LYMPHOLOGY TODAY | RESEARCHES | CURRENT EVENTS

# LIMPHA

NUMBER 3 | AUGUST 2017

# TAPING
## IN LYMPHOLOGY


**Lymphology** in Uzbekistan


**Syndrome** Klippel-Trenaunay-Weber


**Lymphangiosarcoma** - the worst complication of lymphedema

Dear friends!

This issue covers two rare and at the same time complicated topics in the work of a lymphologist - the syndrome of Klippel-Trenaunay-Weber and lymphangiosarcoma.

The causes of the syndrome of Klippel-Trenaunay-Weber remain a mystery for doctors till date. Less than 0.00001% of people in the world face this disease. At the moment, the treatment of this syndrome is symptomatic, and the correct approach to it allows to achieve a significant improvement in the quality of patients' life encountering it.

Lymphangiosarcoma is one of the most terrible complications of lymphedema, which progresses very quickly and leads to death. The most important thing in this case is the fastest diagnosis and timely treatment.

I hope a lot that this will help to improve, and, perhaps, to save a person's life. We will also tell you about the use of taping in the treatment of lymphedema and introduce you interesting specialists from the world of lymphology.

I hope that this issue will be interesting for you!

Good luck to you!

Sincerely, Ivan Makarov
Chief Editor of LIMPHA journal

*Ivan Makarov,*
*MD, lymphologist,*
*chief editor of journal*
*"LIMPHA"*

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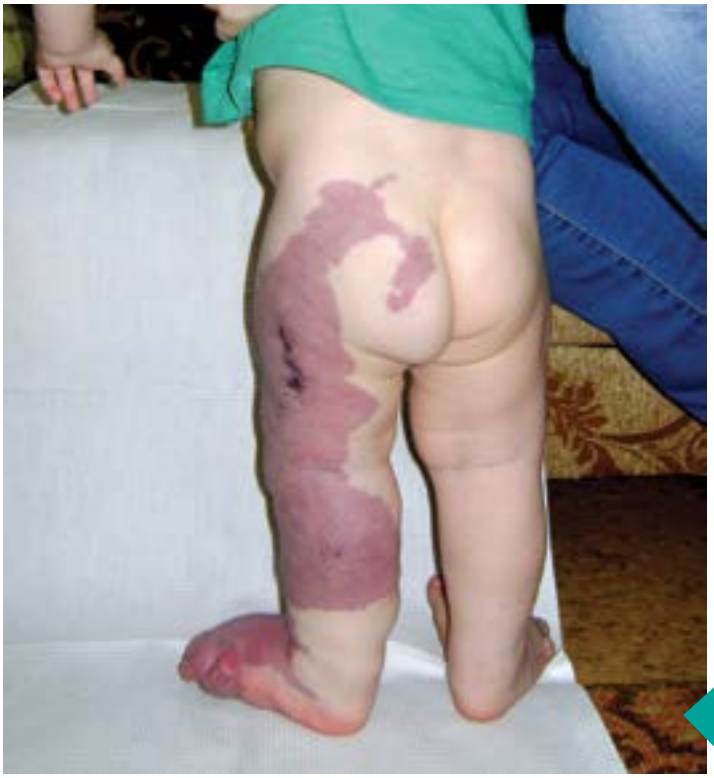

*How to help patients with Klippel-Trenaunay Syndrome*

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# European Society of Lymphology Congress in Stuttgart

*On May 26-27, 2017, The European Society of Lymphology Congress (ESL-2017) was held in Stuttgart (Germany).*

According to the President of the Congress, Professor Etelka Foeldi, this year the main goal of the congress has been to discuss the interdisciplinary approach to treating patients with diseases of the lymphatic system, as well as determining the place of lymphology among other specialties.

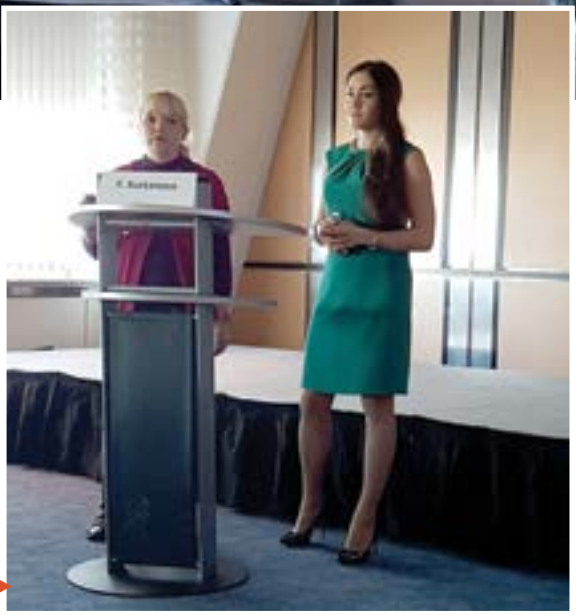
This event brought together lympologists and scientists from all over Europe. From Russia, the congress was visited by doctors dealing with the treatment of patients with lymphedema and other pathologies with the lymphatic system: Alexandra Rovnaya, Ivan Makarov, Aigiz Feiskhanov, Yuliya Demekhova.

*On the photo: Professor E. Foeldi, and G. I. Makarov. Foldi E. and V. S. Makarova are the persones who provided the effective treatment lymphedema in Russia in 1996.*

*Welcome speech of the President of the Congress E. Foeldi*

Ivan Makarov (LIMFA SPC, Moscow) made a presentation on the effectiveness of complex decongestant therapy (CDT) in women with postmastectomy edema in Russia.

Moreover, the clinical psychologist of SPC LIMFA, the head of the special psychology and rehabilitation department of the Moscow State Psychological and Pedagogical University, Yulia Kurtanova spoke about the peculiarities of psychological support for patients with lymphedema.



*Y. E. Kurtanova and A. V. Rovnaya during the report on the 43rd European Society of Lymphology Congress*

*On The 43rd European Society of Lymphology Congress in Stuttgart: Konechni L., Demekhova M., Makarov I., Kurtanova J., Feiskhanov A.*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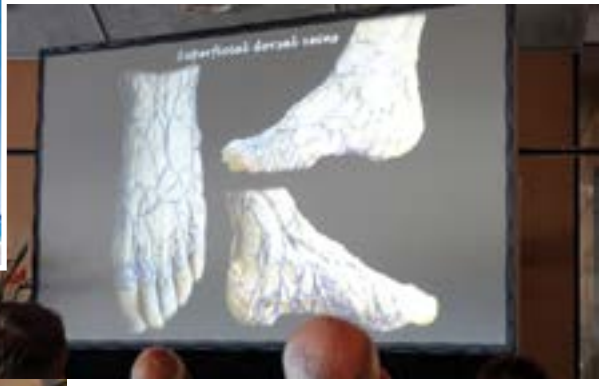
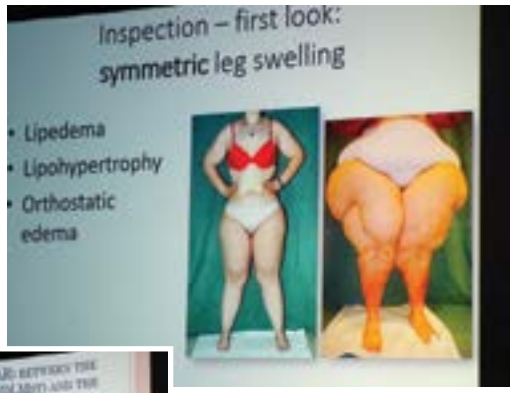
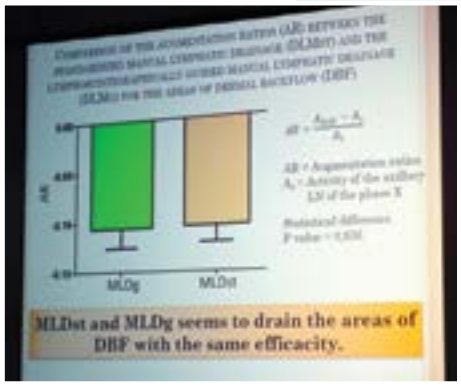
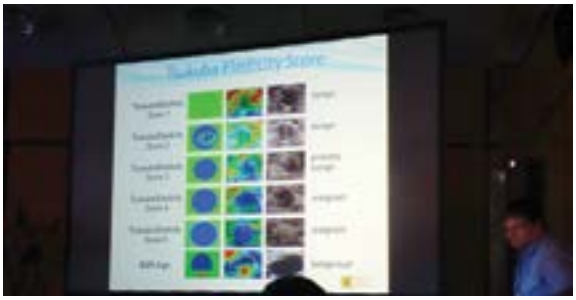
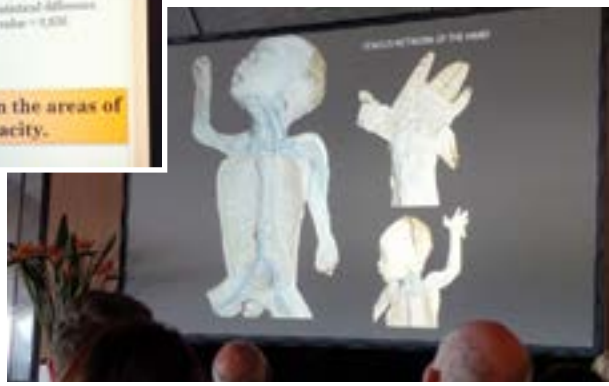

Lateral dorsal vein


Inspection – first look:
symmetric leg swelling
• Lipedema
• Lipohypertrophy
• Orthostatic edema


ACUTE LYMPHANGITIS
CLASSIFICACTION


MLDst and MLDg seems to drain the areas of DBF with the same efficacity.





# World Congress of Lymphology in Barcelona, Spain

## (26th World Congress of Lymphology)

*From 25 to 29 September 2017, Barcelona will host the International Congress of Lymphology.*

The Congress program is almost completely formed and is available on the official website of the Congress: www.lymphologycongress2017.com. The site is also available for registration.

# St. Petersburg Lymphology Forum

*St. Petersburg Lymphology Forum will be held on October 12-13, 2017 in the National Medical Research Center named after V.A. Almazov, at the address: 2 Akkuratova St., St. Petersburg.*

The forum's president is V.I. Konenkov, academician of the Russian Academy of Sciences, scientific director of the Institute of Clinical and Experimental Lymphology. E-mail for registration: conference@almazovcentre.ru. Abstracts are accepted by e-mail: lymph@almazovcentre.ru before September 1, 2017.





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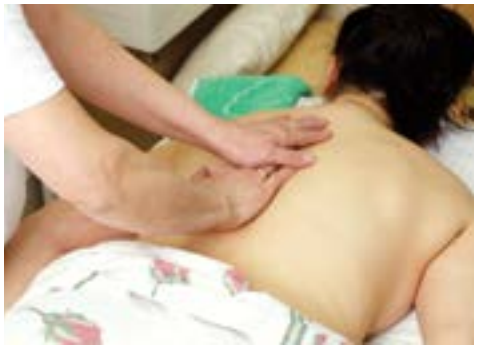
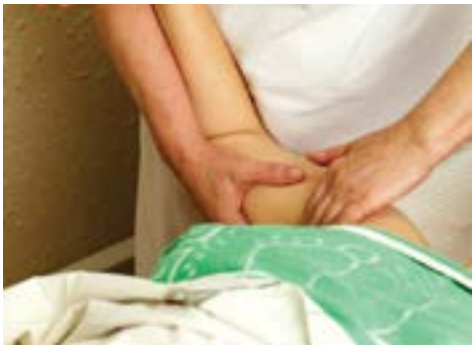
**Ivan Makarov, MD, author of 5 books about problems in treatment of lymphedema for doctors and patients, head medical and educational director of Scientific rehabilitation center of patients with lymphedema LYMPHA, the founder of the school for patients «LIFE WITH LYMPHEDEMA"**





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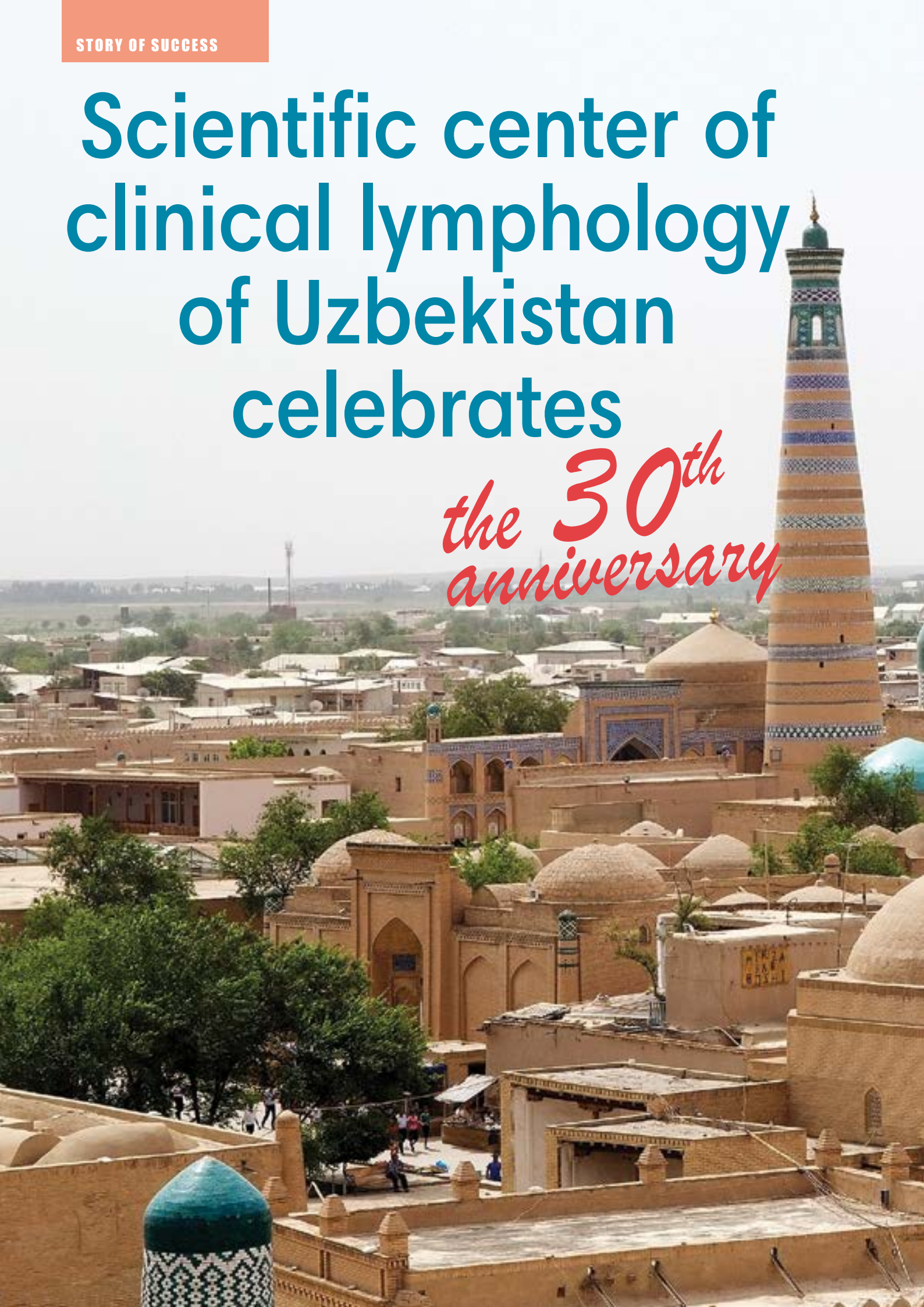
# Scientific center of clinical lymphology of Uzbekistan celebrates the 30th anniversary



Studies conducted over the past decades have revealed that the lymphatic system influences the development and progress of almost all typological pathological processes. This finding has fundamentally changed the attitude towards lymphology - a science that accounts four centuries of history. The discovered properties and possibilities of the lymphatic system have been adopted by clinicians, which allowed developing methods of influence on the pathology in a clinic through the lymphatic system.


