

Lipedema: A Commonly Misdiagnosed Fat Disorder

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Lipedema is a fat disorder that is often misdiagnosed. It was first identified at the Mayo Clinic in 1940, but medical schools do not include it in their curriculum and is therefore poorly understood. It presents as disproportionate and symmetrical accumulations of fat (bilateral), which is often accompanied by orthostatic edema. Early diagnosis and treatment are crucial, as the disease is progressive and can lead to immobility as well as a significant decrease in the quality of life. Lipedema differs from obesity because it does not respond to diet and exercise. This article gives you a glimpse into what lipedema is about and will help you identify some differences between lipedema and lymphedema. It will also help you identify which surgical procedures have been successful in treating the disease.

Lipedema is a fat disorder that was first identified in 1940 by Drs. Edgar Allen and Edgar Hines at the Mayo Clinic. The pathophysiology of lipedema is not clearly understood because it not taught or included in medical school courses (Box 1). Clinical diagnosis of lipedema is defined by the disproportionate and symmetrical accumulation of fat in the lower extremities, which is often accompanied by orthostatic edema (Allen & Hines, 1940).

The onset of lipedema is often seen during puberty, pregnancy, menopause, or other periods of hormonal fluctuations, which suggest a hormonal effect (Fonder, Loveless, & Lazarus, 2007). It is almost exclusively seen in women; however, there are reports of men who have been diagnosed with the disease. Földi and Földi (2007) suggest that 11% of the female population is affected by lipedema; however, it is not known to be prevalent in race or ethnic groups. Familial patterns range from 16% to 45%, whereas 50% of people who are diagnosed with lipedema are also overweight or obese, making it difficult to diagnose at times (as cited in Dayan et al., 2017).

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Those affected by lipedema often find they have column-shaped legs, often described as “tree-trunk legs,” which increase in size as the condition progresses (Figures 1 and 2). The subcutaneous fat deposits end just above the malleoli, or anklebones, with excess fat deposition in a “pantaloons distribution.” The feet are always spared. This causes an abrupt change noticeable at the level of the ankle between the abnormally enlarged legs and the normal-appearing spared feet (as cited in Dayan et al., 2017). This characteristic of an abrupt noticeable change at the ankles is commonly known as the “cuff sign” (Langendoen, Habbema, Nijsten, & Neumann, 2009). This is unlike lymphedema, which often affects both the lower extremities and feet at the same time. However, progression of the disease may lead to poor lymphatic circulation. Once this occurs, the feet will also become edematous. Lipedema fat also begins to accumulate in other areas of the body, which can then affect the arms and abdomen. When the lymphatic system becomes congested, it can cause “lipolymphedema,” which is diagnosed by the presence of dorsal foot swelling. A positive Stemmer’s sign is often noted at this stage (Goodliffe, Ormerod, Beale, & Ramcharitar, 2013; Okhovat & Alavi, 2015). To perform a Stemmer’s test, try to pinch and lift the skin at the base of the second toe or middle finger. If you can pinch and lift the skin, Stemmer’s sign is negative; if you cannot pinch and lift the skin, Stemmer’s sign is positive (Bjork, 2013).

There are a number of key diagnostic differences between lipedema and lymphedema. Lipedema always affects both sides of the body, whereas lymphedema in most cases is asymmetric. Lipedema of the legs does not affect the feet, and there is a negative Stemmer’s sign (in early stages). Lipedema affects the subcutaneous fatty tissue, whereas lymphedema is caused by impaired lymphatic circulation, thus causing an abnormal accumulation of lymph that occurs due to injury, infection, or congenital defects (Hespe, Nitti, & Mehrara, 2015). Although the lymphatic system is not affected in the early stage of lipedema, the accumulating fat continues to put added pressure on the lymphatic vessels. Lymphangiographic imaging shows that the lymph collectors within the proliferated fatty tissue have a coiled or corkscrew-like appearance rather than passing fairly straight toward the lymph nodes as is the case in healthy tissue, which can lead to a reduced transport capacity (Zuther, 2012).

