Opóźniona diagnoza i leczenie pierwotnego obrzęku limfatycznego kończyn dolnych – studium przypadku

Delayed diagnosis and treatment of primary lymphoedema of lower limbs – case study

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Streszczenie:
Leczenie pierwotnego obrzęku limfatycznego stwarza wiele problemów pacjentowi jak i całemu zespołowi medycznemu. Całkowite wyleczenie bowiem z racji uszkodzenia anatomicznych dróg odpływu chłonki nie jest możliwe, a terapia ma na celu jedynie zmniejszenie rozmiarów kończyn, przywrócenie jej funkcji i poprawienie aspektów kosmetycznych. Leczenie jest trudne, wieloetapowe i nawet w przypadku dobrych wyników kosztowne i czasochłonne. Obrzęki pierwotne nie są patologią bardzo częstą, toteż problem związany z ich diagnostyką i leczeniem pozostaje często na marginesie zainteresowania lekarzy. W przeciwieństwie do dobrze zorganizowanej pomocy, którą otrzymują chore z obrzękiem w obrębie kończyn górnych (głównie po leczeniu chirurgicznym raka sutka) możliwości rozpoznania, diagnostyki i wdrożenia postępowania usprawniającego u osób z idiopathic postacją obrzęku limfatycznego kończyn dolnych są istotnie ograniczone. Chciać w ostatnich latach intensywnie propagowana jest wiedza dotycząca zasad stosowania terapii przeciwzastoinowojej nadal w wielu ośrodkach wdrażana jest nieprawidłowo. Nieleczony lub niewłaściwie leczony obrzęk staje się nieodwracalny wskutek rozprzestrzenienia tkanki łącznej, czym różni się od obrzęków na innym podłożu. Narastający obrzęk doprowadzający do stopniowej deformacji i zaburzeń funkcji kończyn, wpływa negatywnie na stan psychiczny pacjenta. Późne rozpoznanie, kosztowne, uciążliwe i długotrwałe leczenie, którego efekt jest niejednokrotnie niezadowalający wpływa istotnie na jakość życia chorego. W pracy przedstawiono jeden z licznych przypadków pacjentek z rozpoznanimi późno i nieprawidłowo leczonymi obrzękami pierwotnymi, które mają być ilustracją typowych zaniedbań w dążeniu do ustalenia diagnozy i wdrożenia leczenia w tej jednostce chorobowej. Podkreślono także rolę badania LS jako złoconego standardu w diagnostyce obrzęków pierwotnych i jej ograniczonej przydatności w ocenie skuteczności leczenia.

Słowa kluczowe:
obrzęk limfatyczny, pierwotny wrodzony obrzęk limfatyczny, rehabilitacja, limfoscintygra

Abstract
Treatment of lymphoedema is problematic for the patient and the entire medical team. Complete recovery, because of anatomical damage to the outflow tract of lymph, is not possible and the therapy is intended only to reduce the size of the limb, restore its function and improve the cosmetic aspects. Treatment is difficult, multistage, unpredictable and even if results are good, expensive and time consuming. Primary lymphoedema is not common disease, so the problem of its diagnosis and treatment is often on the margins of medical concern. In contrast to the well-organized support received by patients with secondary oedema of the upper, diagnosing and implementation of rehabilitation in patients with idiopathic lymphoedema of the lower limbs are significantly limited. Although in recent years is intensely propagated knowledge of the principles of anticongestive therapy, it is still is implemented incorrectly in many centers. If left untreated or improperly treated swelling becomes irreversible as a result of proliferation of connective tissue, what differs from cases of oedema not associated with lymphatics damage. Increasing swelling will lead to progressive deformity and dysfunction of limbs and negatively affects the mental state of the patient. Late diagnosis, costly, cumbersome and long-term treatment with the often unsatisfactory effect significantly affect quality of life. The paper presents one of many our cases of patients with late diagnosed and improperly treated primary lymphoedema and is an illustration of the typical negligence in establishing the diagnosis and initiation of treatment in this disease entity.

Key words:
lymphedema, primary congenital lymphedema, rehabilitation, lymphoscintigraphy
The diagnosis of primary lymphoedema and initiation of appropriate treatment is usually greatly delayed. In spite of continuous progress in diagnostic and therapeutic methods, creation of more and more special medical centres dealing with diagnostics and therapy of oedema, numerous scientific and popular science articles on this subject, we do not observe any improvement in frequency of taking primary lymphoedema into account in diagnostics. General practitioners and even surgeons usually monitor these patients in their clinics for a long time, often suspecting an injury, inflammation or disturbances in venous circulation [1].