The railway station in Andjijan

## The first scientific lymphological centers

With the accumulation of experimental, clinical experience, as well as the introduction of clinical methods of lymphatic therapy, the process of launching scientific centers started in such cities as Moscow, Zugres, Karaganda and others. In 1991, the Laboratory of Lymphology at the Siberian Department of the Academy of Medical Sciences of the USSR (Novosibirsk) was transformed into the Institute of Clinical and Experimental Lymphology under the guidance of academician Y.I. Borodin. One of the first centers was established in Andijan (Uzbekistan) in 1987. The Ministry of Health of the Republic of Uzbekistan created the Republic Center for Clinical Lymphology in Andijan to recognize more than 20 years of scientific research in the field of lymphology conducted by the Department of Hospital Surgery of the Andijan State Medical Institute, headed by professor S.U. Dzhumabaev.

## Features of lymphology in Uzbekistan

The 30-year activity of Uzbekistan's lymphologists can be roughly divided into 3 decades: the first and second decades are devoted to the fundamental experimental and clinical development of the foundations of practical lymphology. The third decade is characterized by the wide introduction of lymphatic therapy methods into practical public health, using standards and protocols of lymphatic therapy.

Nowadays, Uzbekistan has its own school of clinical lymphology. The developed methods are adapted to the peculiarities of the Republic of Uzbekistan with the primary residence of the population in rural areas and the developed system of the primary link of medicine.

The long-term operational activity of the lymphological center is inextricably linked with the names of such great lymphological scientists as B.V. Ognev, J.E. Vyrenkov, I.V. Yarema, V.M. Bujanov, R.T. Panchenkov, Y.I. Borodin, S.V. Lokhvitsky, Y.M. Levin, F.G. Nazirov, I.M. Baibekov, S.A. Dadaev, R.Y. Omirov and others.

## The contribution of Y.E. Vyrenkov

To commemorate Professor Y.E. Vyrenkov, an associate member of the Russian Academy of Natural Sciences, who recently retired from the world, I would like to note the invaluable assistance of this outstanding scientist in the training of personnel, organizing and establishing the Republic Center for Clinical Lymphology in Uzbekistan. His close scientific and friendship ties with the head of the Center - Professor S. Y. Jumabaev, allowed raising lymphology to a high scientific and practical level.

## Achievements in the sphere of lymphology in Uzbekistan

The new stage of reforming the healthcare system of our republic is characterized by the prioritization of new, high technology development at the level of regions and regional centers. It allows developing and introducing economical, universal and at the same time effective methods of lymphatic therapy into practical medicine. Researchers in our republic are working on the development of a number of fundamental directions in clinical lymphology.

**1. Surgery of the lymphatic system, thoracic duct**
The Scientific Specialized Center of Surgery named after academician V. Vahidov of the Ministry of Health of the Republic of Uzbekistan has initiated research activities by P.M. Khamidov, F.G. Nazyrov, V.K. Yuldashchev for the first time in the Republic. The obtained results allowed a qualitative improvement of the results of treating patients with liver and lymphedema pathology.

**2. Direct endolymphatic therapy**
The methods that were previously developed and improved have been introduced by the staff of the Tashkent Pediatric Medical Institute and the Andijan State Medical Institute in the treating of purulent-septic lesions of various organs. Methods for indications are used in surgical practice, they are effective in emergency medicine, in the treatment of pulmonary tuberculosis, oncological and a number of other diseases (S.A. Dadaev, L.B. Nugmanova, S.Y. Urakov, T.A Bobomuratova., B.M., A. Ashurmetov, I.A. Ahmedov, A.S. Saidkhanov, R.Y. Omirov, G.H. Khadzhimatov, F.S. Appazov and others).

**3. Indirect lymphatic (lymphotropic) therapy**
The first studies in the republic in the field of clinical lymphology were related to the substantiation of this method. Technical simplicity, combined with sufficient efficiency, makes it a method of choice for using in the emergency medical care system, in the primary health care system, in the stages of medical evacuation in emergency situations.

**4. Regional lymphatic therapy**
This is a priority development of the Republican Scientific Center of Clinical Lymphology of the Ministry of Health of the Republic of Uzbekistan. The method is characterized by high efficiency, selectivity and regulated impact on the pathological process with the use of small doses of medicines. More than 40 therapeutic methods used in all parts of practical public healthcare have been developed. The accumulated experimental material, clinical experience of applying this method to various organ and system diseases, at different levels of practical public healthcare allow us to evaluate the methods of regional lymphatic therapy not only as a disease treatment technology, but also as a method of injecting medicine into the body. To optimize the expenditure of material resources, reduce the risk of nosocomial infection, minimize the negative impact of medicine on the body, special attention should be paid to some characteristics of regional lymphatic therapy in comparison with traditional methods of administration. Those characteristics include a 1.5-2 fold decrease in medicine consumption, a 2-3 fold decrease in injections, reduction of treatment duration by 10-25% and lowing expenses for inpatient treatment by 25-40%.

## Modern achievements

Nowadays, clinical lymphology in Uzbekistan is characterized by the development of an independent scientific direction, recognized by specialists. It has been introduced into practical public health and resulted into the establishment of a lymphological service in the healthcare system of the republic.

For the first time in the history of practical public health, the staff of the center has developed and introduced a classification of lymphatic therapy. Employees and the Center partners have defensed 10 doctoral and 28 Ph.D. theses devoted to lymphology issues. Moreover, 12 inventions have been issued; more than 800 scientific works and 10 monographs have been published. In cooperation with scientists from the clinic in North Manchester (UK), a "Regional lymphatic therapy" guideline was published in English in 1998. The first handbook on clinical lymphology was published in 1999. Ten scientific conferences (the 2nd All-Union Congress of Lymphology and the 4th International one) were held, and from 1992 to 2014 a specialized scientific and practical journal had been issued. Since 2000, after the death of Professor S.U. Dzhumabaeva, the founder of clinical lymphology in Uzbekistan, this publication has become known as «Lymphology. Journal after the name of S.U. Jumabaev».

In 2001, the center was given the scientific status by the order of the Ministry of Health having only one department of clinical lymphology where more than 2000 doctors and 700 nurses learned different methods of lymphotherapy.

In 2002, the scientific and practical activities and technologies of the Republican Scientific Center for Clinical Lymphology, developed in the field of clinical lymphology, were highly appreciated by the International Fund of International Arch of Europe for Quality and Technology and nominated for the award of the International Arch of Europe for Quality and Technology».

*Prof. E.S. Dzhumabaev*
*Andijan Medical Institute (Uzbekistan)*

In the photo: the founders of clinical lymphology in Russia and Uzbekistan - Professor S.U. Djumabaev, associate member of AMS, Professor B.V. Ognev, associate member of RANS, Professor Y.E. Vyrenkov.

# ALCOHOL and LYMPHEDEMA

**Alcoholic beverages are beverages containing ethyl alcohol (ethanol). Depending on the concentration of ethanol and the number of drinks consumed, they have different effects on our body. But they do influence in any case. And our task is to find out what effect alcohol has on patients with lymphedema, and what should their decision be in the end - to drink or not to drink?**

**We are not going to consider such a disease as alcoholism here, but suggest talking about those cases from everyday life of almost every person, when people occasionally drink alcohol (for example, on holidays). In some countries and among some nationalities, the use of certain alcoholic beverages is a tradition. Therefore, patients with lymphedema need to know how alcohol affects the lymphatic system.**

**For people suffering from lymphedema, it is customary to make certain adjustments to their daily lives. And this is a common practice for patients with any chronic disease, otherwise it can worsen.**

**When you have lymphedema, any detail is important. This applies to food, water temperature for shower, and physical activity. Therefore, it makes sense to find out how alcohol affects the lymphatic system.**

## Alcohol and you

Alcohol begins to affect your body once you make the first sip. It is absorbed into the blood almost immediately through the mucous membrane of the stomach and then continues to be absorbed more slowly in the intestine.

After a few sips, you can feel the warmth all over your body. There is an emotional rise, an improvement in mood. That is how the body reacts increasing the release of serotonin and endorphins, which are responsible for exacerbation of emotions.

The physical sensation of warmth and relaxation, however, arises from the expansion of blood vessels. This process is called vasodilation. This increases the flow rate of lymph and the amount of fluid accumulating in the tissues of your body.

Alcohol has a diuretic effect, stimulates the kidneys to secrete more fluid. In combination with the expansion of blood vessels, this carries some problems for patients with lymphedema, as far as, as noted above, the tissues of our body receive excess lymph. A defective lymphatic system cannot cope with the removal of excess fluid, and this can lead to swelling and a feeling of heaviness in the affected part of the body, especially after drinking at night.

Drinking a lot of alcohol can also weaken your immune system, which in turn can increase the risk of such a complication of lymphedema, like erysipelas. According to the National Institute for Alcohol and Alcohol Abuse (USA): even a single excessive use of alcohol to intoxication can slow the body's ability to produce cytokines, which are substances produced by white blood cells, to protect the body from infections. This can reduce your ability to reflect infections for 24 hours. This is especially dangerous for patients with lymphedema, who already

have weakened immune system.

The time of feeling the effect of alcohol and its consequences depends on the individual characteristics of the organism such as sex, physiology, and genetics. All people are different: someone with a high alcohol content in the blood can seem much more sober than he really is. Keep this in mind to know how alcohol affects you, especially if you have lymphedema.

## What to do?

It is obvious that the most effective way to avoid the negative impact of alcohol on your body is to completely give up drinking it, or significantly reduce its amount.

**But, if you still continue drinking alcohol, here are some tips to help reduce the risk of consequences:**

✓ Drink more water. Remember that alcohol has a diuretic effect, that is, you lose more fluid than you consume. Therefore, during a celebration drink more water, it will saturate your body with moisture and help to avoid dehydration, not to mention that this is the right remedy for a hangover.

✓ Self-massage. If you or your relatives are trained in the correct technique of manual lymph drainage, you may want to use it after drinking alcoholic beverages. It will not only reduce edema, but also help to remove toxins quickly.

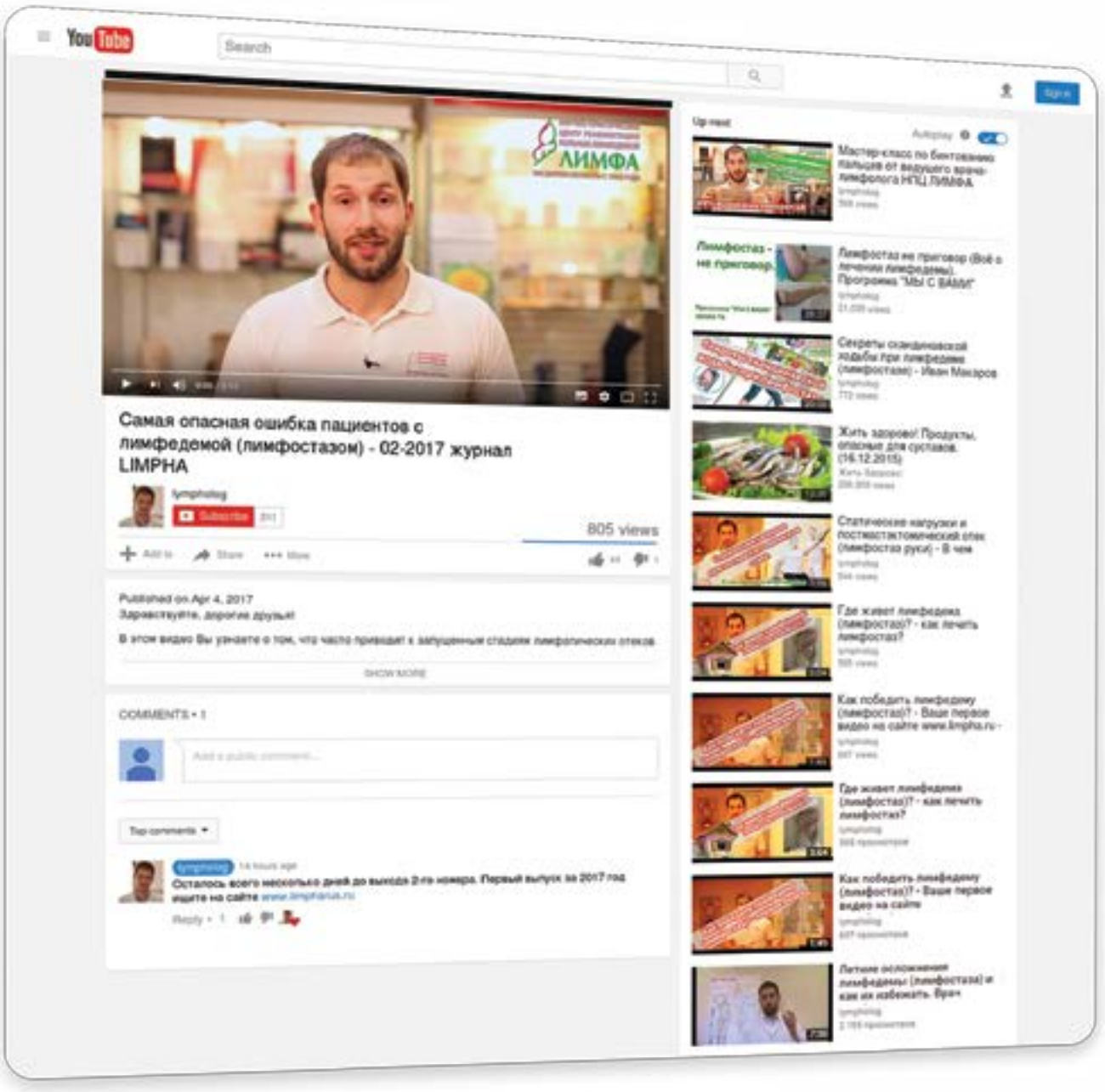
✓ Raising the affected limb to an elevated position can also help you avoid increasing swelling.

✓ Compression jersey. Of course, you should always wear it, because it provides additional support to your lymphatic system. With frequent use of alcohol, knitwear is not always able to retain edema; nevertheless, it will not allow edema to increase significantly.

✓ Be attentive to your body. Watch how your body reacts to alcohol: if your edema are worsening, if the feeling of heaviness in the affected limb increases, if you experience pain or discomfort. Depending on this, you may want to change your mind about drinking alcohol. In addition, your body will thank you for it!

Drinking or not drinking alcoholic beverages is a personal choice. However, any choice should be based on knowledge. Now you know what happens to your body when you drink alcohol, and how it affects your lymphedema. Therefore, you will be able to make your choice basing on awareness.