BOX 1 Courses and Resources

Free CME Course for a maximum of 5.5 AMA PRA Category 1 Credits TM: <https://LipedemaProject.org/cme-setting-the-research-agenda-for-lipedema/>

<https://lipedemaproject.org/>

<https://lymphaticnetwork.org/news-events/the-lipedema-project-course-offerings>

<https://rarediseases.info.nih.gov/diseases/10542/lipedema>

<https://www.fatdisorders.org/>

lipedema-simplified.org

Limb swelling is common in lipedema and is exacerbated by orthostasis. Most patients also experience increased swelling in warm weather, during airline travel, or when sitting for long periods of time (Allen & Hines, 1940). In early stages of lipedema, the skin is soft and not discolored while evidence of edema is usually nonpitting (Fife, Maus, & Carter, 2010). However, when lipedema is accompanied with lymphedema, venous insufficiency, or stasis dermatitis, erythema, ulcerations, hyperpigmentation, cellulitis, and thickened skin can often be experienced (Okhovat & Alavi, 2015).

Lipedema fat is often painful and tender, as experienced with slight pressure on the affected extremities. It has been called the “painful fat syndrome” by those affected by it. Lipedema also triggers easy bruising, which is random with no defined pattern (Allen & Hines, 1940; Wold, Hines, & Allen, 1951). The extremities are heavy and enlarged from increased fat accumulations, which typically progress and develop at the medial knees, leading to decreased mobility and gait disturbances. This can later lead to hip or knee damage with increased discomfort and disfigurement as the fat continues to proliferate (Goodliffe et al., 2013).

Although general obesity responds to diet and exercise, there is little response with lipedema fat because it is resistant to diet and exercise. Thus, many patients



FIGURE 1. Stage III, Type III Lipedema exhibiting the disproportion between the upper body and large lower body from the buttocks to ankles. (This lady also shows signs of Stage IV, affecting the arms.) © The Lipedema Project. 2017-2018. All rights reserved. Used with permission.

have been unsuccessful at losing weight (Fife et al., 2010). When seeking advice from family physicians, patients are often told that there is nothing medically wrong and that they just need to be more serious about maintaining a healthy lifestyle. A common complaint by those who have the disease is that physicians do not believe they have worked hard at dieting and exercising because of their continued lack of success with losing weight. Weight comes off the unaffected areas but not the areas where the lipedema fat exists. This leads to feelings of guilt and failure as patients are made to feel they are not trying hard enough. The emotional despair is real, and hopelessness and depression can be overwhelming. A patient's self-esteem drops significantly and social activities decline. Often, people seek comfort through social media sites where various lipedema groups are formed and information is shared, along with encouragement and understanding, which is offered by fellow “lippy ladies” (M. Caruana, personal communication, September 2018).

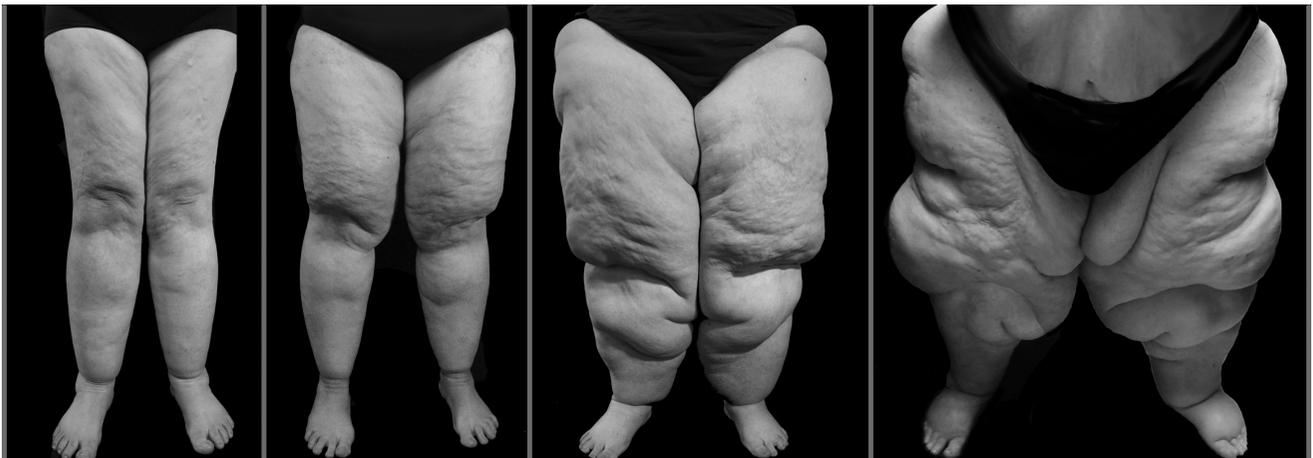


FIGURE 2. The four stages of lipedema. The disease progression through the stages is clearly noted. © The Lipedema Project. 2017-2018. All rights reserved. Used with permission.