As a result of the progress in oncological surgery, secondary lymphoedema is observed more and more often, due to iatrogenic damage to lymph vessels and removal of the affected lymph nodes. Also recurrent infections, venous insufficiency, and filariasis may contribute to the increasing number of lymphoedema patients. Globally, over 300 million people suffer from oedema, and their number is growing [2]. Secondary lymphoedema is currently of primary interest to lymphological research centres, which are formed mostly in association with oncological departments. Because of intensive development of vascular surgery, focused on blood vessels, the active treatment of lymph vessels unfortunately is often outside the scope of interest of physicians. Proper lymph circulation, however, is necessary for drainage of interstitial spaces. The human body, weighing on average 65 kg, includes 3 dm3 of plasma in vascular circulation and 12 dm3 of interstitial fluid. About 8-12 dm3 of lymph is produced per 24 h, including 4-8 dm3 that are reabsorbed to blood vessels as ultrafiltrate. Thus lymph vessels do enormous work [3].

Primary lymphoedema is not frequent, so problems with its diagnosis and treatment is often of marginal interest to physicians. It affects about 1.15 children and young adults per 100 000 population. They are mostly girls during puberty [4]. Primary lymphoedema is caused by inborn anatomical malformations of lymph vessels. In many cases their genetic background has been confirmed [2]. For clinical purposes, several types of primary lymphoedema are distinguished, based on the age of onset.

1. Congenital lymphoedema, appearing at birth or soon after birth (before 2 years of age). Often familial, affecting 6-12% of patients with primary lymphoedema. Cases of congenital lymphoedema may be linked with abnormal lymph transport in intestines (intestinal lymphangiectasia), causing diarrhoea with considerable protein loss.

2. Lymphoedema praecox, occurring usually during puberty, before 35 years of age. Rarely familial, usually sporadic, affecting 83-94% of patients with primary lymphoedema [4, 5]. Much more frequent in females, suggesting involvement of oestrogens in its etiopathogenesis [6].

3. Lymphoedema tarda, occurring after the age of 35 years and affecting up to 11% of patients with primary lymphoedema [7, 8, 9].

As mentioned above, gene defects have been confirmed recently in many cases of primary lymphoedema, e.g. in Milroy disease in children, manifested as bilateral lower extremity oedema below knees and in boys also hydrocoele
testis, a mutation was found in the FLT4 gene, which codes for vascular endothelial growth factor receptor, VEGFR-3 [10,11,12,13,14]. In Nonne-Milroy-Meige syndrome, affecting mostly girls during puberty, no genetic defect has been identified so far. Oedema is then bilateral, symmetric, and involves legs above the knees. Some chromosomal aberrations are associated with primary lymphoedema too (Turner syndrome, Klinefelter syndrome, etc.). In familial lymphoedema, autosomal dominant inheritance is documented [15,16]. However, in spite of confirmed genetic background in many cases, primary lymphoedema is usually sporadic [4,5], which makes proper diagnosis more difficult. Familial lymphoedema accounts for only 5-10% of cases.

Clinical manifestation is usually observed only when interstitial fluid volume is doubled, as compared to the normal value. The accumulated interstitial fluid is rich in proteins and contains also lipids. It leads to increased colloid osmotic pressure and water retention. Initially the oedema is reversible, noticeable as asymmetry of limb circumference in the morning, but disappears during the day. Prolonged lymphostasis leads to accumulation of fibroblasts, adipocytes, and keratinocytes in the swollen area. Also collagen and adipose tissue deposition is increased. In connective tissue, fibrosis is observed [17,18], especially in primary lymphoedema. The oedema is then present permanently and its hardening can be noticed. Patients then complain of tenderness and difficulties in using the limb. The outline of the limb and its functionality change. In such cases usually secondary lymphoedema is diagnosed, associated with arteriovenous malformations.