*Origin: https://thelymphielife.com/2017/06/02/bottomsup-the-effects-of-alcohol-on-lymphedema/*

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**This is the opportunity to meet with patients daily struggling with lymphedema, learn their secret of success and how they cope with their disease! This information about the leading doctors in the area of lymphology and clinics that help patients with diseases of the lymphatic system.**

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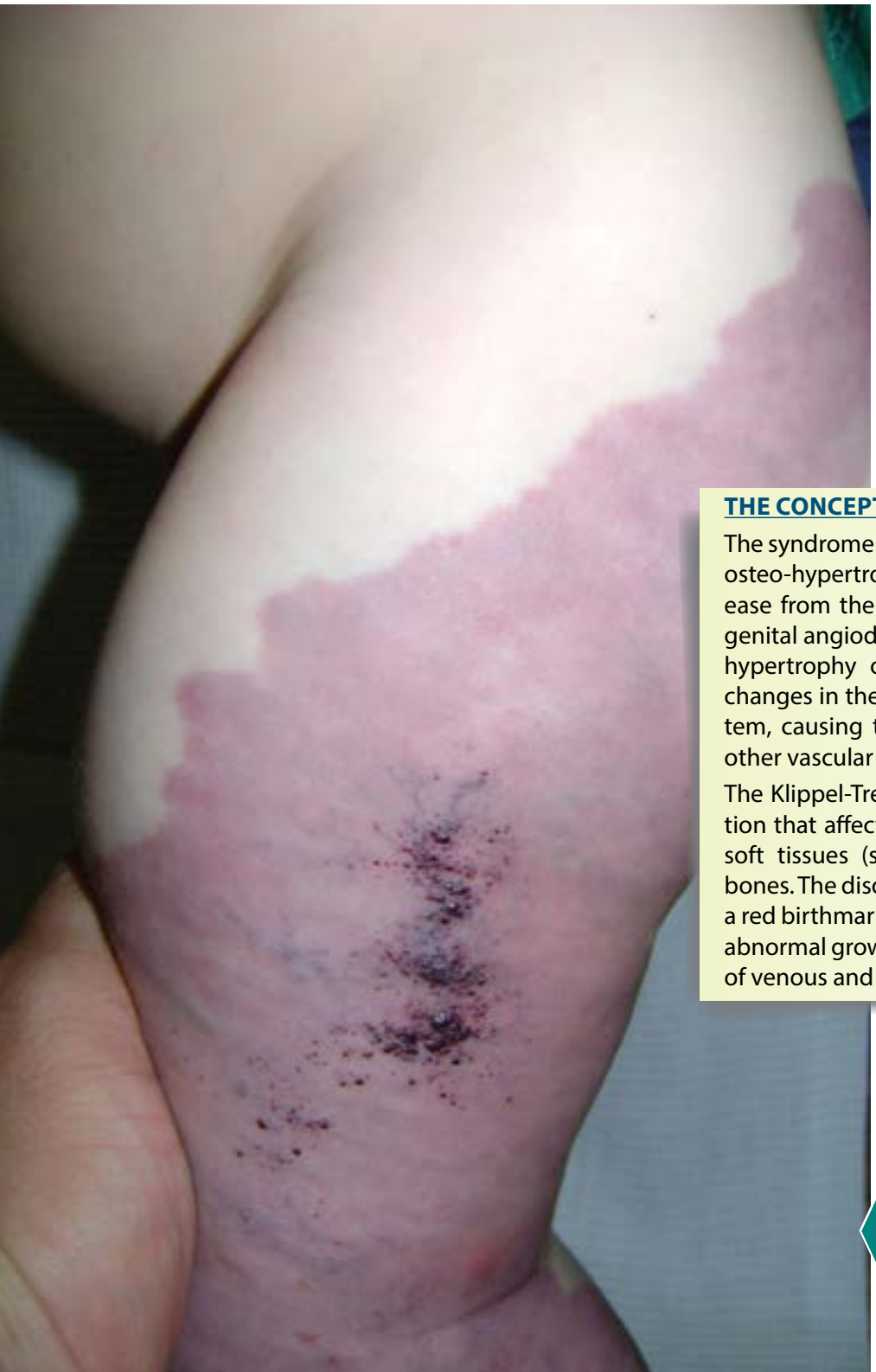
# Lymphedema

## as a complication of the Klippel-Trenaunay-Weber syndrome



*Bykova E.A., MD*

**# Keywords:**
Klippel-Trenaunay-Weber syndrome, lymphedema, children

### THE CONCEPT

The syndrome of Klippel-Trenaunay-Weber (varicose osteo-hypertrophic nevus) is a rare congenital disease from the group of phakomatoses, it is a congenital angiodysplasia characterized by asymmetric hypertrophy of the extremities, the presence of changes in the vessels, including the lymphatic system, causing the edema of the affected limb and other vascular complications [1 , 2].

The Klippel-Trenaunay-Weber Syndrome is a condition that affects the development of blood vessels, soft tissues (such as the skin and muscles) and bones. The disorder has three characteristic features: a red birthmark, called a port stain or a wine spot, an abnormal growth of soft tissues and bones and vices of venous and lymphatic vessels.

The wine spot - characteristic symptom of Klippel-Trenaunay-Weber syndrome

There are several disputes about the terminology. This condition was first described by French physicians Maurice Klippel and Paul Trenaunay in 1900 and named naevus vasculosus osteohypertrophicus. Frederick Parks Weber described the cases in 1907 and 1918, which were similar, but not identical to those described by Klippel and Trenaunay .

In 1965, Lindenauer suggested that in the presence of an arteriovenous fistula the term «Parkes-Weber Syndrome» should be used instead of it [5]. More recently, Cohen has supported a division that retains these terms distinctly [6].

In ICD-10, the term «Klippel-Trenaunay-Weber Syndrome» is currently used.

## Epidemiology

The Klippel-Trenaunay syndrome is a fairly rare disease, according to some estimates, it occurs in 1 out of 30,000 newborns [4,5]. It is believed that the Klippel-Trenaunay syndrome affects at least 1 in 100,000 people worldwide. This syndrome is registered with equal frequency, both among men and women, and the prevalence of the disease does not depend on race. [1,2,6]. The majority of patients have other congenital anomalies, in particular the development of hip dysplasia and syndactyly, occurs in 30% of patients [4].

The first symptoms of the disease appear even in childhood, but most often the only sign at an early age is lymphedema and the appearance in the area of the affected limb, the so-called «wine spot,» the capillary malformation (anomaly) of the nevus type. As the disease progresses, the length of the limb increases due to the development of hypertrophy of the bone tissue, as well as the increase in the affected limb in the volume, due to a significant increase in muscle tissue. The defeat of lymphatic vessels is manifested in such patients by the development of lymphedema, which, together with an edematous syndrome, causes the appearance of dry skin, papillomas, hyperpigmentation, is the reason for the decrease of local immunity and the development of infectious processes, including erysipelas.

Diagnosis of the disease is established on the basis of the presence of a triad of symptoms, which includes lymphedema, partial gigantism and the presence of capillary malformation as a «wine spot» of different localization. However, only 63.5% of patients show a complete triad of the disease. For this reason, the diagnosis in such patients can be established with two of the three triad elements [3,20].

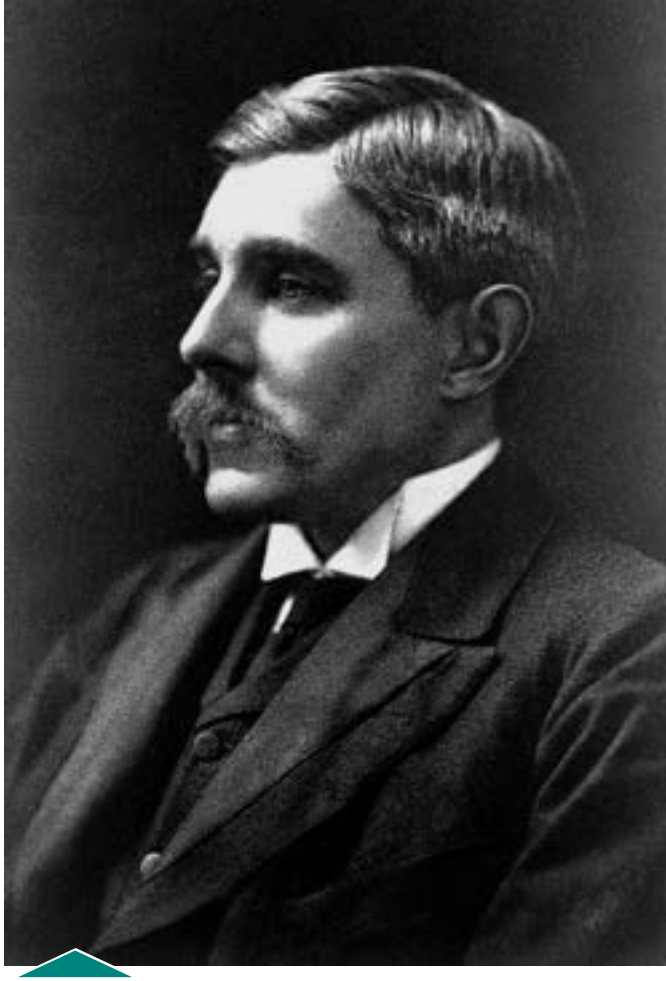
*The Klippel-Trenaunay syndrome occurs in 1 out of 30,000 newborns*

The disease is often accompanied by the occurrence of bleeding, in particular, there may be bleeding in the gastrointestinal tract. The source, in this case, most often, is the varicose veins of the esophagus, caused by portal hypertension, which in turn arises as a result of cavernous transformation or hypoplasia of the portal vein [7,8]. Bleeding associated with intestinal lesions most often occurs in the distal parts of the colon [8,9]. Transient thrombophlebitis, thrombocytopenia, severe anemia, congestive heart failure, and often a complication of the course of the disease in these patients [9].

## Mechanism of development

Venous malformations in this syndrome often manifest as preservation of embryonic veins, for example, the sciatic vein in such patients does not undergo regression, but the most common finding is embryonic lateral marginal vein, which occurs in 68-80% of patients [10]. This vein extends over the outside of the foot and rises up the outside of the lower limb. The vein usually has a thickened wall, is located subcutaneously and is incompetent all along, due to the absence of venous valves. Drainage in this case is carried out in the lateral branch of the femoral vein or in the internal iliac vein [10,11]. Lymphatic malformations cause the development of primary lymphedema and occur in 70% of cases [11]. Also, a frequent sign of lymphatic malformation in this category of patients is the presence of so-called lymphangiomas - benign tumors of the lymphatic system. In 50% of cases, lymphangioma can be detected at birth. In 75% of cases, lymphangiomas are located on the head and neck (61%) or the axillary region (13%). The remaining 25% are distributed along the trunk (11%), in the limb region, (11%), mediastinum (1%), abdominal cavity and genitalia (3%) [2].

The reason for this syndrome is that the deep trunk veins become impassable under certain conditions. Scientists have described numerous congenital arteriovenous canals with abnormally dilated veins, penetrating muscular, bone and skin tissues. Since the anomaly covers large areas of skin integument, the subcutaneous veins turn out to be hypertrophic enlarged. As a consequence, the skeleton and soft tissues in such areas are subject to abnormal changes: they increase and lengthen.

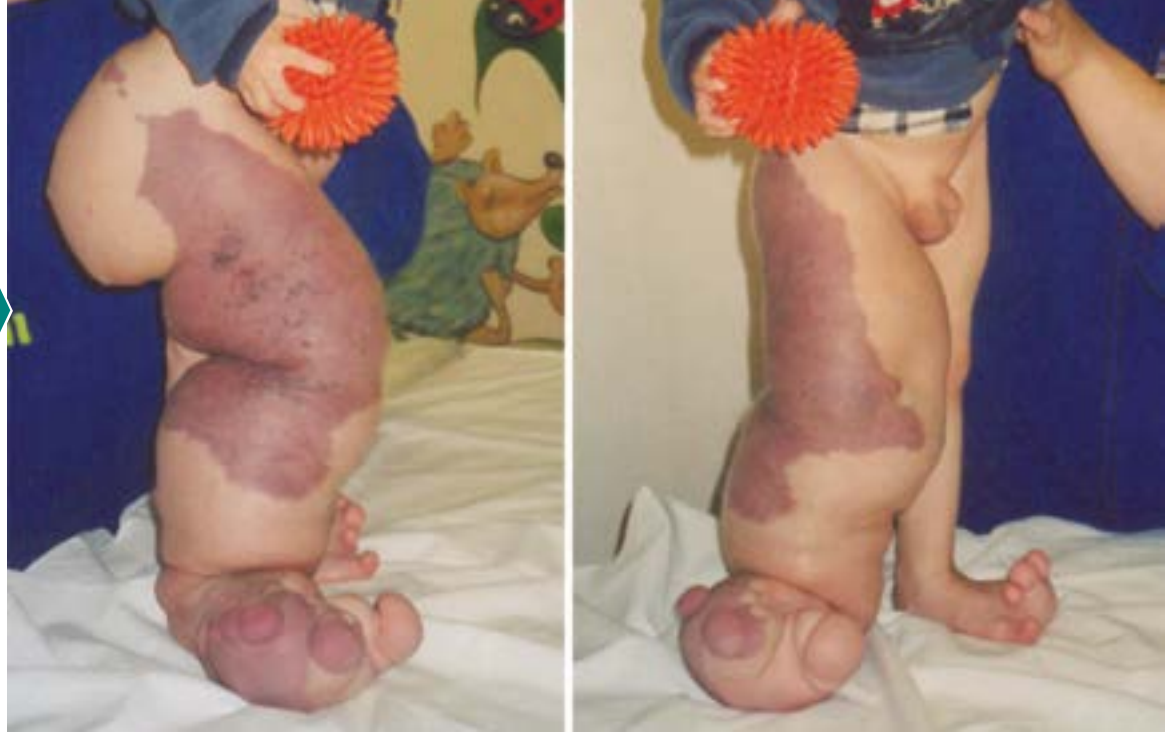


Frederick Parks Weber (1863-1962)

*Diagnosis of the disease is established on the basis of the presence of a triad of symptoms, which includes lymphedema, partial gigantism and the presence of capillary malformation as a "wine spot" of different localization*

### GENETICS

The Klippel-Trenaunay-Weber Syndrome may be caused by mutations in the PIK3CA gene. This gene provides instructions for obtaining the p110 alpha (p110α) protein, which is one part (subunit) of an enzyme called phosphatidylinositol-3-kinase (PI3K). PI3K plays a role in chemical signal transmission, which is important for Klippel-Trenaunay-Weber Syndrome may be caused by mutations in the PIK3CA gene. This gene provides instructions for obtaining the p110 alpha (p110α) protein, which is one part (subunit) of an enzyme called phosphatidylinositol-3-kinase (PI3K). PI3K plays a role in chemical signaling, which is important for many cellular activities, including cell growth and division (proliferation), cell movement (migration), and cell survival. These functions make PI3K important for the development of tissues through-



The child with the syndrome of Klippel-Trenaunay-Weber

out the body.

Mutations of the PIK3CA gene associated with the Klippel-Trenaunay-Weber syndrome change the protein p110α. The modified subunit makes PI3K abnormally active, which allows the cells to grow and divide continuously. Increased cell proliferation leads to abnormal growth of bones, soft tissues and blood vessels.

The Klippel-Trenaunay-Weber Syndrome is one of several proliferation syndromes, including the melanodermal-capillary malformation syndrome, which is caused by mutations in the PIK3CA gene. Together, these conditions are known as the fouling spectrum associated with the PIK3CA (PROS).

Because not all of the Klippel-Trenaunay-Weber Syndrome have a mutation in the PIK3CA gene, it is possible that mutations in unidentified genes can also cause this condition [21].

The syndrome of Klippel-Trenaunay-Weber is almost always sporadic, and this means that it occurs in people who do not have a history of frustration in their family. Studies show that this condition is the result of genetic mutations that are not inherited. These genetic changes, called somatic mutations, occur randomly, in one cell, in the early stages of development, before birth. As the cells continue to divide during development, the cells emerging from the first abnormal cell will have a mutation, and the other cells will not. This mixture of cells with a genetic mutation and without it is known as mosaicism [6].

There is some evidence that this may be due to translocation to t (8; 14) (q22.3; q13) [21].

## ETIOLOGY

The etiology of the disease is still unknown, there are several theories, according to one of them, a hereditary character of the disease is assumed, in which the dominant gene responsible for the development of angiopathy and partial gigantism is inherited. For a long time, there was a suggestion of deep vein obstruction and, as a consequence, an increase in venous pressure in such patients, but recent studies have shown that most patients have unobstructed venous drainage. Currently, most authors tend to believe that the Klippel-Trenaunay-Weber syndrome is caused by sporadic damage to the myoblasts mesoderm, which occurs during intrauterine development, which is responsible for the development of microscopic arteriovenous communications in the limb bud.

In a series of 252 patients in the Mayo Clinic, 63% of patients had all three symptoms, and 37% had 2 of 3 symptoms. Wine spots were observed in 98% of patients, varicose veins or venous malformations - in 72%, and limb hypertrophy - in 67%. Atypical veins, including lateral veins and persistent sciatic vein, were present in 72% of patients. Finally, deep venous anomalies included aneurysmal dilatation, hypoplasia, aplasia, and missing or incompetent valves.

***The Klippel-Trenaunay-Weber Syndrome is a condition that affects the development of blood vessels, soft tissues (such as the skin and muscles) and bones***

Diagnostics. As a follow-up, an ultrasound examination of soft tissues is recommended to assess the thickness of the muscular layer, as well as radiographic examination of the limb bones to reveal partial gigantism. All patients are recommended to perform ultrasound examination of the arteries and veins of the lower extremities to assess the degree of venous system damage and exclude thrombotic complications.

***Angiomas - "wine spots", most often they are on one leg and cover a large area***

Magnetic resonance lymphangiography (MRL) with dimeglumin gamatronic as a contrast showed that 31 of 32 patients had abnormalities of lymphatic vessels and / or lymph nodes, including hyperplasia (11/31), hypoplasia or aplasia (20/31) of lymphatic vessels and lymphedema (31/31) of affected extremities. In patients with the Klippel-Trenone-Weber syndrome, there was a high combined lesion of the lymphatic system and veins in the affected limbs [10].

