Lipoedema, still an under-diagnosed condition Practical guideline

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A SHORT DISEASE HISTORY

Patient M. is 31 years old. She has been struggling with obesity for years (fig.1 & 2). Despite all possible diets, she can't get her figure under control. Even the nutritionist she visited on GP's advice got frustrated. When loosing weight, her love handles diminished, her tummy flattened and her face started looking starved. She could hear people whisper the words "steel legs" when she was out on the street. And that was true. Her legs felt heavy and weary, pants didn't

fig. 1: Forced spread stand of the legs due to increased volume leads to abnormal way of walking

fit: too large at her waist and too narrow around her legs. Even the widest high boots didn't fit anymore. Those legs didn't belong to her. Sports becoming increasingly difficult, extra weight was quickly put on. Had she only known this...

LIPOEDEMA

Lipoedema is a (metabolic) disorder of the adipose tissue affecting the legs and sometimes the arms, generally leading to fat accumulation and increased volume.

In case of lipoedema, the metabolism of pathological fat cells is disturbed: whereas normally fat in the cells would be burned (lipolysis) and fat volume be decreased, lipids are stored in the tissues instead of being resorbed, resulting in increased fat volume.

The volume gained as a result from this disorder almost exclusively concerns women and often starts after puberty. The progression of the disease can sometimes be influenced by pregnancy.



fig. 1: Forced spread stand of the legs due to increased volume leads to abnormal way of walking

This might indicate a possible disfunction at hormonal and receptors level. Though the evolution of lipoedema can vary in gravity and secondary morbidity, it is always a progressive illness. 2

There are no hints as to whom could be affected by lipoedema, however there is clear evidence indicating that hereditary factors play a major role. It is estimated that 10% of the female population is affected by a more or less severe form of lipoedema of the legs, i.e. upper legs, though mostly under legs as well. 30% of this group also develop lipoedema of the arms, mostly upper arms.

Besides pointless diets and recurrent stigmatisation (obesity), patients often undergo lymphatic drainage at the manual therapist or physiotherapist. Complex decongestive therapy (CDT), a combination of manual lymphatic drainage, intermittent pneumatic compression and compression garments¹³ has an effect on the physical pain. However, wearing elastic compression stockings can be experienced as unpleasant or painful. The increased hypertrophy of fat cells results in secondary lymphoedema. However, there where the initial hypothesis was that capillary compression would lead to drainage restraint of the lymphatic vessels, it now seems more likely that the pathologic fat cells excrete inflammatory molecules. This causes lymphangiosclerosis, easy bruising and pain.

The effect of diets on the size of the legs frustratingly staying away as well as the subsequent limited mobility often lead to secondary obesity, which in turn leads to related diseases such as hypertension, diabetes, cardiovascular disease and mental disorders. Three main factors are to blame when talking about limited mobility: first of all the pain, secondly the mass around legs (and arms) that has to be carried and moved along, and finally having to move with legs further spread apart as a result of the increasing volume on the inner thigh. This last factor could also lead to secondary knee pathology, as seen in many lipoedema patients. The increasing volume of the upper arms results not only in limited mobility, but also in shoulder and neck pathology.

In time, lipoedema mostly leads to a growing need for medical care and frequently results in working incapacity and social isolation due for a great part to a negative self-image³, limited mobility and pain, severely undermining the quality of life¹. More and more research is being done on the disease, treatment, morbidity and quality of life⁴.

Therapy consists in the least traumatic possible removal of the greatest part of the abnormal tissues as well as in the reconstruction of proportions while keeping connective tissue, lymphatic and vascular structures untouched⁵.



fig. 3: The stature of the legs clearly differs from the trunk's stature despite the mild form of lipoedema. Palpation reveals a thick and firm subcutaneous layer.

STANDARD PATIENT HISTORY

Generally starting at puberty, lipoedema is characterised by an increased leg -sometimes arm-volume when compared to other parts of the body. Diets are often more frustrating than efficient; weight loss affects the rest of the body, but has no significant influence on the size of either legs or arms.

The skin of the legs is sensitive. Contact or pressure can be painful, easily causing bruising. Clothes hardly ever fit, the difference in size between the waist and the legs can vary from 1 to 2 or even 3 sizes. High boots don't fit either.

As the disease evolves, sports and exercise become increasingly difficult due to the growing volume of the legs and the pain caused by quick movement, like by jogging for instance. Symptoms can also include the two-body-syndrome, i.e. the feeling that the legs don't belong to the rest of the body, not to mention social isolation.

PHYSICAL EXAMINATION

A very thick (fig. 3) and disproportionate (fig. 4) subcutaneous fat layer can clearly be seen from the hips or upper legs down to the knees or ankles. This can also be present in the upper and/or under arms (fig. 5)

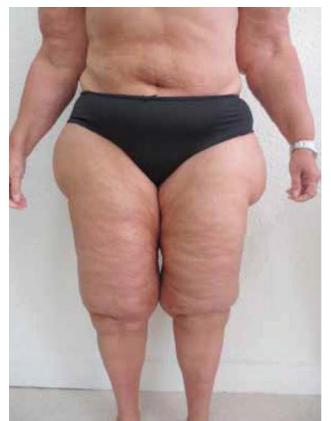


fig. 4: Typical though less common transition from lipoedema of the upper legs to normal under legs

The tissues feel firm to palpation while the upper skin of the legs often has an irregular and uneven aspect.