Primary lymphoedema aggravates progressively and in the absence of treatment may lead to many complications: recurrent infections, especially streptococcal (erysipelas) and fungal, recurrent infections of lymph vessels with accompanying fever, or even a tumour – lymphangiosarcoma in the swollen area [19,20]. Pain, chronic changes causing disability, and remarkable changes in aesthetic appearance, result in a substantial decrease in the patient's quality of life [21,22]. The lack of perspectives for future improvement in primary lymphoedema, emphasized quite frequently by general practitioners, gives rise to the feeling of hopelessness, especially in young people. The patients frequently experience fear, anxiety, depression, difficulties to adapt to new situations, and problems with professional, family, and sexual life [23,24,25]. About 10% of patients lose their job [20]. Because of the very late diagnosis, patients receive proper treatment with a great delay, frequently after a few years or are not treated at all [26]. The patient is usually convinced that the problem is mild and little can be done to help. In fact, however, currently thanks to the progress in rehabilitation and modern treatment schemes, the patients can be maintained in a good physical and mental condition if the therapy is started early (consensus).

To identify the cause of oedema, various imaging methods can be used. The invasive lymphography is now replaced by lymphoscintigraphy (LS). It is a non-invasive method, easily repeatable, inexpensive, having no negative effect on the
vessel endothelium [27]. Since 1950, when it was used for the first time, it has been continuously improved, using new, more accurate radiotracers. It is currently a gold standard in diagnostics of primary lymphoedema, and according to some authors is useful in monitoring of treatment outcome [28]. Interpretation of the LS results in oedema patients requires some experience and depends not only on the skills of the radiologist but also on the depth of tracer injection, radiotracer dose and type, and muscle pulp activity [29, 30]. It is not difficult to diagnose primary lymphoedema on the basis of a properly performed LS, as the LS image of this disease has some characteristic features [31]. Imaging of lymph vessels and nodes with the use of LS is currently more and more common in lymphological centres, although work on its proper standardization is continued [26]. Sensitivity and specificity of this method in diagnosing of lymphoedema is estimated at 92% and 100%, respectively [32]. It is believed that currently LS is the best method for confirmation of lymphoedema as well as functional and anatomical assessment of lymph vessels [33, 34]. The migration of colloid particles, labelled with technetium-99m isotope, precisely images the rate of lymph flow, with simultaneous evaluation of lymph nodes. In a properly working lymphatic system, the tracer moves at a normal rate in the lymph vessels, symmetrically in both extremities. A lack of radiotracer flow from the injection site, delay of its flow, asymmetry between the extremities, broadening or blurring of lymph vessels and regional lymph nodes, are signs of the disease. Pathological changes include also lymph backflow in smaller lymph vessels, mostly at the level of the skin and subcutaneous tissue (dermal backflow).

Considering all the advantages of LS, the few contraindications to it, and perfect suitability of this method for imaging of oedemas, it is surprising that it is not taken into account in routine diagnostics, especially with respect to recognition of primary lymphoedema.

In this paper we report a case, selected from many similar cases among our patients with greatly delayed diagnosis and wrongly treated primary lymphoedema. Their histories illustrate typical neglect in both diagnostics and initiation of treatment of this disease. We also emphasize the role of LS as a gold standard in diagnostics of primary lymphoedema and its unsuitability for follow-up assessment.

**Case report**

Patient M.C., 46 years old, working as an accountant. At the age of 30 years, after increased physical activity (longer journey), she noticed oedema near the right ankle, which was not associated with any other symptoms. The size of the oedema initially varied, but later it continuously increased. A general practitioner referred the patient to an orthopaedic surgeon who, in turn, referred her to a vascular surgeon, to make an accurate diagnosis. On the basis of Doppler ultrasound findings, venal insufficiency was detected and Detralex was prescribed. The patient was not offered any diagnostic or therapeutic treatment. The oedema was gradually aggravating, the left lower extremity was inflamed, swollen, with limited mobility in the knee and
ankle. Her general condition was deteriorating continuously.
The course of the disease was complicated by a streptococcal infection (erysipelas). As late as 8 years since the onset of the disease, after another visit to the surgeon because of aggravation of the changes and pain, LS was performed. It revealed disturbances in the structure of lymph vessels and lymph nodes of the right leg. A filariosis test gave negative results, so she was diagnosed with lymphoedema praecox.

The patient was referred to the Department of Physiotherapy at the Medical University of Gdansk, where since 2006 she has attended decongestive lymphatic therapy (DLT). It included manual lymphatic drainage, compression pump treatment, special lymphoedema bandaging or compression stockings (compression class II, followed by class III), and physical therapy. She also learned simple manual lymphatic drainage techniques (to be used by herself), proper skin care, and remedial exercises. During the therapy, an ultrasound was performed, which detected progressive fibrosis of the subcutaneous tissue in the swollen area.