**DIFFERENTIAL DIAGNOSIS** is carried out with:

- Dermatological manifestations of the protein syndrome
- Muffucci Syndrome

Thrombus venous veins in the Klippel-Trenaunay-Weber Syndrome should be distinguished from an inflammatory pelvic disease. The syndrome of Klippel-Trenaunay-Weber can be associated with arterial and venous malformations in the lower segment of the uterus [4]. The Sturge-Weber syndrome in combination with the Klippel-Trenaunay-Weber syndrome has been described in more than one patient [6].

## SYMPTOMATOLOGY

Cutaneous vascular malformation manifests itself at birth, but venous varicosity and hypertrophy of the limbs may not be obvious initially.

Although the causes and processes associated with the Klippel-Trenaunay-Weber syndrome are poorly understood, a congenital defect is diagnosed by the combination of these symptoms (often about 1/4 of the body, although in some

cases more or less affected tissue may be present).

The following symptoms are classic symptoms of the disease [1]:

1. Angiomas - vascular spots («wine spots»). Most often they are on one leg and cover a large area. Color angiomas can be different: from light to dark purple hue. Under the influence of physical factors (friction, impact), the skin in the area of «birthmarks» is easily damaged, bleeding appears.

2. Varicose veins of the superficial veins. Clinical manifestations of this trait are the tortuosity, thickening, enlargement and soreness of the vessels of the lower limb.

3. Hypertrophy of the affected leg. Because of the defeat of deep veins and their proliferation, the limb increases in diameter. In some cases, the bone system is included in the process. Then the affected leg can be longer than the healthy limb, which can lead to local gigantism or contraction.

4. Improperly developed lymphatic system.

Note: in some cases, patients may be present without «wine stains». Such cases are very rare and can be classified as an atypical syndrome of Klippel-Trenaunay-Weber.

Most people with the Klippel-Trenaunay-Weber Syndrome are born with a wine spot. This type of birthmark is caused by a tumor of small blood vessels near the surface of the skin. Wine spots are usually flat and can range from pale pink to maroon. In people with the Klippel-Trenaunay-Weber Syndrome, a wine spot usually covers part of one limb. Affected area can become lighter or darker with age. Sometimes the wine spots produce small red blisters that open easily and bleed.

Deformities of the veins - the third important feature of the Klippel-Trenaunay-Weber syndrome. These abnormalities include varicose veins, which are swollen and twisted veins near the surface of the skin, and which often cause pain.

The syndrome of Klippel-Trenaunay-Weber can affect blood vessels, lymphatic vessels, or both. Most often this is their combination. Those who have venous complications are subject to a more severe lifestyle, due to increased pain and complications [3].

The first manifestations of the Klippel-Trenaunay-Weber syndrome are noticeable already in the period of newborn. In rare cases, they occur later - in childhood.

Sometimes one of the signs of the disease is absent or does not appear externally (for example, varicose veins). In some cases, the pathology progresses rapidly, and the lesion passes from the extremities to the trunk and upper humeral girdle.

There is an elongation or shortening of the length of the affected limb, which can lead to problems with walking. Varicose veins show up on the extremities, more often on the lower leg and hips. It can affect not only superficial, but deep veins, which increases the risk of thrombophlebitis, thrombosis and can cause pulmonary embolism. Patients often experience muscle spasms and complain of pain when walking. Paresthesias and ulcers on the skin may also appear. When involved in the process of the gastrointestinal tract or internal organs, episodes of bleeding (internal, rectal, hematuria) are noted. When there is lymphostasis, there is a marked edema of the limb, the appearance of pain sensations [3,15].

*Most people with the Klippel-Trenaunay-Weber syndrome are born with a wine spot*

In a study of 252 patients in the Mayo Clinic in Minnesota, 63% of patients had all three symptoms, and 37% had 2 of 3 symptoms. Port wine stains were observed in 98% of patients, varicosities or venous malformations in 72%, and hypertrophy of the extremities in 67%. Atypical veins, including lateral veins and persistent sciatic vein, were present in 72% of patients. Finally, deep venous anomalies included aneurysmal dilatation, hypoplasia, aplasia, and missing or incompetent valves [13].

Arteriovenous fistulas, a feature that distinguishes the Klippel-Trenaunay-Weber Syndrome from Parkes-Weber syndrome, are rarely found in the affected limb. If they are present, they can sometimes be palpated as pulsating mass, trembling or fetus during physical examination [5,12].

Hyperthermia and the positive sign of Branham (bradycardia with compression on the artery closest to the blemish) are also indicators of arteriovenous malformation.

Hypertrophy of the limbs may be secondary to the increased length (bone lesion) and / or increased girth (involvement of soft tissues). Hypertrophy can be evaluated at birth. This usually progresses during the first years of life. A greater degree of hypertrophy can be observed in patients with coexisting arteriovenous malformation. Although lymphedema is also observed in patients, true hypertrophy of affected soft tissues is present [5,25].

*Sclerotherapy involves injecting a chemical into abnormal veins to cause thickening and blockage of target vessels*

Up to 12 cm of discrepancies have been reported. Sometimes the involved limb can be atrophied, rather than hypertrophied.

Other features include lymphatic obstruction, spina bifida, hypospadias, polydactyly, syndactyly, hyperhidrosis, hypertrichosis, paresthesia, decalcification of the involved bones, chronic venous insufficiency, stasis dermatitis, poor wound healing, ulceration, thrombosis, angiosarcoma and emboli. [17, 18]

Brain anomalies include hemorrhage, infarction, venous malformations, arteriovenous malformations, cavernous, aneurysm, hydrocephalus, vascular plexuses of anomalies, atrophy, calcification, cortical dysplasia and convulsions. [13] Pulmonary embolisms secondary to thrombosis of the venous extremities are a risk in patients with this syndrome. [14] Cerebral infarcts are rare, as are brain tumors.

Surgical examination demonstrated atresia and agenesis of deep veins, compression due to fibrous bands, aberrant arteries, abnormal muscles or venous vaginas. Varicose veins can remain stable in size or gradually expand. Usually they report pain and lymphedema. These symptoms may worsen during pregnancy.

### COMPLICATIONS

Hypertrophy of the limb can lead to a subsequent scoliosis of the spine, gait irregularities and compromise functions. Patients with the Klippel-Trenaunay-Weber Syndrome tend to develop degenerative joint disease at an early age [10].

## Treatment

The treatment of the Klippel-Trenaunay-Weber syndrome is conservative and symptomatic. When managing these patients, an individual approach is necessary. For the treatment of capillary malformations, for cosmetic purposes, sclerotherapy and laser therapy are used.

*The treatment of the Klippel-Trenaunay-Weber syndrome is conservative and symptomatic*

Currently, not infrequently, these patients undergo various surgical interventions to reduce soft tissue edema, as well as phlebectomy due to angiovenous malformation [14]. Surgical treatment, often not only does not bring the desired result, but can worsen the patient's condition. Thus, the development of fibrotic and sclerotic changes in the postoperative period may lead to an increase in edema, the development of trophic changes in the skin of the affected limb. In connection with this, the surgical intervention in the affected limbs, in these patients, is recommended only for emergency indications [14]. In the treatment of edematous syndrome in this disease is currently recommended to conduct con-

servative therapy, which includes the procedure of manual lymphatic drainage massage [15].

Compression clothing is indicated for chronic venous insufficiency, lymphedema, recurrent cellulitis and occasional bleeding from capillary or venous limb developmental disorders. Squeezing clothing can also protect the limbs from injury. Intermittent pneumatic compression pumps can also benefit. However, in some patients with absent or hypoplastic deep venous systems, elastic compression may increase venous congestion and cause discomfort.

Some researchers indicate that pain in treatment can be a very important aspect of patient care from the Klippel-Trenaunay-Weber and recommend that such a contingent of patients be referred to a pain clinic and / or an interdisciplinary group comprising a pain specialist [3].

It is reported that radiotherapy helps in some cases of the Klippel-Trenaunay-Weber syndrome. Radiation can help cause regression of hemangiomas. However, the results can develop slowly [1,16].

Cytoreduction or cytoreductive surgery has been the most widely used treatment for the syndrome and has been used for decades. Progress has been made in this method over the past two decades, but it is still an invasive procedure and has many complications. Since there are currently other options for patients with the Klippel-Trenaunay-Weber Syndrome, this method is usually used as a last resort.

According to the Mayo Clinic (Minnesota), which reported the largest experience in the treatment of the Klippel-Trenaunay-Weber syndrome with extensive surgical interventions. For 39 years in the Mayo Clinic, a surgical department was performed and 252 consecutive cases of the Klippel-Trenaunay-Weber syndrome were evaluated, of which only 145 (57.5%) could be treated by primary surgical operations. The immediate success of treatment for varicose veins was only 40%, elimination of vascular malformation was possible by 60%, removal operations by 65%, correction of bone deformation and correction of limb length (epiphiseosis) had 90% success. All procedures demonstrated a high incidence of relapse in follow-up. Clinical studies in Mayo suggest that the initial surgical treatment of the Klippel-Trenaunay-Weber syndrome has limitations, and it is necessary to develop non-surgical approaches to ensure a better quality of life in these patients. A major operation, including amputation operations, does not appear to be of any use on a long-term basis.

Sclerotherapy is the treatment of specific veins and vascular malformations in the affected area. The procedure involves injecting a chemical into abnormal veins to cause thickening and blockage of target vessels. Such treatment can resume normal blood flow. This is a non-surgical medical procedure and not so aggressive. Sclerotherapy with the use of ultrasound guidance is the newest treatment technology, which can potentially close many large vascular malformations. [22,23]. Cytoreductive operations can lead to large deformations and have a high potential for repetition and damage to the nerves [14].

Compression therapy has been increasingly used over the past ten years. The biggest problem with the Klippel-Trenaunay-Weber syndrome is that the blood flow and / or lymphatic flow can be difficult and will be combined in the affected area. This can cause pain, swelling, inflammation, and in some cases even ulceration and infection. Among older children and adults, compression clothing can be used to alleviate almost all of them, and in combination with increased lesion area and proper management can lead to a patient's life comfort without any operation. Compressed clothing items are also used recently after discharge from the hospital in order to preserve the results of the procedure. For the early treatment of infants and toddlers with Klippel-Trenaunay-Weber syndrome, the use of special clothing for compression is impractical due to the rate of growth. When children can take advantage of compression therapy, you can use wraps and lymphatic massage. Although compression clothing or therapy is not suitable for everyone, they are relatively cheap (compared to surgical intervention) and have few side effects. Possible side effects include a small risk that liquids can simply be moved to an undesirable place (for example, in the groin) or that compression therapy additionally prevents blood circulation to the affected limbs. Other procedures are also available, including other surgical procedures and massage. Currently, however, many of the symptoms can be cured, but as for the Klippel-Trenaunay-Weber syndrome, the search for different methods and approaches to treatment continues [11].

***Compression therapy has been increasingly used over the past ten years in patients with the Klippel-Trenaunay-Weber syndrome***

Endovascular laser therapy of the large saphenous vein receives support for the treatment of varicose veins in the general public and in patients with the syndrome of Klippel-Trenaunay-Weber [16]. This therapy was used alone and in combination with other surgical interventions. This is a new and minimally invasive approach for the treatment of certain varicosities.

In the Klippel-Trenaunay-Weber syndrome, laser treatment of a hemangioma can be effective for lightening the color of the wine spot. At present, a pulsed dye laser with a flash lamp is the method of choice for vascular lesions. Laser therapy is also indicated in case of ulceration. Ulceration of hemangiomas can be painful and impair functional abilities. When treating the laser, ulcers often heal faster. Laser treatment is most effective at early implementation, since it can improve the long-term appearance of the wine spot and, thereby, improve the function. As a rule, many procedures are required to achieve the desired effect. Laser treatment only helps with the surface component of the hemangioma [16].

## Prognosis

The syndrome Klippel-Trenaunay-Weber has rather favorable prognosis for life, if timely to conduct surgical treatment. The exact prognosis of the course of the disease is difficult, due to multiple concomitant diseases and congenital malformations in these patients. According to various data, in 8-12% of cases in patients with the Klippel-Trenaunay-Weber syndrome there is a venous thrombosis of different localization, which in 1-2% of cases, for example, with the development of pulmonary embolism, can lead to death [9]. Pulmonary embolism can occur in 10% of patients, especially after surgery. Bleeding, accompanied by the development of severe anemia, decompensation of heart failure may also worsen the prognosis in such patients [2]

Violations in the hand or foot in the Klippel-Trenaunay-Weber Syndrome can predict the presence of abnormalities in the deep venous system. The mortality in the Klippel-Trenaunay-Weber syndrome and the Parkes-Weber syndrome according to different researchers is 1% [16].

Patients with the Klippel-Trenaunay-Weber Syndrome should be monitored at least once a year and more often, according to the indications.

***The syndrome Klippel-Trenaunay-Weber has rather favorable prognosis for life, if timely to conduct the right treatment***

Clinically you can observe a stable course of the disease. Klippel-Trenaunay-Weber Syndrome is not always a static process of the disease and can progress.

# Results and discussion

We present the data of three patients observed in the SPC «LIMPHA» with the syndrome of Klippel-Trenaunay-Weber. All patients have a lesion of one lower limb, a prominent lymphedema attracts attention during examination, the affected limb is hypertrophied due to the phenomena of partial gigantism, there are capillary malformations in the form of a «wine stain» or «port spots», as well as pale discharge spots.

Patient Z., 4 years. The child from the first pregnancy against a background of toxicosis, during pregnancy, the mother repeatedly diagnosed threats of abortion in the early stages, anemia, late gestosis of mild degree, cholestatic hepatosis of pregnant women. At birth, weight 3750, length 52 cm. According to Apgar scale 8/8 b.

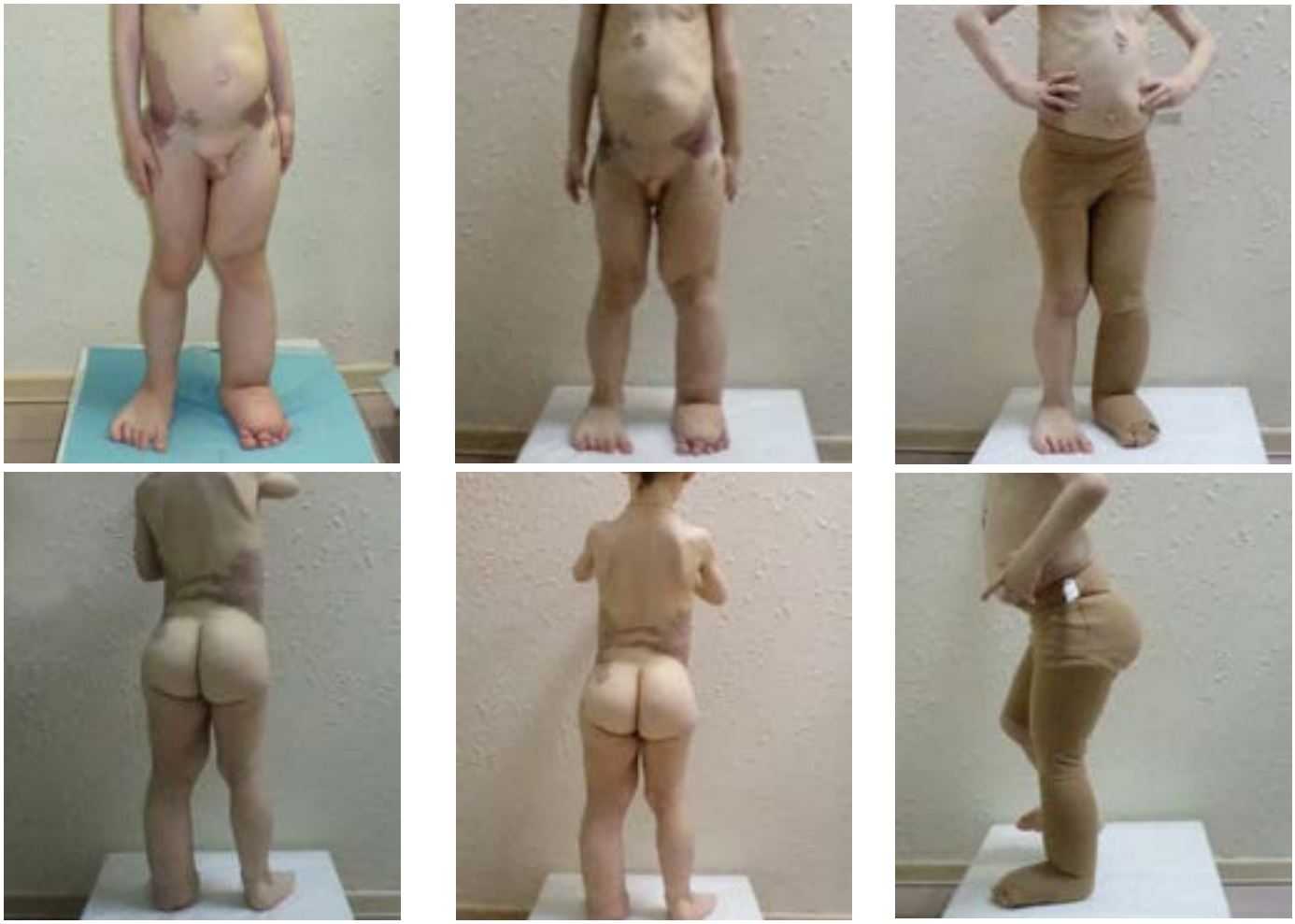
From an anamnesis, according to the mother of the patient, it is known that the first edema of the left lower extremity was detected by ultrasound examination at 32 weeks of gestation. He was born with an edema of the left lower extremity, expressed by extensive angiovenous dysplasia in the region of the back, the lateral surfaces of the trunk. Also in the history of frequent erysipelas, recurrent, usually 2 times a year.