STAGES AND TYPES OF LIPEDEMA (DAYAN ET AL., 2017)

There are four stages of lipedema:

- i. Smooth skin; increased fat in underlying tissues
- ii. Indentations and nodules present
- iii. Thickening/hardening of subcutis; large nodules and fat pads, especially on knees and thighs
- iv. Lipedema with lymphedema (lipolymphedema)

Four types of lipedema (Dayan et al., 2017):

- i. Affect the buttock and hips;
- ii. Affect the buttocks to knees, with fat accumulations on the medial side of the knees;
- iii. Affect the buttocks to ankles;
- iv. Affect the arms; and
- v. Affect the lower legs.

The causes of lipedema include but are not limited to (Dayan et al., 2017):

- Increased permeability and weakened capillaries leading to leakage of proteins and bruising;
- Increased size of adipocytes (increased size and increased diameter than unaffected adipocytes);
- Hormonal effects; and
- Heredity.

There is no cure for lipedema, but there are treatment options that might help slow down the progression. Proper diagnosis and treatment will help prevent complications and lipolymphedema (Dayan et al., 2017). Patients should be educated about the importance of exercise and physical activity, with movements that focus on pumping the leg and calf muscles to help increase lymphatic drainage and venous flow to prevent edema (Okhovat & Alavi, 2015). Lymphatic yoga, Pilates, and rebounding (low-impact exercise performed on a mini trampoline) are a few helpful activities that focus on improving lymphatic drainage, in addition to deep breathing and stretching (Fetzer & Wise, 2015; Wounds UK, 2017). The use of a vibration plate will also help move lymphatic fluid and reduce swelling. In early stages of lipedema, patients should also be advised to wear compression stockings or garments to help prevent and maintain lymphatic circulation. Other treatments include massage that focuses on moving lymphatic fluid, such as manual lymphatic drainage (MLD). The MLD massage helps move fluid and reduce swelling. It is often paired with compression garments, bandaging, or wrapping the limbs to minimize fluid buildup.

Although it is thought that bariatric surgery may help lipedema, Fife et al. (2010) suggest that patients with lipedema do not respond well to this procedure. Traditional “dry technique” liposuction can cause complications such as swelling to the lymphatics, with bleeding and damage to the lymph vessels, causing some critics to advise against liposuction therapy. However, tumescent liposuction,

which uses an infiltration solution, has improved the result of liposuction by facilitating fat removal while vasoconstricting the blood vessels with the epinephrine to reduce bleeding (as cited by Dayan et al., 2017). Another procedure that has been used successfully is water jet-assisted liposuction (WAL), which consists of a “fan-shaped water jet” that immediately separates the fat cells from the rest of the tissue while suctioning fat along with tumescent solution. Water jet-assisted liposuction differs from tumescent liposuction because less fluid is injected initially and then a large amount of fluid is pulsed in and out during the suctioning. With this technique, the tissues do not become fully tumescent or distended with fluid, which makes it easier to assess the contour of the limb during suctioning (Dayan et al., 2017). Tumescent and WAL liposuction have helped reduce lipedema fat, improve appearance of the affected limbs, and improve quality of life.

The distorted appearance of lipedema fat is very visible, but the psychological effects are unseen. We live in a society that has a negative opinion about fat in general and about obesity. Our society is quick to cast judgment by falsely assuming that lack of will power, along with poor diet and lack of exercise, is the reason for someone’s large shape or size. Although this is often true, at times it is not the case. As health care professionals we need to be aware of the signs and symptoms of lipedema so that we can help those who are experiencing physical and psychological pain and suffering caused by a disease that has been allowed to progress because the medical community has failed to diagnose the condition properly. As plastic surgical nurses, we see patients seeking liposuction procedures for weight loss. We should recognize the clinical diagnostic signs of lipedema. We should educate our fellow nurses, doctors, and surgeons to clearly understand the differences between lipedema and obesity and make it common knowledge so that those affected can find early diagnosis and attain the necessary treatment, reducing their progression and maintaining their quality of life as long as possible (M. Caruana, personal communication, September 2018).