After 7 years of regular physiotherapy (2006-2013), the patient’s subjective condition was continuously improving. The patient said that the leg seemed to be less heavy, the oedema was softer, and limb functionality had also improved (Figure 1). During the treatment, no infection has occurred. She felt less pain and her mental condition was better.

Fig. 1. Patient M.C. Lower limbs after 7 years of rehabilitation physiotherapy
For objective follow-up assessment, LS was repeated but it did not detect any differences, in comparison with the LS performed 7 years earlier. LS images of the lower limbs and the abdominal cavity were made in anterior projection 3 h after administration of the radiotracer. In both images (year 2006 and 2013), remarkable abnormalities of lymph flow are visible in the lower right limb and the degree of lymph flow impairment is similar. They both show severe impairment of radiotracer uptake in inguinal lymph nodes and iliac on the right side and extensive diffusion of the radiotracer in dermal lymphatics. Additionally, in the earlier image (2006), lateral flow towards the other limb was observed at the level of the lesser pelvis (Figure 2).

Fig. 2. Patient M.C. – lymphoscintigrams in 2006 and 2013. In both images no normal lymphatics are present. Extensive dermal backflow of lymph on the right in both studies. Only small number of inguinal and iliac lymph nodes.
LS was performed after intradermal injection into the 1st and 2nd interdigital web spaces of 0.5 ml of suspension of 99mTc-labelled human albumin colloid with a particle size of < 80 nm in activity 37 MBq. Images were recorded at 30 min, 1 h, and 3 h after administration of the radiotracer, in the whole-body anterior projection, with a scanning speed of 12 cm/min, 256 × 256 matrix and low-energy high-resolution collimators, using Siemens MS-2 gamma camera in 2006 and Siemens Symbia T6 in 2013. At the beginning of the LS in 2006, additionally a series of dynamic images was recorded, encompassing the distal part of the thigh and the proximal part of the calf (10 × 1-min frames). Then in the projection of the thigh and the right calf, some of the tracer accumulated in many dilated, tortuous lymphatic collaterals. Activity of the tracer increased in the right limb during the 10 min monitoring, in contrast to the left limb, where radiotracer ascent was fast, resulting in a considerable decrease in its activity (Figure 3).

![Image](image_url)

**Figure 3.** M.C. – dynamic lymphoscintigram of lower limbs in 2006, dynamic acquisition time 10 min. Visible accumulation of the radiotracer in the network of dilated lymphatics in the right limb; quantitative analysis (radiotracer activity curves) confirms lymph obstruction on the right side (increasing trend in the right limb)

Patient condition after the therapy was additionally assessed by comparing the circumference of the healthy limb and of the oedematous limb every time before and after physiotherapeutic treatment. The measurements were made in a standing position at selected points on both limbs: 1st at the height of the groin, 2nd at 10 cm above the patella base, 3rd at the height of the patellar groove, 4th at 10 cm below the patella, 5th at 2 cm above the ankle, and 6th at 1/3 of the foot length from the toe tip. To assess the significance of differences in limb circumference before and after treatment, the values were subjected to the Shapiro–Wilk test, and as their distribution was not normal, the nonparametric Wilcoxon signed-rank test was used.
Table 1. Patient M.C. Statistical significance of differences in limb circumference in selected measurement points

<table>
<thead>
<tr>
<th>Measurement point</th>
<th>p-level</th>
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<tbody>
<tr>
<td>Groin</td>
<td>0.008</td>
</tr>
<tr>
<td>Thigh</td>
<td>0.021</td>
</tr>
<tr>
<td>Knee</td>
<td>0.87</td>
</tr>
<tr>
<td>Calf</td>
<td>0.097</td>
</tr>
<tr>
<td>Ankle</td>
<td>0.03</td>
</tr>
<tr>
<td>Foot</td>
<td>0.02</td>
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</table>

Differences between measurements before and after intensive treatment at the height of groin, thigh, ankle, and foot are significant ($p < 0.05$). The smallest differences are noticeable at the level of the knee joint.

Figure 4. Patient M.C. – measurements of the circumference of the oedematous limb in the course of intensive physiotherapy, in comparison to the healthy limb
To determine if the applied therapy gives long-term effects, the method of least squares was used to fit a linear function to the data. In table below (table 2) slope coefficients calculated for all measured point in lower limb.