It is observed in the SPC «LIMPHA» from 2013, with the diagnosis: Syndrome Klippel-Trenaunay-Weber. Congenital lymphedema of the left lower extremity of III degree, sporadic form complicated by erysipelas (last in August 2015). Partial gigantism of the right foot.

When examining the patient, there was an increase in the volume of the left lower limb due to a significant edema of soft tissues, muscle and bone hypertrophy. Edema is a moderate dense, deforming contour of the lower limb. The symptom of the Stemmer is positive. Lymph nodes are not enlarged. The temperature and sensitivity of the skin are not disturbed, D = S. There was extensive capillary angiodysplasia of the antero-lateral surface of the abdominal wall and the lumbosacral region.

Passed 4 courses of complex decongestive therapy with a positive effect in the form of soft tissue softening, decrease of the left lower extremity in the volume. During the 4th course, the decrease in the left lower limb in the volume was from -2.5 cm to -6.0 cm. Constantly using compression knitwear of flat knit by individual measures: a stocking of the 2nd class of compression.

**Dynamics on the background of treatment (4 courses of complex decongestive therapy - April 2016)**



Before treatment | After treatment | In compression garment

Patient S., 9 years old. The child from 3rd pregnancy, proceeded with the threat of abortion at 9 weeks - polyhydramnios. 2nd childbirth at 39 weeks. Weight at birth 3700 g, length 52 cm. Perinatal damage to the central nervous system of hypoxic-ischemic origin, moderate degree, hip dysplasia was diagnosed.

At birth, there was an increase in the volume of the right lower limb, gluteal, inguinal areas to the right and vascular drain spots in the same areas. In the future, with the growth of the child, a proportional increase in the spot and the right lower limb. Was examined in the interdisciplinary center of vascular anomalies (Regensburg, Germany), where a combined capillary lymphovenous malformation with accompanying hyperplasia of the right lower quadrant was performed, radiofrequency ablation and sclerosing of the large vena cava were performed.

It is observed in the SPC «LIMPHA» from 2016, with the diagnosis of the Klippel-Trenaunay-Weber syndrome, combined capillary lymphovenous malformation of the vessels of the right lower limb. Chronic lymphovenous insufficiency of the 2nd degree. Partial hypertrophy of the right lower limb.

Upon admission, the moderate increase in the right lower limb, the right gluteal, inguinal regions due to swelling of soft tissues and muscle hypertrophy attracted attention. Edema moderately tight throughout the limb. The skin is pale pink, over the entire surface of the skin of the right iliac region, the right buttock, the right hip and lower leg, multiple discharge pale spots of capillary angiodysplasia. The symptom of the Stemmer is positive on the right.

She received 1 course of complex decongestive therapy, against the background of treatment there was a decrease in the right lower limb in the volume from 2,1 cm to 4,4 cm. Constantly wears compression garment of flat knitwear, manufactured by individual measures.

**Dynamics on the background of treatment**
**(1 course of complex physical anti-therapeutics in 2016)**

**Before treatment**

**After treatment**

**In compression garment**

Patient S., 9 years old. A child from 1st pregnancy, in the first and second trimester of pregnancy, the mother had toxicosis, anemia, frequent acute respiratory viral infections, with fever, pronounced intoxication at 4 months. In the third trimester of pregnancy, the mother diagnosed with exacerbation of chronic pyelonephritis, severe gestosis, complicated by pulmonary edema. Childbirth operative. The condition of the child at birth is severe, due to asphyxia in childbirth. On the Apgar scale 5/7 points. On the 2nd day after birth, the appearance of edema of the right lower limb was noted, bandage was performed with elastic bandage.

In 2006, a diagnosis was made: Klippel-Trenaunay-Weber syndrome. Consulted in the Russian Children's Clinical Hospital in 2013, revealed: arteriovenous malformations of the vessels of the lower extremity, according to angiography data from 05.02.2014: arteriovenous fistulas are not defined.

Also, in the history of correlated heart disease (from 02.2006) - defect of the interatrial septum, abnormal drainage of pulmonary veins, the patient is regularly observed at the cardiologist. It is observed in a pulmonologist with the diagnosis: bronchial asthma, atopic form, mild persistent flow, remission period.

In the SPC «LIMPHA» is observed since February 2013 with the diagnosis of the Klippel-Trenaunay-Weber syndrome, arteriovenous malformation of the vessels of the right lower limb. Chronic lymphovenous insufficiency of the 4th degree.

Concomitant: Congenital heart disease: defect of the interatrial septum. Condition after correction of the defect in 2006. Dilated cardiomyopathy. Heart failure 1-2 degrees. Bronchial asthma, atopic form, mild persistent flow, remission period. Respiratory failure 0-1 degree.

Upon admission, there was a significant increase in the right lower limb, the right gluteal region due to swelling of soft tissues, muscle hypertrophy. Edema marked, moderately dense throughout the limb. Skin covers are swarthy, along the entire surface of the skin of the right iliac region, the right buttock, the right hip and lower leg, multiple discharge pale spots of capillary angiodysplasia. The symptom of the Stemmer is positive.

She received 3 courses of complex decongestive therapy with a positive effect. On the background of treatment - soft tissue softening, decrease of left lower limb in volume, so after the 3rd course of treatment in 2015 there was a decrease in the volume from 4,5 cm to 8,0 cm. After the course of treatment, he uses compression garment of flat knitting made on individual measures.

**Dynamics on the background of treatment (3rd course of complex physical anti-therapeutics, 2015)**

**Before treatment**

**After treatment**

**In compression garment**

All patients were recieved complex physical decongestive therapy, including manual lymph drainage, as well as compression bandage of the affected limb. Against the backdrop of treatment, positive dynamics was noted, softening of soft tissues and reducing of the affected limb in volume were observed in all patients. To prevent the development of edema, patients are advised to use daily compression garment flat knitted, matched to individual measures.

## The view of the chief editor

With such pathologies as the Klippel-Trenaunay-Weber Syndrome, lymphedema is one of the complications. Moreover, the increase of an affected limb in the volume is mainly due to tissue hypertrophy, rather than the accumulation of a fluid rich in protein, as in the classical versions of lymphatic edema. The increased load on the lymphatic system also happens due to varicose veins.

To reduce edematous component and decrease the risk of complications associated with inadequate functioning of the lymphatic system, it is advisable to use Complex Decongestive Therapy (CDT), which includes manual lymph drainage, compression therapy, skin care and exercise therapy.

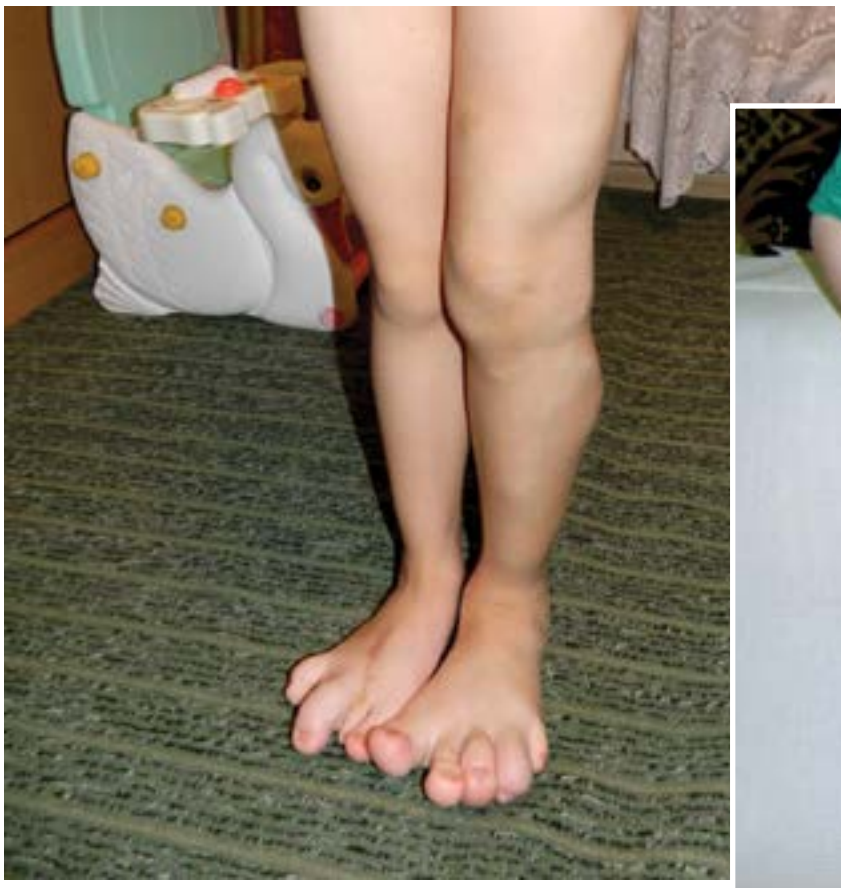
Many years of experience of our foreign colleagues have repeatedly proved that the use of CDT for such patients favorably affects lymphatic vessels, varicose veins, relieves inflammation, pain, stagnation, prevents the appearance of trophic changes on the skin.

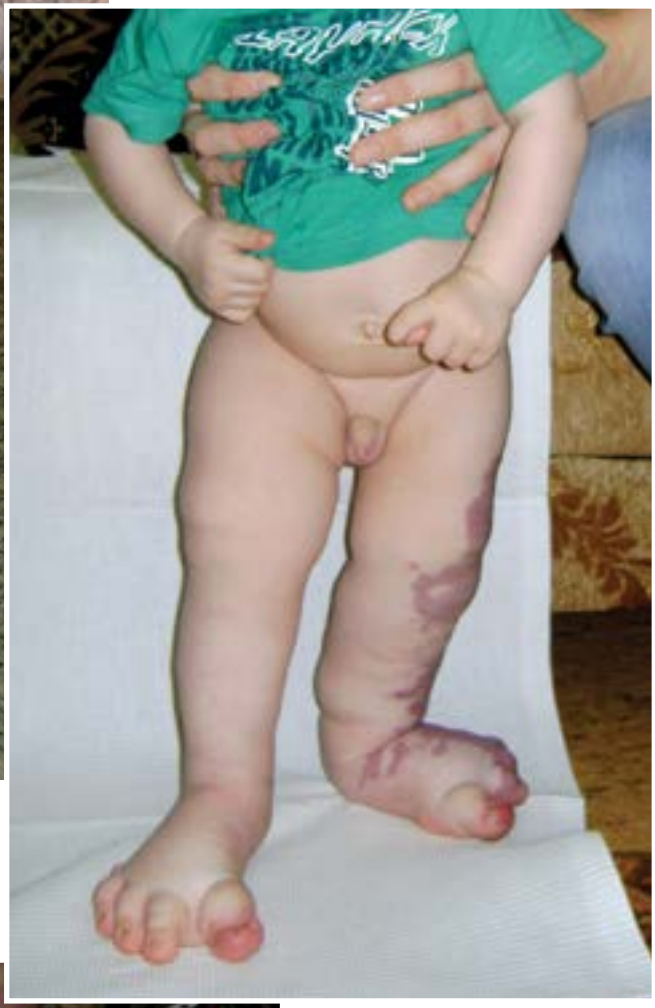
According to the photos of the patients treated in our center, which can be found in this article, you are able to see that edema has not significantly decreased after several courses of CDT (only 2-4 cm in circumference), but they are necessary to hold the result and partly to restrain hypertrophy of tissues.

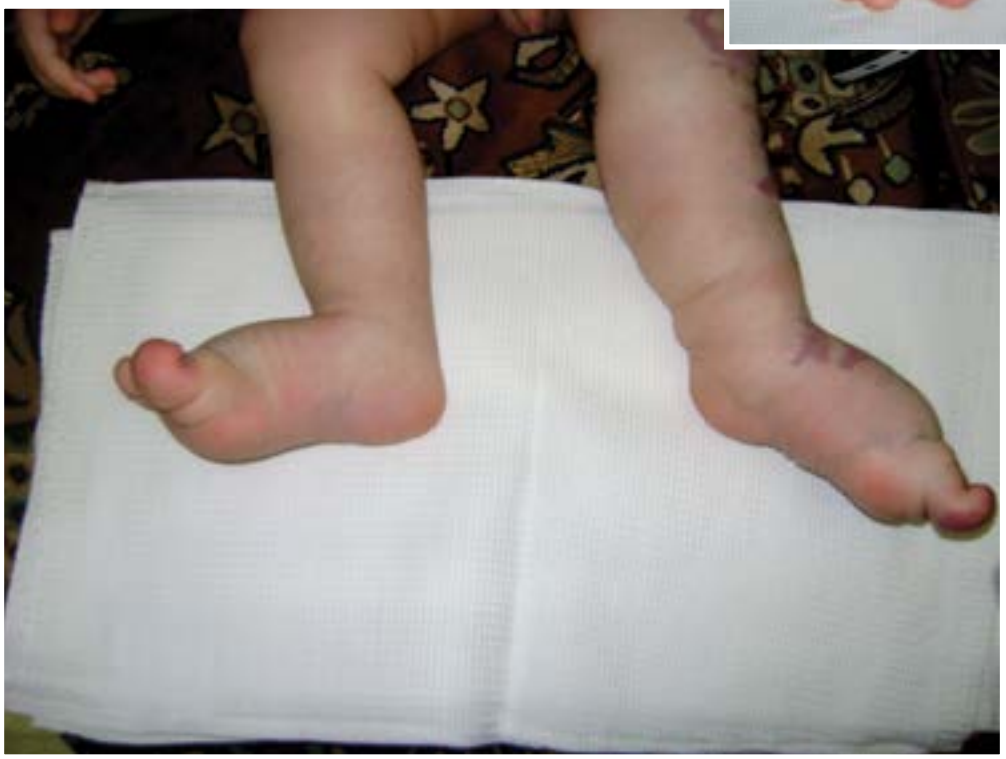
Constant wearing of compression knitwear of flat knitting, sewn by individual measures, fixes the result from the treatment, not allowing to build up swelling and protecting from injuries and infections.

