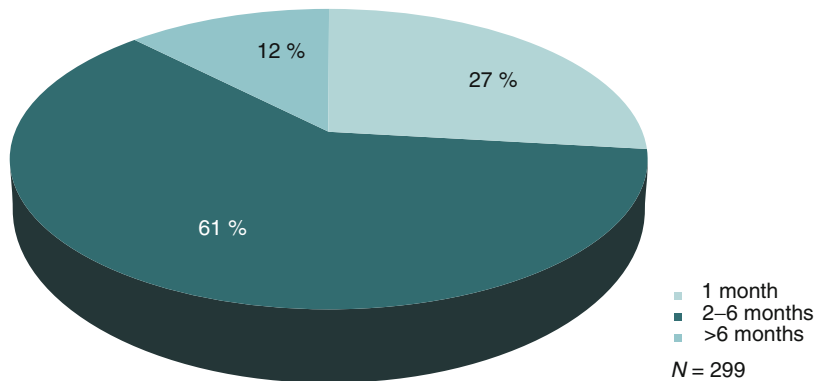


Fig. 79.8 Cologne Lipoedema Study 2012: length of postoperative discomfort

- For how long did patients experience postoperative discomfort?



1-week intensive outpatient follow-up treatment at the patient's residence: 1 h of lymphatic drainage per day and wearing the appropriate compression garments 24 h a day. In the second, third, and fourth

week of the follow-up treatment, the frequency is halved. The first week is accompanied by antibiotics as well as a heparinization in order to avoid side effects. Symptoms occurred in less than 0.2 % of

Fig. 79.9 Cologne Lipoedema Study 2012: postoperative discomfort

➤ How many patients still experience discomfort (bruising, etc.)?

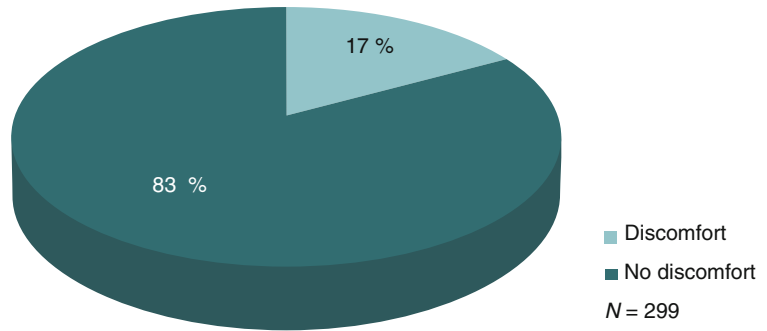


Fig. 79.10 Cologne Lipoedema Study 2012: satisfaction cosmetic result

➤ How satisfied are patients with the cosmetic result?

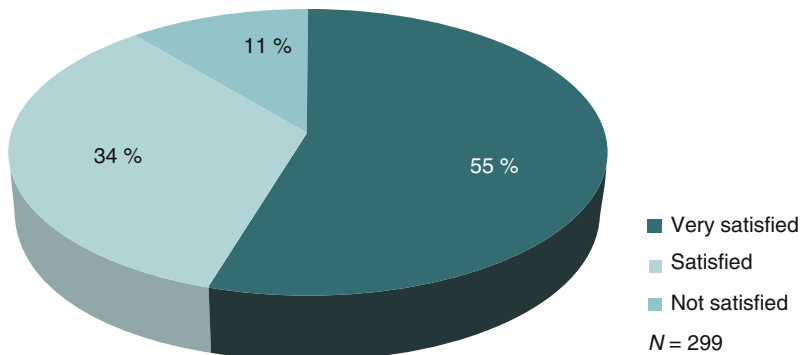
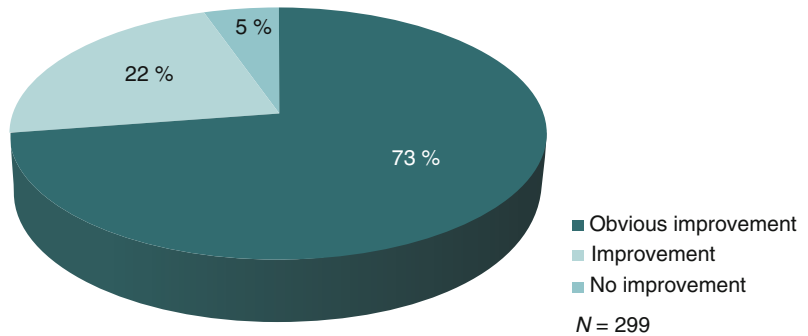


Fig. 79.11 Cologne Lipoedema Study 2012: improvement of life quality

➤ How do patients evaluate improvement to their quality of life?



patients: panniculitis, hematoma, muscle aches, wound infections, erysipelas, thrombosis, and pulmonary embolism. Despite the large amount of tissue to be operated on, the authors believe that the operation must be carried out as outpatient surgery, but carefully planned, due to the low side effect rate (Fig. 79.11).

Since 97 % of the patients did not undergo further complex decongestive therapy, the authors of the 2012 Cologne Lipoedema Study allowed themselves to continue talking of “healing” the disease lipohyperplasia dolorosa by means of lymphological liposculpture (Fig. 79.12).

Fig. 79.12 Cologne Lipoedema Study 2012: surgery recommendation

➤ How many patients would have the surgery again and/or recommend it?