The results in Table 2 indicate that all the estimates are close to zero. This suggests that the applied physiotherapy did not cause any permanent decrease in limb circumference in the selected measurement points. Thus no long-term effect can be achieved now, unfortunately.

**Table 2. Patient M.C. Values of slope coefficient a in selected measurement points**

<table>
<thead>
<tr>
<th>Measurement point</th>
<th>Slope coefficient a</th>
</tr>
</thead>
<tbody>
<tr>
<td>Groin</td>
<td>0.0019</td>
</tr>
<tr>
<td>Thigh</td>
<td>0.0022</td>
</tr>
<tr>
<td>Knee</td>
<td>-0.0004</td>
</tr>
<tr>
<td>Calf</td>
<td>0.0014</td>
</tr>
<tr>
<td>Ankle</td>
<td>0.0026</td>
</tr>
<tr>
<td>Foot</td>
<td>0.0005</td>
</tr>
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</table>

**Discussion**

Establishing a proper diagnosis and initiation of proper treatment of primary oedema remains a challenge for many physician specialists. It seems that this is mostly due to the lack of diagnostic vigilance of general practitioners, who in the course of diagnosing do not take into account the possibility of detection of inborn malformations of lymph vessels. In the case of limb oedema, usually the patient is tested for systemic diseases or traumas. A detailed clinical examination and a detailed case history are very helpful in explaining the causes of oedemas associated with heart, kidney, and liver diseases, oedemas associated with hypoproteinemia in cases of severe malnutrition, as well as endocrine disturbances (hypothyreosis). Available data on the factor triggering the oedema, the rate of its increase, accompanying symptoms from other organs, skin condition, warmth, haematoma, presence of pulse on the oedematous limb, consistency of the oedema, as well as family history, allow to approach the proper diagnosis very precisely [28, 35]. As a rule, however, most often without appropriate justification and additional tests, secondary oedema is diagnosed. In the case presented above, the true diagnosis was greatly delayed.

Patient, M.C., was diagnosed with an 8-year delay. Initially her lymphoedema was misdiagnosed as post-traumatic oedema, treated with home cures (compresses, etc.). After a long time, the patient visited a general practitioner, who referred her to an orthopaedic surgeon, and finally to a vascular surgeon, who started treatment for venous insufficiency, without additional tests. LS was performed as late as 8 years after the onset of the oedema, and only then primary lymphoedema was diagnosed.

For many years it has been known that LS is a gold standard in imaging and differentiation of various causes of oedema [35]. As a simple method, almost completely non-invasive, it nearly entirely replaced lymphography, which required the use of oil contrast, causing complications [27]. Symptoms of primary lymphoedema can appear at various ages, from birth to 35 years or even later, and lymphoscintigrams may differ and require inter-
pretation, depending on patient age (child, adolescent or adult). Most authors report that LS may exclude or confirm lymphoedema with nearly 100% certainty [2, 36, 37]. Admittedly, although there is a consensus about the great usefulness of LS in identification of lymphoedema, there are some doubts if it can serve to distinguish primary lymphoedema from secondary lymphoedema [29, 30, 36, 37]. According to most authors, the most characteristic feature of primary lymphoedema is a complete lack of radiotracer transport, apparent absence of lymph nodes [27, 31, 37]. The latest consensus on diagnosis and treatment of primary lymphoedema emphasizes that diagnosis of late-onset primary oedema (lymphoedema tarda) requires special inquisitiveness [35]. In such cases some authors believe that LS is not sufficient as a single method for unambiguous diagnosis of primary lymphoedema [38, 39, 40], and suggest that it should be supplemented with duplex ultrasound [41, 42]. It seems that broadening of diagnostic methods is particularly necessary if a surgery is planned.

In our patient, LS images were typical for primary lymphoedema and did not cause any diagnostic problems for the radiologist. In presented patient, the LS showed that the radiotracer was nearly completely absent in inguinal and iliac lymph nodes in the oedematous limb. Low-intensity radiotracer flow was detectable in tortuous lymphatic collaterals. Collateral flow was also observed in the pelvis to lymph vessels of the contralateral limb. Severe impairment of radiotracer uptake was also visible in region of inguinal and iliac lymph nodes in the oedematous limb, accompanied by extensive diffusion of the radiotracer in dermal lymphatics.