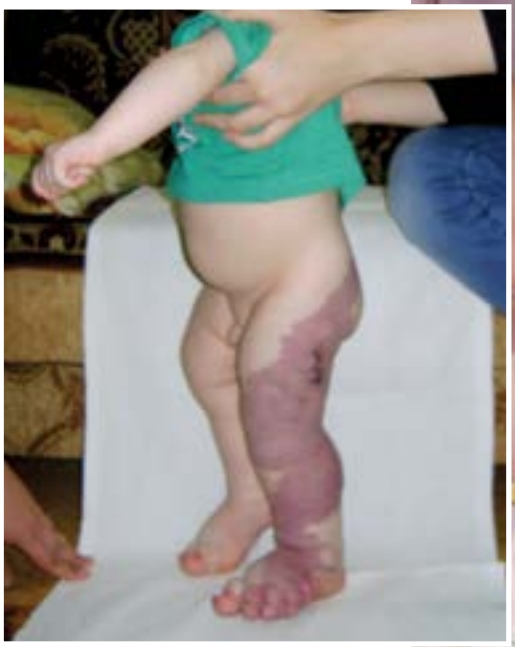
*Makarov I. G.,*
*the head of the medical and educational directions in Scientific practical center of rehabilitation of patients with lymphedema «LIMFA»*

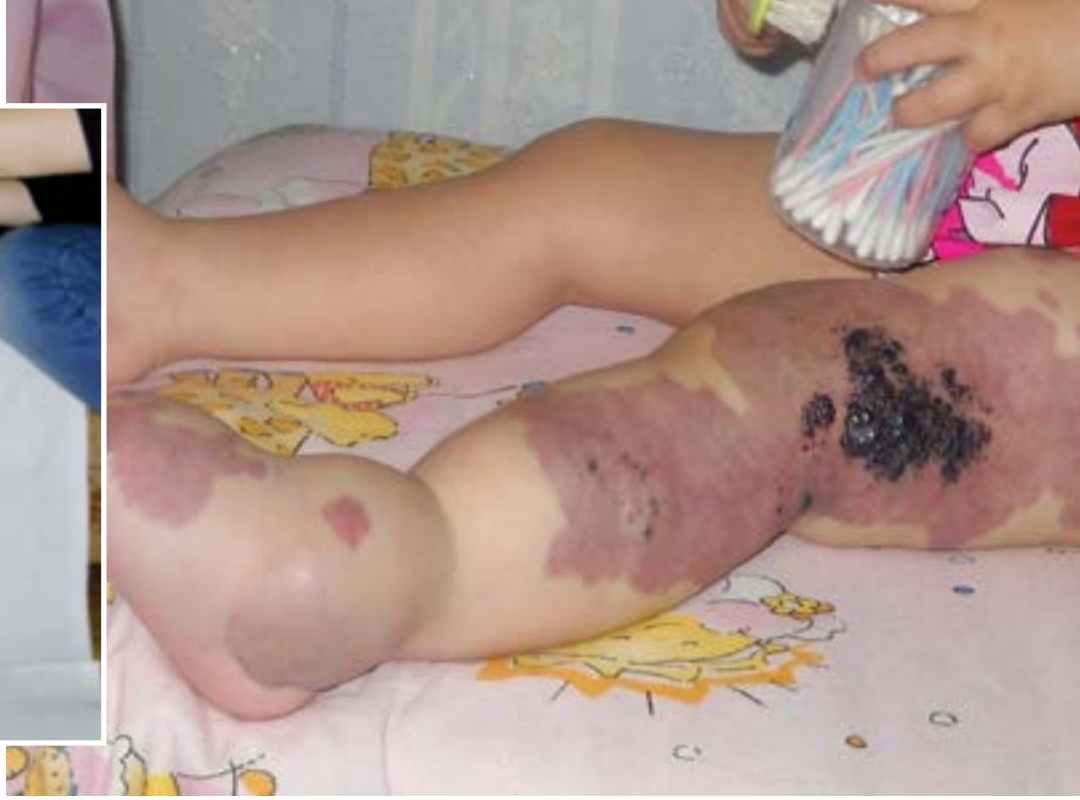
# Clinical variants of the Klippel-Trenauney-Weber syndrome











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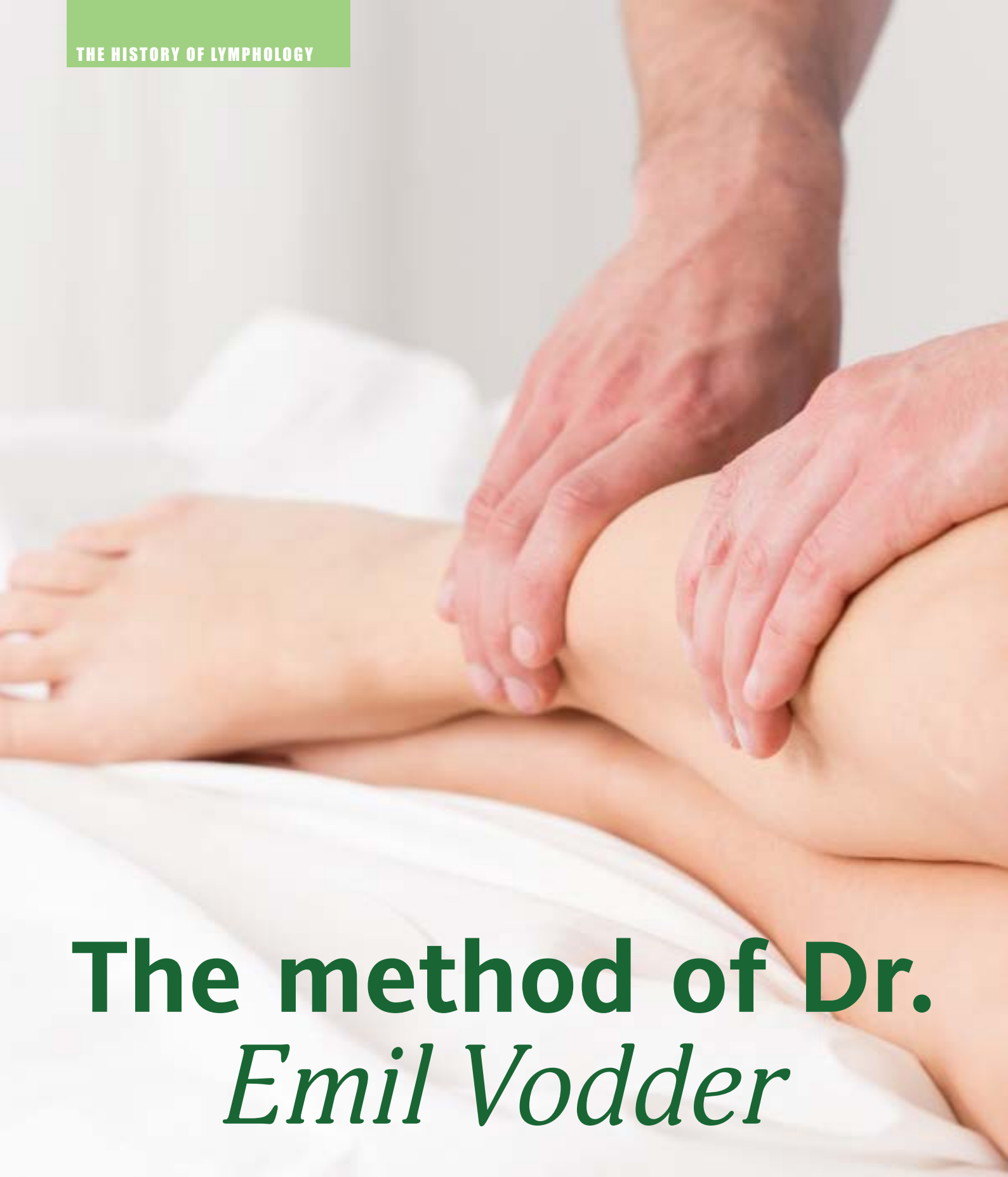

# The method of Dr. *Emil Vodder*

One of the founders of the Complex physical decongestant therapy is undoubtedly Emil Vodder. After Alexander Vinivarter invented the technique of manual lymph drainage in the late 19th century, Professor Vodder perfected this method and introduced it to the public in 1936 in Paris.

## The beginning of the way

Emil Vodder was born in Copenhagen on February 20, 1896. He was a Danish philologist and physiotherapist. Together with his wife, Estrid Vodder, he developed manual lymph drainage. Also, for the first time he identified lymphology as a separate branch of medicine.

After graduation, Vodder studied painting and art history, and to compare, he studied 10 languages.

Later, he had been working in the Royal Navy Map Archive for 6 years. Then, in his free time, he studied singing techniques and playing the cello.

At the University of Copenhagen, he was engaged in biology, mineralogy and botany, which was the beginning of his passion for medicine, cytology and microscopy.

At the end of the eighth university term Emil Vodder fell ill with malaria, that is why he had to interrupt his studies. After recovery, he was not admitted to finish his studies.

In 1929, Emil Vodder and his wife moved to the French Riviera.

*Emil and Estrid Vodder*

## Studying the works of other scientists

In Cannes, Vodder got interested in the lymphatic system. He knew many scientists who described «mystical pure water» many centuries ago. He read the works of these researchers. Gaspare Azelli (1581-1626) was one of them - he studied the lymphatic system on dogs. These were the times of opening many vessels and systems. For example, Jean Pecke (1622-1674) described the heal cistern and the thoracic lymphatic duct, which leads to the venous system. Olaf Rudbeck (middle of the 17th century) discovered the lymph vessels of the thick and rectum and confirmed that these vessels go to the heal cistern, as Azelli previously described. Rudbeck was the first to discover that the lymph from the tissues goes through the lymphatic vessels into the chest lymphatic duct then back to the circulatory system.

In the 18th century, it was investigated that the lymph nodes are located throughout our body, and that the function of the lymphatic system is to absorb the tissue fluid. For a long time this fact had been questioned.

Vodder specifically studied the life of his fellow citizen, a Danish scientist, Thomas Bartolin. In 1637, he was admitted to the University of Lieden (the Netherlands). Further, he continued his scientific research at the Anatomical Theater (The Netherlands), where he discovered the botanical garden, the library, and the hospital with patients for him. It was an ideal option, because in Northern Europe there were only plague houses and madhouses at that time. One of the aspects of Bartolin's study of Azelli's lymphatic vessels was the visualization of them by injecting a colorant. Bartolin was the first to describe the lymphatic system as a whole. He published his discoveries in the field of the lymphatic system in his work «Vasa lymphatica».

*In the 18th century, it was investigated that the lymph nodes are located throughout our body, and that the function of the lymphatic system is to absorb the tissue fluid.*

Studying the relevant literature, Emil Vodder convinced himself that a man is a biological unit, and also that lymph is omnipresent as a living environment. Vodder always quoted Drinker's prophecy: «The lymphatic system is the most important organic system for human and animal life.»

In the days of Vodder and Drinker, the interstitial fluid of loose connective tissue was called as lymph. If we consider that lymph (which means - loose connective tissue) is susceptible to micro-oocytes, this can be the cause of many diseases.

## The invention of manual lymph drainage

In 1929, when Vodder was engaged in the treatment of his patients, he started to make hypotheses. He palpated enlarged cervical lymph nodes of patients suffering from acne, migraine, sinus problems. He believed that the real reason for these ailments is a squeezing in the throat due to the increase of lymph nodes. Vodder suggested that such lymph nodes cannot effectively perform their cleansing function. And to normalize the situation, he invented a light stroking massage to open the «sluices» and remove excess water from them. Thanks to this massage, the condition of Vodder's patients improved significantly. Then he began to think about whether he really found the key to solving the problems of patients with a disease of the lymphatic system.

*Anatomy of the lymphatic system of a hand (according to Sappey)*

It must be understood that at that time in the 1930s there was a secret «taboo» in the medical society for interfering with the lymphatic system due to a lack of knowledge about it. But this did not stop Vodder, and in 1932 the couple began to actively study the lymphatic system, and developed special cautious massage movements with their hands along the lymphatic tract.

## Research in Paris

In 1933, Emil Vodder and his wife moved to Paris, where they continued their studies. They specially allocated time for studying the anatomy and physiology of the lymphatic system. In a large anatomical atlas, the Vodders found a collection of remarkable engravings by the famous French anatomist Sappey. These engravings have become the fundamental basis for the systematic and clear development of practical procedures. It was necessary to think through a new manual on the technique of practical implementation, describing circular movements, very light pressure, to avoid hyperemia under any circumstances.

*He invented a light stroking massage to open the «sluices» and remove excess water from them.*

In 1936, after four years of research, Emil Vodder introduced the technique of manual lymph drainage (MANUAL LYMPH DRAINAGE AD MODUM VODDER) to the world public at a congress in Paris, France.

Estrid and Emil Vodder devoted the rest of their life to demonstrations and teaching this method.

*Manual lymphatic drainage of the foot*

## Returning to Copenhagen

After 11 years in France, Vodder and his wife returned to Copenhagen during the break in World War II. In the early 50's, Vodder began to receive invitations to teach his method in European countries.

*One day Vodder said: «I discovered my method too early. Nobody understands me».*

In the 60s the German doctor-therapist, Dr. Asdonk, heard about Emil Vodder's activities and became interested in them. Therapists owe much to Dr. Asdonk. As a doctor, he recognized the importance of Vodder's method and compiled the first list of indications for manual lymph drainage. Paradoxically, the lymphedema was not included.

## "I opened my method too early ..."

In 1966, Dr. Wittlinger, Asdonck and Vodder founded the Association of Manual Drainage of Dr. Vodder. They organized the first congress, which was a great success. The problem at that time was that no doctor or scientist could understand how manual lymph drainage could positively affect the lymphatic system. One day Vodder said: «I discovered my method too early. Nobody understands me».

*Professor Mislin said: «If Vodder had not invented his method, it would have to be invented urgently.»*

In the 1970s, professor Mislin from Sweden, after conducting a study on lymphangia, managed to prove the effect of manual lymph drainage on lymphatic flow. This work confirmed that lymphatic drainage techniques stimulate the lymphatic system, increasing the speed of lymph movement and enhancing the work of lymphangions. Professor Mislin said: «If Vodder had not invented his method, it would have to be invented urgently.»

## Manual lymph drainage today

Vodder has always had a complex approach to a man. His main thesis was: «If one part is sick, then the whole body is sick.»

Nowadays, manual lymphatic drainage, developed by Dr. Vodder, is the basis of Integrated physical decongestant therapy, which is effectively used to treat lymphoedema of various etiologies all over the world.

In addition, the method of manual lymph drainage is used in a variety of areas of medicine. Since German health insurance recognized that technique 25 years ago, manual lymph drainage has become the most frequently prescribed procedure in this country.

## Recognition of the Vodder's method

Dr. Vodder's knowledge was not limited to the lymphatic system. In the 1930s, he studied the human immune system. He found that the lymph nodes play an important role in protecting the body.

Vodder was often offered to sell the rights to his method, he would have become a very rich man in that case. However, he responded: «I will not sell my life.»

In 1985, the German Association of Massage and Physiotherapy awarded Emile Vodder a medal in gratitude for his work of a lifetime. Along with this event, the professional association recognized the method invented by Vodder and gave it the name «Manual lymphatic drainage of Dr. Vodder».

Emil Vodder died in February 1986, shortly before his 90th birthday.

He undoubtedly greatly influenced and enriched physical therapy with his research and inventions. Thanks to Dr. Vodder, patients with lymphedema all over the world receive significant relief of their condition through manual lymph drainage nowadays. Many scientists, physicians and physiotherapists contributed to the recognition of Vodder's method, which he created 70 years ago, all over the world.

*Teaching the methods of manual lymph drainage nowadays*

*The professional association recognized the method invented by Vodder and gave it the name «Manual lymphatic drainage of Dr. Vodder».*

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# LYMPHANGOSAROMA as complication of an illegal lymphodema

[1] Scientific and Practical Center "LIMPHA"
Budojapova T.S., MD, lymphologist.

**Lymphangiosarcoma** (syn: Stuart-Treves syndrome) is extremely malignant, often rapidly metastasizing endothelioma. Most often develops against a background of chronic untreated lymphedema, penetrates the subcutaneous layer and, in rare cases, fascia. Lymphangiosarcoma develops against a background of reduced local immunity and occurs in any form of lymphedema. The majority of cases are made by patients after treatment of breast cancer (Stewart-Treves syndrome). However, single cases of lymphangiosarcoma are described on the background of congenital, idiopathic, post-traumatic and infectious lymphedema. In some cases, as a consequence of radiation, regardless of whether lymphedema is present [1]. Example: patient L., 24 years old, with primary early lymphedema of right lower extremity III st. (elephantia), the left lower limb of II st. M.Foeldi, sporadic form complicated by lymphatic drainage, presence of chillous reflux into the skin, erysipelas; untreated. (Figure 1).

**Etiology:**

The main cause of lymphangiosarcoma is untreated lymphedema, which, in turn, is the cause of such complications as erysipelas, trophic skin changes, lymphorrhea, etc.

Also, some authors as an etiological factor put forward a theory of genetic predisposition, which is not currently proven. An integral cause is radiation therapy (radiation), used in combination treatment of the majority of oncological diseases, which causes aseptic inflammation of all vessels, including lymphatic and soft tissues in the area of irradiation.