REFERENCES

- Allen, E. V., & Hines, E. A. (1940). Lipedema of the legs: A syndrome characterized by fat legs and orthostatic edema. *Proceedings of the Staff Meetings of the Mayo Clinic*, 15, 184–187.
- Bjork, R. (2013, March/April). *Positive Stemmer’s sign yields a definitive lymphedema diagnosis in 10 seconds or less*. Retrieved September 17, 2018, from <https://woundcareadvisor.com/positive-stemmers-lymphedema-diagnosis-under-10-seconds/>
- Dayan, E., Kim, J. N., Smith, M. L., Seo, C. A., Damstra, R. J., Schmeller, W., et al. (2017). *Lipedema—The disease they call FAT: An overview for clinicians*. Boston, MA: Lipedema Simplified Publications, The Friedman Center for Lymphedema Research and Treatment at The Center for Advanced Medicine at Northwell Health in collaboration with Lymphatic Education & Research Network (LE&RN).
- Fetzer, A., & Wise, C. (2015). Living with lipoedema: Reviewing different self-management techniques. *British Journal of Community Nursing*, 20, S14–S19.

- Fife, C. E., Maus, E. A., & Carter, M. J. (2010). Lipedema: A frequently misdiagnosed and misunderstood fatty deposition syndrome. *Advances in Skin & Wound Care*, 23(2), 81–92. doi:10.1097/01.ASW.0000363503.92360.91
- Földi, M., & Földi, E. (2007). Lipedema. In M. Földi & E. Földi (Eds.), *Földi's textbook of lymphology: For physicians and lymphedema therapists* (2nd ed., pp. 417–427). Munich, Germany: Urban & Fischer.
- Fonder, M. A., Loveless, J. W., & Lazarus, G. S. (2007). Lipedema, a frequently unrecognized problem. *Journal of the American Academy of Dermatology*, 57(2), S1–S3. doi:10.1016/j.jaad.2006.09.023
- Goodliffe, J. M., Ormerod, J. O. M., Beale, A., & Ramcharitar, S. (2013). An under-diagnosed cause of leg swelling. *BMJ Case Reports*, 2013, bcr2013009538. doi:10.1136/bcr-2013-009538
- Hespe, G. E., Nitti, M. D., & Mehrara, B. J. (2015). Pathophysiology of lymphedema. In A. K Green, S. A. Sumner, & H. Bororson (Eds.), *Lymphedema: Presentation, diagnosis, and treatment* (pp. 9–18). Switzerland: Springer.
- Langendoen, S. I., Habbema, L., Nijsten, T. E., & Neumann, H. A. (2009). Lipoedema: From clinical presentation to therapy. A review of the literature. *British Journal of Dermatology*, 161(5), 980–986. doi:10.1111/j.1365-2133.2009.09413.x
- Okhovat, J., & Alavi, A. (2015). Lipedema: A review of the literature. *The International Journal of Lower Extremity Wounds*, 14(3), 262–267. doi:10.1177/15347346145554284
- Wold, L., Hines, E. A., & Allen, E. V. (1951). Lipedema of the legs: A syndrome characterized by fat legs and edema. *Annals of Internal Medicine*, 34(5), 1243–1250. doi:10.7326/0003-4819-34-5-1243.
- Wounds UK. (2017). *Best practice guidelines: The management of lipoedema*. Retrieved September 17, 2018, from <http://www.wounds-uk.com/best-practice-statements/best-practice-guidelines-the-management-of-lipedema>
- Zuther, J. (2012, December 13). *A closer look at lipoedema and the effects on the lymphatic system*. Retrieved September 14, 2018, from <http://www.lymphedemablog.com/2012/12/13/a-closer-look-at-lipedema-and-the-effects-on-the-lymphatic-system>

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