It is debatable, however, if LS allows objective follow-up assessment of lymphoedema in patients offered intensive decongestive lymphatic therapy (DLT). As a method that can be repeated many times because of the minimum invasiveness, LS is used for this purpose in some centres [28, 29, 31, 37]. Other authors do not find any changes corresponding with improved lymph flow in LS performed during treatment [43]. The same applies to our patient. She reported improvement in the subjective well-being, decreased tenderness of the oedema, reduced size of the oedema, which allowed the first patient to wear shoes of the same size, and generally improved the functionality of the limb. LS was performed several times, at the beginning and during the therapy, but gave nearly identical results. This may be due to the fact that DLT leads only to a decrease in water components within the oedema, but does not reduce the retention of proteins, lipids, and other components of the oedema fluid. The therapy may also help by putting pressure on the capillaries and reducing the filtration, and thus limiting lymph formation [37, 43]. The observations made so far indicate that LS is a gold standard for identification of lymphoedema, but is not useful for follow-up assessment. Further improvements of the LS method are currently disputed, primarily on its standardization with respect to the applied radiotracer type, uniform radioactivity doses, injection volume, injection location (intradermal or subcutaneous), and time intervals of the imaging. The protocol should take into account the patient’s activity during the LS [44]. When these principles are applied, results from various lymphological centres will be fully comparable. In recent years, many works concerning principles of lymphoedema staging have been published. In the past, the major factors taken into account were: oedema size, tenderness, hardness, limb shape change, excluding the patients’ economic status (job) and lowered quality of life [45, 46, 47]. New
versions of the grading system will additionally be based on clinical assessment of the patient and the LS. The system should take into account the frequency of recurrent infections, degree of fibrosis, skin condition, and the patient’s compliance in the long and arduous therapy. The patient’s compliance to the treatment of primary lymphoedema is often limited by the common statements of physicians, already at the moment of establishing of the diagnosis, that the disease is incurable and all efforts will be fruitless [35]. Current treatment of chronic lymphoedema is based primarily on DLT [48, 49, 50] and it aims mainly to improve the functionality of the oedematous limb and the quality of life [51, 52]. Patients need to be informed about the necessity of skin care to prevent streptococcal infections. Attention is paid also to psychological aspects (self-acceptance, in spite of the often huge deformations) as well as preparation for work and life in the society. Currently surgical interventions are offered more and more often [53, 54], including plastic surgery of lymph vessels. Decisions about surgical treatment must be made by a multidisciplinary team [54], which will assess the chances of success.

Because of the unsuitability of LS and ultrasound as methods for follow-up assessment, we attempted to measure changes in our patients’ oedematous limb circumference before and after DLT. The measurements showed that soon after treatment, in some of the measurement points, limb circumference was reduced significantly, but the changes were short-term. In spite of the lack of objective criteria of oedema improvement in the course of intensive therapy, the patients subjectively felt much better, both physically and mentally. Limb mobility/functionality was improved considerably, and the swollen tissues were much softer. Skin condition was satisfactory all the time, with no infection. These patient was selected because of perfect co-operation and continuation of the therapy for many years without breaks (she wore compression garments day and night, and took them off only for hygienic purposes). The metric method is often used by physiotherapists for follow-up assessment. It is not precise, and there are no well-defined standard measurement points on the limbs, while casual placement of the measurement tape and its pressure of the tissues increase the measurement error. Johnston emphasized a lack of an objective tool for oedema assessment, which makes co-operation of various research centres impossible [55].

**Summary**

1. Diagnosis of primary lymphoedema is often greatly delayed (8-10 years).
2. Late initiation of treatment, when fibrosis is advanced and recurrent infections are observed, greatly worsens the prognosis.
3. Lymphoscintigraphy is a gold standard in diagnosing of primary lymphoedema, but in spite of its low invasiveness, physicians (especially general practitioners) rarely use it for diagnostic purposes.
4. Lymphoscintigraphy is not useful for follow-up assessment of primary lymphoedema.
5. Our attempt to assess oedema size in the course of DLT by a metric method has shown only short-term, transient decrease in circumference of the oedematous limb.
6. The therapy must be maintained all the time, periodically at home, by the patient applying simple manual lymphatic drainage techniques, learned in a medical centre.
7. In spite of the objective lack of confirmation of a decrease in oedema in the course of DLT, the patients reported a remarkable improvement of their mental and physical condition, better functionality of the oedematous limb, reduced tenderness, and pain in the limb.

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