**Diagnosis:**

Extremely important in diagnosis is the patient's history, which will include prolonged progressive lymphatic edema, most often untreated. In the clinical picture, initially, small hemorrhagic spots appear (vascular asterisks, purpura), which look like bruises, but the color change associated with the successive decay of hemoglobin does not occur, and these «hematomas» are not accompanied by pain syndrome (Fig. 2). With the passage of time, the spots gradually increase, infiltrated and turn into plaques (Figure 3). Subsequently, plaques can grow in size and merge with each other, forming nodes (Fig. 4). From the nodes large conglomerates are formed with elements of decay (Fig. 5).

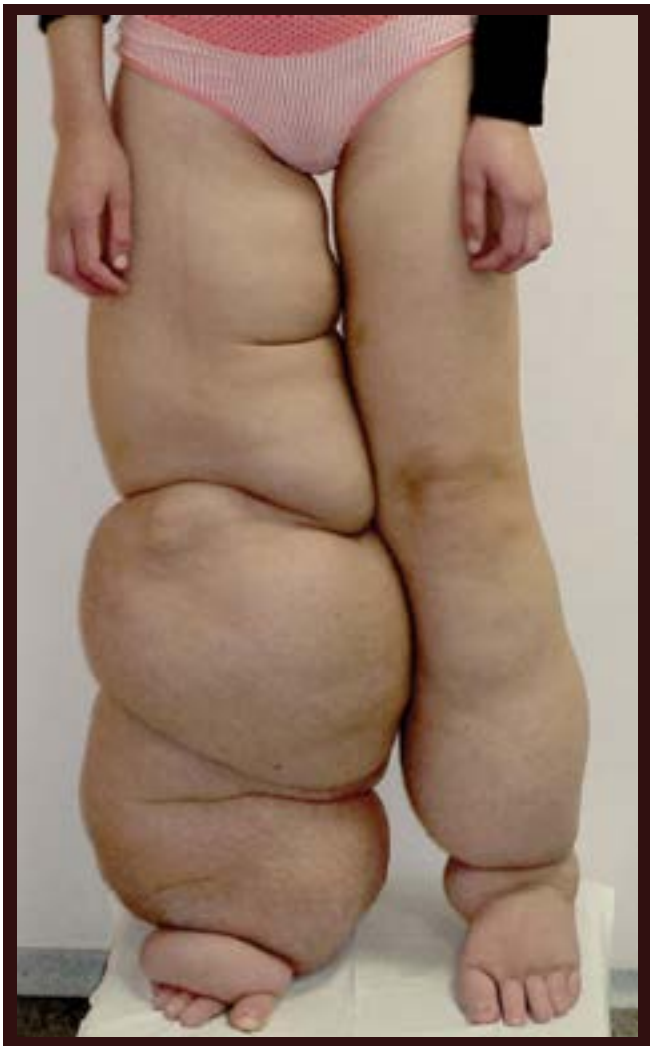
Diagnostics uses such methods of examination as: biopsy with histological examination for reliable diagnosis verification, CT / MRI for detection of metastases; positron emission tomography for detecting the degree of subcutaneous spread of the tumor.

With histological examination: in typical cases, under a light microscope, one can see a tumor with a tendency to hemorrhage, invading the skin and subcutaneous fat. Small or large solid tumor infiltrates are seen, consisting of polymorphic tumor cells with vesicular nuclei and protruding nucleolus. There are numerous mitotic divisions. In addition to solid tumor infiltrates, it is possible to see angiomatous or sinusoidal aggregates of tumor cells forming cavities of various sizes, inside of which are red blood cells or elements of cellular decay. Also, papillary tumors that protrude into the cavity of the cavity are common.

> Lymphangiosarcoma develops against a background of reduced local immunity and occurs in any form of lymphedema

**Differential diagnosis includes a number of diseases that need to be excluded:**

- Kaposi sarcoma (Kaposi angiosarcoma or multiple hemorrhagic sarcomatosis) is a multiple malignant neoplasm of the dermis (skin), against the background of secondary immunodeficiency (AIDS).
- malignant skin tumors (melanoma),
- Angioendothelioma (endothelial cancer of blood vessels),
- Cancer metastasis in the skin,
- Angiolymphoid hyperplasia, Kimura's disease (granulomatosis, combined with hyperplastic changes in lymphoid tissue and eosinophilia)



1Fig. No. 1. Patient L., 24 years with a primary lymphedema of both lower extremities

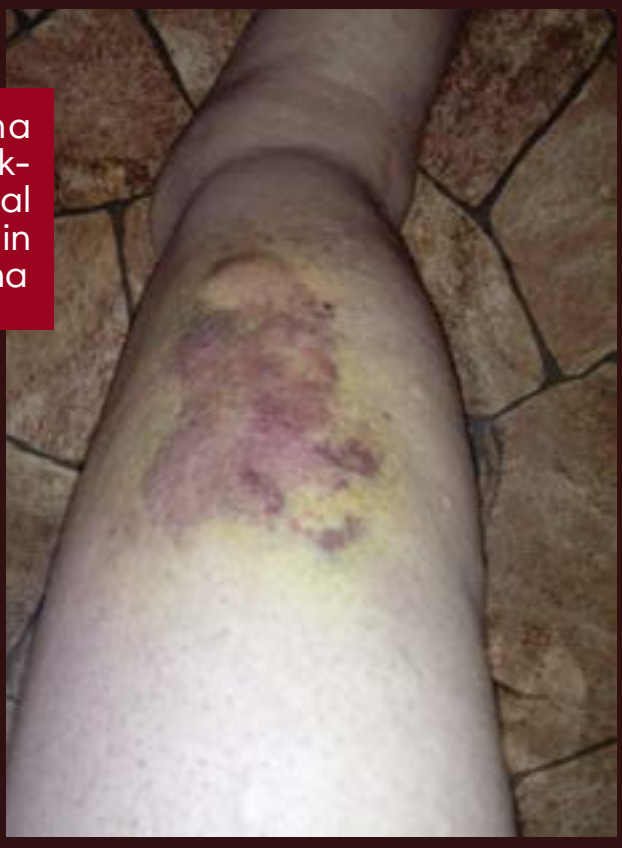


Fig. No. 2. Lymphangiosarcoma in the stage of hematomas

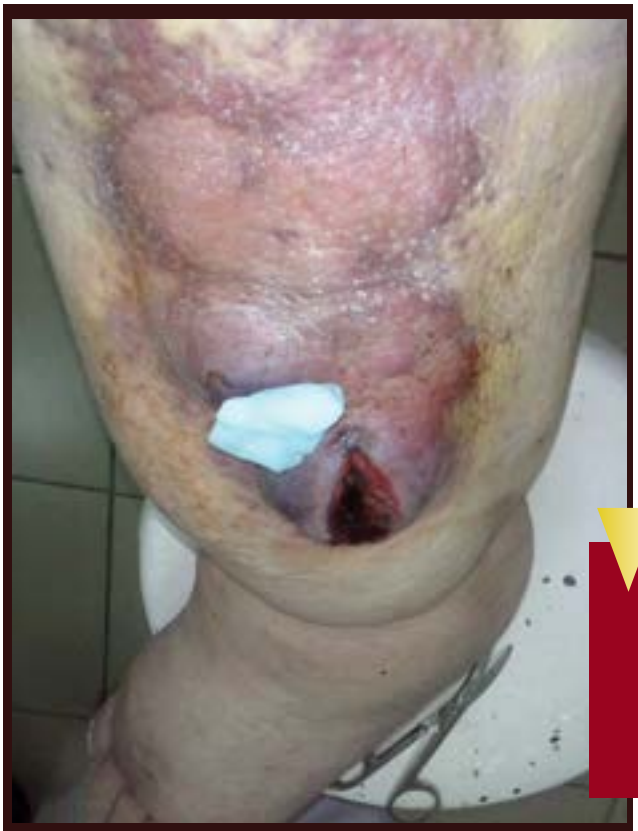


**Fig. No. 3. Lymphangiosarcoma in the stage of plaques**

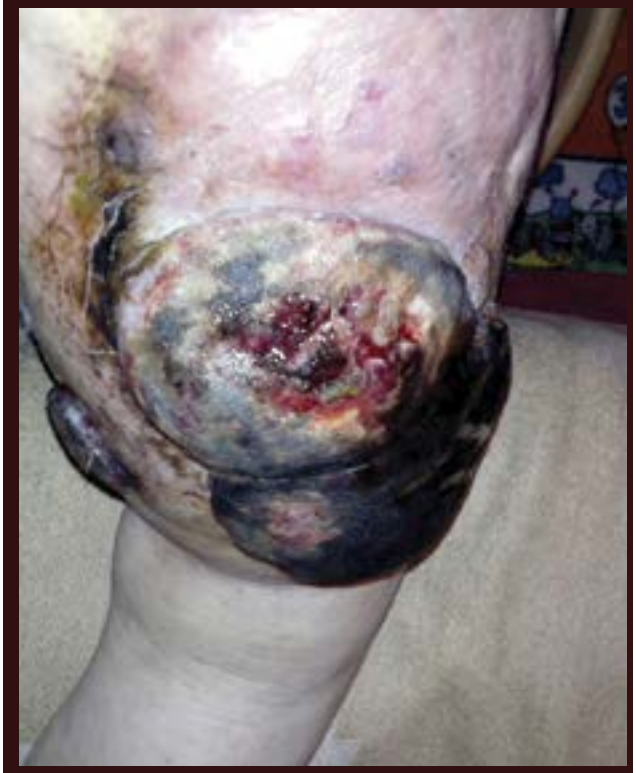


**Fig. No. 4. Nodes of lymphangiosarcoma**

Surgical treatment - early amputation or extended excision of the tumor focus is the primary treatment for lymphangiosarcoma. Of course, such an intervention is crippling for patients, but, in turn, has a positive effect on life expectancy. Chemotherapy / radiation therapy / immunotherapy is used necessarily in combination with surgical treatment.

As a primary prevention, regular «Complex physical decongestion therapy (CFT)» is necessary, which includes: manual lymph drainage, compression therapy (bandage and knitwear), skin care and exercise therapy. The method of CFPT allows to reduce edema, to achieve stable remission of lymphedema, to prevent the development of complications, to improve the quality of life of patients. [4]

**Secondary prevention:**

- Rapid diagnosis;
- Early combined treatment.

Forecast: extremely unfavorable.

Extremely important in diagnosis is the patient's history, which will include prolonged progressive lymphatic edema, most often untreated.

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Surgical treatment - early amputation or extended excision of the tumor focus is the primary treatment for lymphangiosarcoma

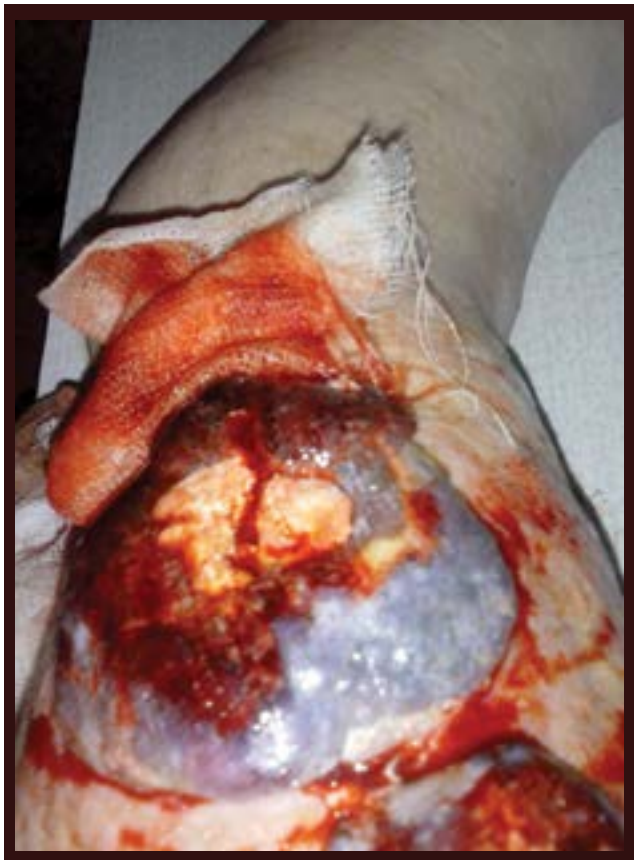

**Fig.5. Nodes lymphangiosarcoma with elements of decay**

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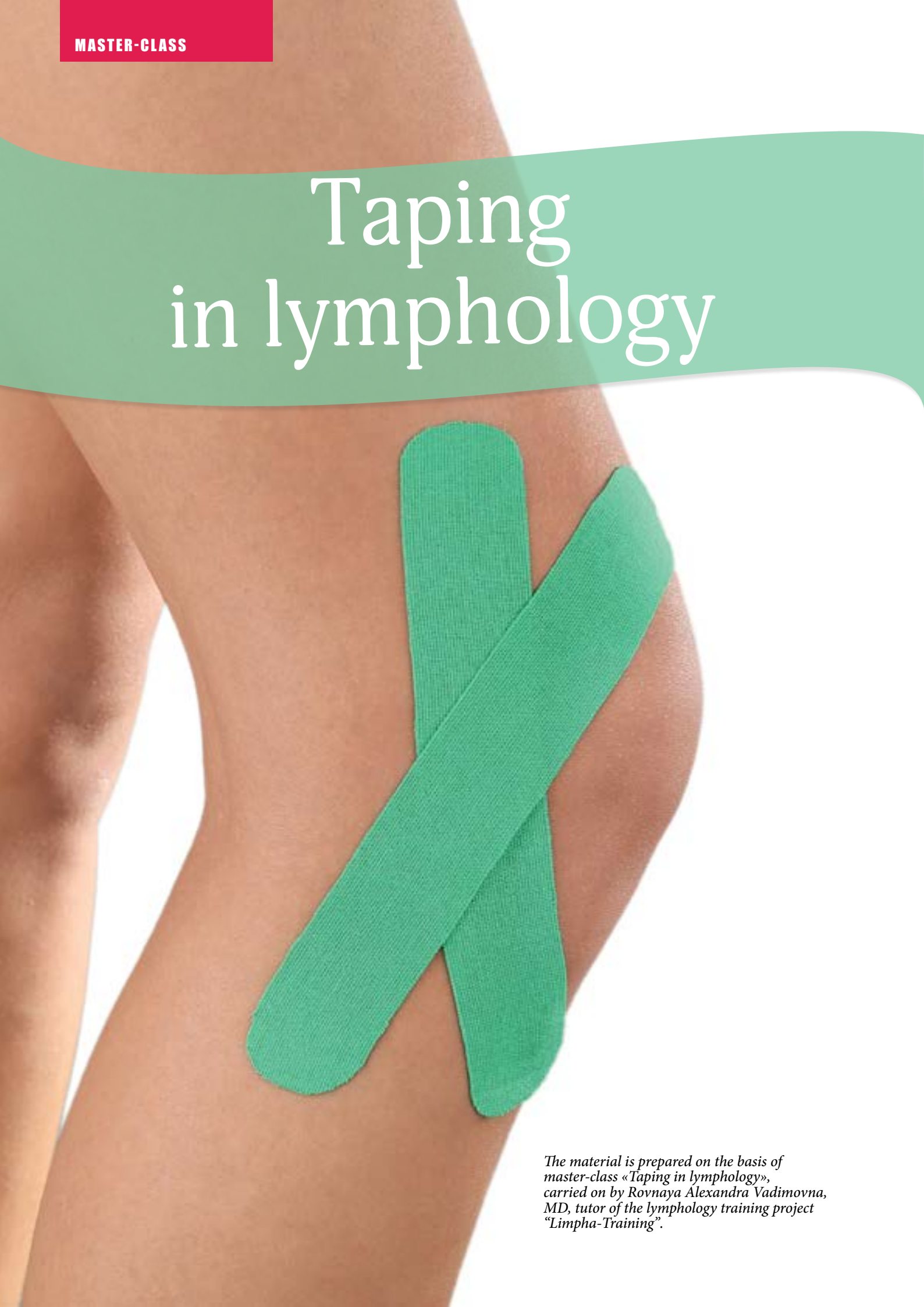
# Taping in lymphology



*The material is prepared on the basis of master-class «Taping in lymphology», carried on by Rovnaya Alexandra Vadimovna, MD, tutor of the lymphology training project "Limpha-Training".*

## A bit of history

Taping is a procedure of applying special patches (tapes) to prevent and treat various microcirculation disorders and diseases of the musculoskeletal system, to ease the load on the muscles, ligaments, joints.