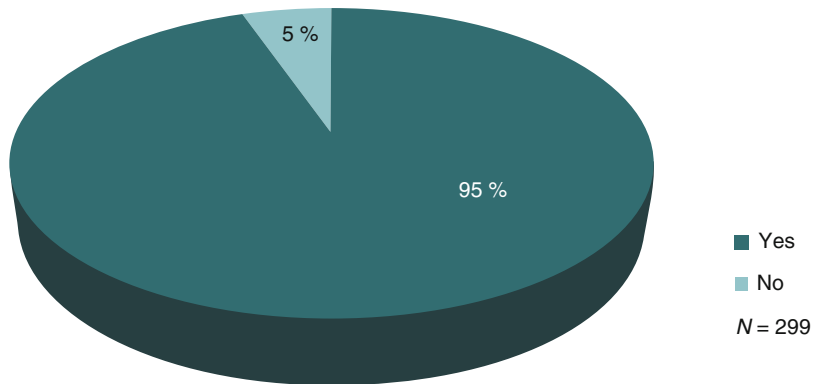


Fig. 79.13 Cologne Lipoedema Study 2012: findings at a glance

➤ Findings at a glance

Lipoedema patients examined since 1997	3140
Lipoedema surgeries since 1997	1645 surgeries on 592 patients
Response rate	(299 patients) 57 %
How many patients are still undergoing CDT?	3 %
Satisfaction with cosmetic result	89 %
Evaluation of improvement to quality of life	95 %
How many would have the surgery again/recommend it?	95 %

It is reported, encouragingly, that colleagues Schmeller (in Lübeck) and Rapprich (in Darmstadt) have had similarly good results in the surgical treatment of lipoedema. Professor Schmeller [15] reported on 114 patients and Rapprich [14] on 27 cases. Regardless of the surgical procedure's excellent results on lipoedema, it should be noted that it is not advisable to try to cure the lymphatic disorder "lipohyperplasia dolorosa" by means of lymph surgery alone. Furthermore, up to 80 % of the patients at the CG Lymphe center in Cologne are treated successfully using primarily nonoperative procedures, even though the curative treatment is increasingly becoming more accepted (Fig. 79.13).

Conclusions

Surgical treatment on patients with a lymphatic illness is a special one. It should certainly not be confused with plastic surgery procedures.

Both preoperative and above all postoperative patient care require qualified, conventional lymphological procedures. Patients with lipohyperplasia dolorosa should be examined carefully on the legs and arms; they should be offered both conventional and surgical therapy.

Healing rates following surgical procedures are established at 97 % in the largest sample size study with 592 patients and a follow-up term of 15 years.

References

1. Cornely ME. Dicker durch Fett oder Wasser – Lipohyperplasia dolorosa vs. lymphödem. *Hautarzt*. 2010;61(10):873–9.
2. Allen EV, Hines EA. Lipedema of the legs: a syndrome characterized by fat legs and orthostatic edema. *Proc Staff Meet Mayo Clin*. 1940;15:184–7.

3. Wienert V, Földi E, Jünger M, Partsch H, Rabe E, Rapprich S, Schmeller W, Stenger D, Stücker M, Waldermann F. Lipödem. Leitlinien der Deutschen Gesellschaft für Phlebologie. *Phlebologie*. 2009;38:164–7.
4. Brauer WJ. Altersbezogene Funktionslymphszintigraphie beim Lipödem und Lipolymphödem. *LymphForsch*. 2000;2:74–7.
5. Marsch WC. Ist das Lipödem ein lymphologisches Krankheitsbild? *J Lymphologie*. 2001;1:22–4.
6. Meier-Vollrath I, Schneider W, Schmeller W. Lipödem – neue Therapiemöglichkeiten für ein oft übersehener Krankheitsbild. *Dermatol Prakt Dermatol*. 2007;13:297–304.
7. Cornely ME. Lipödem und lymphödem. In: Plewig G, Prinz J, editors. *Fortschritte der praktischen Dermatologie und Venerologie 2002*. Berlin: Springer; 2003. p. 255–63.
8. von Runnebaum B, Rabe T. Gynäkologische Endokrinologie. Grundlagen, Physiologie, Pathologie, Prophylaxe, Diagnostik, Therapie. Berlin: Springer; 2012. p. 5.
9. Cornely ME. Liposuktion (liposculpture). In: Weissleder H, Schuchhardt C, editors. *Erkrankungen des Lymphgefäßsystems*. Köln: Viavital; 2000. p. 384–97.
10. Cornely ME. Lipedema and lymphatic edema. In: Shiffman MA, Di Guiseppa A, editors. *Liposuction: principles and practice*. Berlin: Springer; 2006. p. 547–55.
11. Sattler G, Hasche E, Rapprich S. Neue operative Behandlungsmöglichkeiten bei benignen Fettgewebskrankungen. *Z Hautkrh*. 1997;72:579–82.
12. Habbema L. Safety of liposuction using exclusively tumescent local anesthesia in 3,240 consecutive cases. *Dermatol Surg*. 2009;35(11):1728–35.
13. Bender H, Cornely ME, Pleiß C, Risse JH. Lymphszintigraphie beim Lipödem. Einfluss einer Liposuktion. *Vasomed*. 2007;19:60–2.
14. Rapprich S, Dingler A, Podda M. Liposuktion ist eine wirksame Therapie beim Lipödem – Ergebnisse einer Untersuchung mit 25 Patientinnen. *J Dtsch Dermatol Ges*. 2011;9:33–41.
15. Schmeller W, Meier-Vollrath I. Langzeitveränderungen nach Liposuktion bei Lipödem. *LymphForsch*. 2010;14(2):17–28.
16. Cornely ME. Liposculptur bei Lipödem – ein Eingriff in höchster Sicherheit. *J Lymphol*. 2002;1:1–3.
17. Cornely ME. Liposuktion bei Lipödem (cellulite) – follow up bei 140 operierten Patienten nach 7 Jahren. *Akt Dermatol*. 2004;10:3–21.