The deformities are mostly symmetrical, showing a distinct transition from affected to normal (fat) tissues.

A further stage in lipoedemic evolution can lead to "cuff formation" around the ankles (fig. 2) and obvious fat pads under the inner side of the knees. Trunk, face and non-affected parts of the body have a normal aspect.

The forced straddle stand of the legs due to increased inner thigh volume can be more or less noticeable (fig. 1). This can possibly be the cause of knee complaints as seen in many lipoedema patients.

TREATMENT

Besides extensive explanation about the disease, possible treatments and expectations, tumescent liposculpture is currently the only adequate (surgical) treatment of lipoedema⁷.

The tumescent technique consists in injecting the right amount of a liquid made of physiological salt solution, lidocaine, epinephrine and natriumbicarbonate, in order to create a watery subcutaneous environment from which the adipose tissue can be removed easily without damaging the lymphatic or connective tissue structures.

This technique is an efficient treatment ensuring quick recovery, optimal contraction of skin and connective tissues and minimal risks of secondary lymphoedema. Fat tissues are more evenly removed thanks to the preoperative increased volume obtained by the tumescent solution.

Per treatment session, a same area is symmetrically treated on each side, left and right.

Lipoedema patients need two to four treatments in order to achieve an efficient reduction of the upper legs and maximal results, preventing lipoedema to recur on the same places in the future.

Under legs can be treated in one session, provided they are treated as a whole and not only calves or ankles.

Removing large amounts of fat doesn't enhance the risks of complications⁹.

Risk-bearing factors¹⁰ are related to the extent of the treated area, the use of too little (tumescent) solution, as well as undergoing the treatment under general anaesthesia. Those factors can increase the risks of thrombosis, skin necrosis and lidocaine intoxication. The maximal lidocaine dose in combination with liquid pressure is a sensible restriction as to the dimension of the area that can be treated in one session.

Unfortunately, this treatment is not covered by health insurances in the Netherlands.

Secondary lymphoedema can show at a later stage.



fig. 5: Lipoedema of the upper arms and to a lesser extent the under arms

OTHER TREATMENTS/THERAPIES/ADVICE

There is a number of treatments ⁷ and advice which can be helpful, though only to some extent.

Worth mentioning are lymphatic drainage as a keystone in the treatment, diet counselling in case of obesity component, exercise counselling in order to train muscular strength, adapted footwear to support sagged feet, and psychological guidance as for how to deal with the disease in the best possible way.

A regular calorific intake should be observed as (high calorie) peaks result in increased fat deposition in the abnormal fat cells which subsequently cannot be evacuated.

RESULTS AND EFFECTS

Weight and size decrease of the legs (and/or arms) enables moving much more easily11. Clothing will fit better. As mobility increases, frustrations and social hindrance often disappear. Patients regain selfconfidence, psychological well-being, enhancing weight loss in patients with secondary adipositas The pain due to lipoedema is considerably reduced, or definitely gone¹². Next to pain reduction, the appearance of oedema and haematoma diminishes¹⁴. It appears that 22% of the patients preoperatively treated with CDT don't need any postoperative therapy anymore¹², including not having to wear compressive devices for the rest of their life. The quality of life eventually improves. Skin excess mostly doesn't show as the skin usually progressively retightens during the process. However, the removal of large amounts of fat can leave the skin somewhat wrinkled. Such a side effect doesn't weigh in comparison to the benefits of the treatment, though. Taking away a large part of the lipoedemic tissue in the treated areas has lasting results: it will not come back. Even though remaining fat cells could continue to grow, treatment always guarantees an improvement for the future. Adapting one's lifestyle (if necessary) can contribute to the prevention of a relapse.

TREATMENT, A SUMMARY

- Extensive preoperative patient information and instructions (oral and written);
- Preoperative screening (ASA-score, blood test, current allergies and medication);
- Preparation: photographs, disinfection, markings;
- Tumescent anaesthesia (20'-40'): making simultaneous use of several needles, infiltration of anaesthetic tumescent solution via pump and perfusion system, regulating pressure as to not be painful for the patient. Target is a tensed, livid looking skin within the depicted area. Processing time is about 40'.
- Liposculpture using vibrating microcannula's² specifically adapted to the area to be treated. Access to the fatty tissue through a couple of 5-10mm incisions.
 Working from the deeper fatty layers to the surface in a fanlike pattern. This process lasts 1 to 2 hours, depending on the dimension of the treated area.
- Post-treatment⁵: post-operative compression and lymphatic drainage. Absorbing bandage on treated areas and compression garments for 24 hours. All devices can then be taken off and patient may shower. Compression garment should from then on be worn at least one week day and night, then 2 weeks during the day. CDT may be applied on indication.
- Future treatment: 4-6 weeks until next treatment, provided fairly complete recovery of adjacent area.

SUMMARY

Lipoedema is still an under-diagnosed condition, mainly due to lack of familiarity with the clinical picture and history. However, the condition results in high morbidity with a substantial impact on the quality of life. This article gives us tools on how to recognize these patients to ensure they receive the necessary attention.

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