The name comes from the English word «tape» - a sticky tape, a sticky adhesive.

Initially, tapping was used in sports medicine. A rigid, inelastic tape was placed to fix the ligaments or joints in case of trauma, to reduce mobility.

The development of such a direction as taping that is a part of kinesiology - the science of movements, which is especially popular in Japan. Japanese first developed the idea of putting an elastic stretchable tape over the skin, they believed that would create additional support for muscles, ligaments, and at the same time would affect sensitive skin receptors. And, if it is stuck according to special methods, then the upper layer of skin will rise above the subcutaneous fat, and then the microcirculation processes in these underlying layers will improve. This method had actively been developing in Asia in the 70-80s, and at the same time began to develop in the United States.

Taping technique became widely spread after sports events at the beginning of the XXI century, when it was noticed that many of the athletes performed with pasted colored tapes.

There were rumors that these patches were impregnated with medicinal substances. But this is not so. Tape is just an adhesive material made of cotton, which should be stretched along and slightly diagonally, but not across. In that case, it repeats the elasticity of skin. But the elasticity of tape is still limited. As a result of this limited ability to stretch, you can create support or give the position of a specific part of the body.

> *Tapes can be of different colors. Only black color should be paid attention to: in a hot climate with increased insolation it will provide a more warming effect precisely because of the ability of black color to accumulate heat. The Japanese, who originally developed the taping, pay a great attention to color therapy. Red color and red tones mean the activation of covered parts - this is an increase in blood flow, and an increase in muscle tone in a certain place. The blue color and cool shades of other colors have, on the contrary, relaxation, miorelaxation, anti-inflammatory effect. As for lymphedema, it is better to use both blue (reducing inflammation) and red (activating the lymphatic system, stimulating lymphangions) tapes alternately, thus alternating warm and cold shades.*

## Application of taping

You can use tapes in completely different areas. For example, in sports medicine in order to help athletes to quickly recover from injury, or to strengthen some physical indicators. At the moment, in some sports, kinesiotherapy has been banned, as it was leveled to light outfit. In medical practice, taping has also become widespread in such areas as neurology, orthopedics, traumatology and lymphology.

## Tape

Tape is a plaster. The standard tape width is 5 cm, some firms produce 2.5 cm for children and 10 cm for covering large areas. It has a cotton base. This means that it breathes and does not interfere with perspiration. It can be worn while swimming or taking a shower. At the same time, tape has an elastic base, that makes it is capable of stretching, thereby it repeats the skin's ability to stretch elastically. On the inner surface of a tape a layer of acrylic glue is applied (this kind of glue is hypoallergenic in the shape of waves. The stretching capability of tapes is most often 130%, in some cases it can reach 180%. Tape stretches only along. It is also important for the stabilization of certain structures. This ability of serving as a second skin provides support to those parts to which it is stuck.

## Operating principle

Tape is stuck to the skin.

1. Stimulation of skin receptors. Human skin is very sensitive due to numerous receptors. A skin tape carries signals through these receptors to the brain. The impulse from the skin surface along the efferent receptors enters the central nervous system (CNS). This causes the activation of certain areas of the cerebral cortex, which sends the impulses back to the receptors on the skin (afferent impulses). These impulses become larger, and they influence different receptors. For example, athletes improve coordination and accuracy of movements by stimulating proprioceptors, which are responsible for the positions of the body in space.

2. The effect of decompression. A tape is correctly pasted when it has the tension of fabric, picking up the skin. This micro lifting, when the upper layer of the skin is lifted above the underlying structures, first, mechanically reduces the pressure on the receptors responsible for pain impulses, and secondly, creates more space for microcirculation and for lymph flow.

3. Pain relief. Different types of sensitivity depend on different types of nerve fibers. There are so-called slow nerve fibers - the impulses pass slightly slower through them. And, there are fast nerve fibers where the impulses, accordingly, pass quickly. However, the area of the brain perceiving them is the same. If there is a slight pain, these pain impulses are transmitted through slow nerve fibers. To interrupt them, you need to give some sort of impulse that would go over the fast fibers. The impulses from the tape go through the fast nerve fibers, outstripping the pain impulse, which follows the slow pulse. Thus, the pain signal is blocked by the earlier pulse from the tape.

*Carrying out allergen test: you need to take a small piece (2 cm) of tape, stick it on the inner surface of the forearm and hold for 15 minutes. If there was no itching, skin irritation, pain sensations, then everything is all right, and you can start the procedure of taping.*

**To sum up, it is possible to list the main effects of the taping:**

1. Maintaining muscles and ligaments
2. Regulation of muscle tone
3. Anesthesia
4. Improvement of microcirculation and lymphatic transport.

## How to apply tape?

**1. Allergen test**

First, you need to execute test for allergies. Despite the fact that acrylic adhesive is low allergenic, absolutely any person can face a sporadic allergic reaction.

Carrying out allergen test: you need to take a small piece (2 cm) of tape, stick it on the inner surface of the forearm and hold for 15 minutes. If there was no itching, skin irritation, pain sensations, then everything is all right, and you can start the procedure of taping.

However, even after an allergen test, the patient may begin to complain of itching, pain and discomfort. It happens, if a specialist stretched tape too hard before sticking.

**2. Patient preparation**

Next, patient preparation and deciding where to stick the tape follows. The skin should be dry and clean. If necessary, the area for taping must be degreased. To do this, it is best to use alcohol wipes if a patient does not have an allergy to alcohol. Then, pay attention to the hairline. Firstly, tapes should not be tightly attached to the hairline, and secondly, removing this tape will be very painful for a patient. Accordingly, in order not to cause unnecessary suffering, the area for taping needs to be shaved with the permission of a patient.

**3. Cutting the tape**

First, you need to take a tape and try on the size. Prepare all the necessary pieces in advance. Scissors must be sharp, because tapes need to be cut without breaking, very accurately. If the edges of a tape are crumble, then it will quickly become unstuck.

Tapes cannot be cut off on a patient.

Before sticking these strips, you need to round off all edges. The fact is that if you leave the edges, the patient will always touch them when taking off / putting on clothes, therefore they will quickly get unstuck.

**4. Applying tape on skin**

Experts distinguish several parts in any strip of a tape. Those parts that are on the edges (the last 3-4 cm) are conditionally called anchors and bases. They are always stuck without tension. The tension of a tape is generated in the interval between the bases; it can reach a maximum of 130% of the original length.

To get the desired effect, skin should be slightly folded at the expense of a tape either by stretching a tape itself in the middle of a strip, or by stretching the skin and underlying tissues.

When a tape is applied to the skin, it is necessary to «activate» the glue by smoothing a tape out on skin so that heat is generated.

You should not rub tape too much on children - simply stick it and warm by hand, as children's skin is more tender and sensitive.

You should not keep a tape for its sticky part. From the moment when a sticky part is attached to your fingers, it begins to absorb fat and sweat from your skin, and it will not get stuck to a patient well. That is why tape has a paper substrate. A specialist should always hold exactly this part.

Tape should not end on a tape, the effect will be better in this case. To do this, you need to measure out all the pieces in advance.

There are very sensitive and tender areas (for example, elbow fossa), where it is necessary to adhere tape with caution or try to stick the base outside this area. It will not affect the direction of lymph flow, but can cause irritation and redness of skin. Moreover, the tape in these areas will be stretched and flexed when unbending a limb.

You cannot stick tape in the area of the carotid artery, as it can affect the carotid sinus, causing arrhythmias.

**5. Wearing tapes**

Tape should be worn for four to five days. All the effects of taping, with the exception of anesthesia, are cumulative and not immediately apparent. The pain can abate almost instantly.

Tape should be changed every 5-6 days.

You can apply all the methods of manual lymph drainage and compression bandage with a tape being stuck. These techniques work well in the complex.

*Tapes cannot be cut off on a patient.*

A person can wash with a tape, but it is prohibited to dry it with hot air of a hair dryer. Wipe a taep with a towel in the direction from the middle to the ends, only in that case a tape will remain in place.

If a small piece of tape has peeled off from a patient, then he

can trim this piece himself/herself neatly. This will not affect the taping effect.

## *Removing the tape*

Before removing the tape, the patient's skin should be tightened and fixed. Stabilizing your hand on the fixation point, remove a tap strictly according to the hair growth by slow and smooth movement. If a patient feels very painful, then it is better to wet the tape with warm water. For example, you can shoot tapes while taking a shower.

For young children, a tape is soaked with baby oil before being removed and it can be easily peeled off. After removing a tape, a cream with panthenol is applied to children's skin.

## *Contraindications to taping*

- Acute deep venous thrombosis
- Acute thrombophlebitis
- Damage to skin (any in the place of presumed taping)
- Skin infections and inflammation (including radiation dermatitis, sunburn after ultraviolet)
- Parchment Skin Syndrome
- Malignant diseases
- Allergy to taping materials
- Edema due to cardiac and renal insufficiency (since in renal and heart failure a patient has a water balance and a propensity to accumulate fluid, tapes, raising and increasing the space under skin (between skin and subcutaneous fatty tissue) will contribute even easier accumulation of liquid, so taping can be done only with compensated renal and heart failure and in the absence of edema)
- Acute infectious disease, accompanied by fever and intoxication.

## *Taping in lymphology*

There are many techniques of taping, including those to improve lymphatic transportation. Tapes can be used in the treatment of lymphedema to activate lympho-lymphatic anastomoses, to stimulate lymph transportation in the upper and lower extremities, in the abdomen. In addition, tapping is used in the area of fibrotic changes in the skin and subcutaneous fat.

If a patient with lymphedema has one injured arm or leg, then due to the fact that it is heavier than the other limb, the posture will change, and pain can occur in the joints, in the back. You can also fight with the help of taping with these phenomena. These are decompression techniques for pain points or joint support, as well as postural taping (working directly to correct the position of a body).

**Important! Tapping is an auxiliary method of treatment. Taping lymphatic drainage technique will not solve the problem of lymphedema. However, applied in the complex, it is able to improve the results of treatment, to alleviate a patient's condition and to make the work of a specialist in complex physical anti-therapeutics more effective.**

## *Lymphatic drainage technique for taping*

In the case of lymphatic drainage technique, tape is stretched and glued without tension. You can stretch skin, for example, on the chest - by tilting a patient in the opposite direction, on an arm - by flexing hand or extending fingers, but you cannot stretch a tape yourself.

When there is no way to stretch a part of the body by changing the position, for example, in the abdomen or buttocks, then you need to stretch skin with your hand and stick a tape without tension.

When applying lymphatic drainage techniques, you should not stick all 5 cm of tape. It must be cut into «noodles» (from 5 small strips) and base. Almost all tape manufacturers draw lines on the reverse side of a tape for this purpose, making it possible to cut it straight on the strips.

Adhesion of a tape in the higher and lower pressure range helps to give direction to the lymph flow.

It is not necessary to glue the whole area with tapes. In lymph drainage taping, it is important that there are areas without tape, i.e. the technique of taping should be intermittent. The lymph flow will be organized more effectively in this case.

> *Tape should be changed every 5-6 days.*

**Lymphatic drainage technique for tape adhesion to upper limb lymphedema**

In case of radiation fibrosis it is better to use tapes along this zone

Mesh of "noodles"

### *Method of taping*

First, the base is stuck into the area of local lymph nodes. If you work with a brush and forearm, the base will be stuck to the area of the elbow joint. If the axillary node is preserved, then the base is stuck closer to the armpit.

If a group of lymph nodes is removed somewhere, there should not be a sticking of a tape base. In this case, you need to bypass the area of the removed lymph nodes and fix a base higher. Tape will be superimposed on the pathway used to improve lymphatic transport in manual lymphatic drainage . Thus, if it is necessary to direct local lymph flow to the area of the healthy axillary lymph node, then the tapes should be stuck in this direction.

Next, «noodles» go through the anastomosis and are stuck to the area of an armpit. However, not in the armpit itself, but on a healthy side without reaching an armpit. Sticking noodles along the lymphatic vessels gives a certain direction of a lymph flow. It is always necessary to go through the tape area of lymphatic anastomosis. During manual lymphatic drainage, anastomoses are also activated. Anastomosing vessels will be opened longer and will shrink better if a tape is stuck on the top.

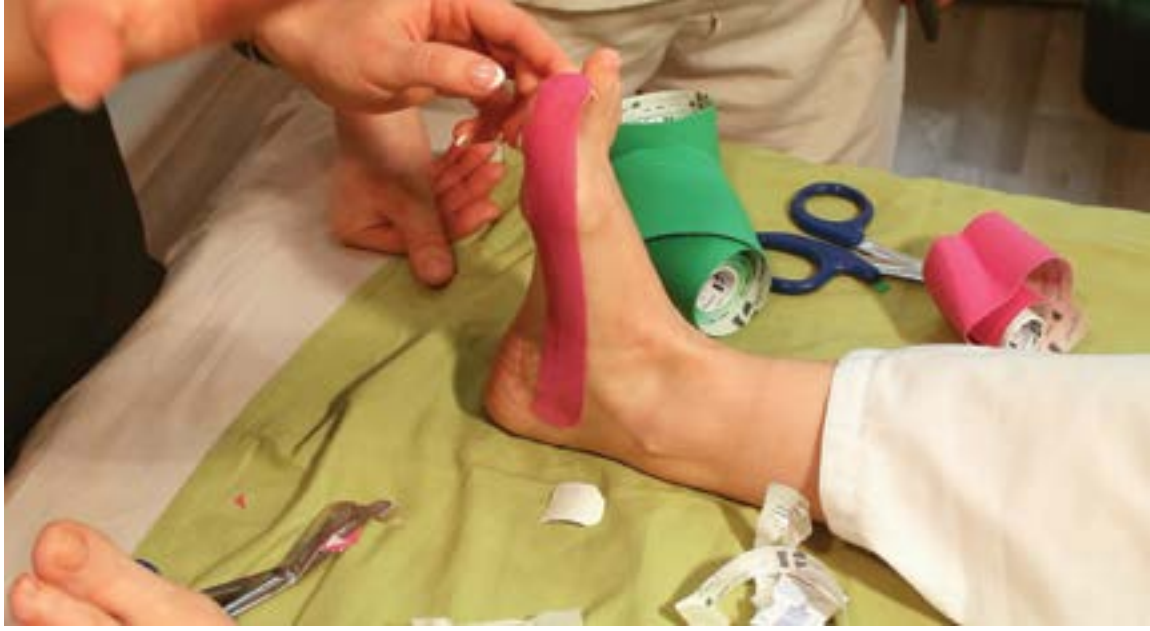
Women with secondary lymphedema (postmastectomy edema) in the area of edema, often face consequences of radiation fibrosis. In this case, you do not need to stick this area, because the there is more tender and prone to damage when peeling off. If radiation therapy is not applied to this place, or if it has passed without consequences, then tiring can be a good aid to complex physical decongestion therapy. In the area of a scar and the area of radiation fibrosis, lymph should be helped to find ways to bypass this place. In this case, you can use tapes along this zone to create the right direction for the lymph drainage.

If there is a very strong edema, prone to fibrosis in some area, then the «noodles» are stuck in the form of five lines with five more lines on the top at an angle of almost 90 degrees, creating a semblance of a grid.

Such a grid will work effectively in the abdomen, and even in the face and neck area along the lymphatic pathways, from the posterior lymph nodes to the subclavian and supraclavicular. Base is stuck in the supraclavicular or subclavian area for the effect.

It is very important to be well aware of the ways

Lymph drainage technique of taping with lymphedema of lower extremities



Taping through a nail plate

of the lymph drainage when applying taping on any area.

Most of all, the effect of taping is noticeable in acute swelling with trauma.

### *Which stages of lymphedema experience effective taping results?*

It is obvious that the greatest effect can be achieved in the initial stages. At the 1st degree of lymphedema, which is not prone to progression, it is possible to apply tape even as a replacement for compression knitwear in order to prolong the effect of manual lymph drainage.

More serious stages of lymphedema can also be treated by this method, but the results will not be so significant. However, taping will undoubtedly contribute to enhancing the effect of complex physical decongestant therapy.

It often happens that there are places where the swelling is better or worse. For example, there are women with secondary lymphedema with a long-lasting edema in the scapula region. Bundling this area is almost impossible, and taping is very helpful in this case.

If a patient underwent sectoral resection of the tumor, rather than a radical mastectomy, then tapping can be used in the area of the mammary gland. This can be especially effective after the course of manual lymph drainage.

### *What are the peculiarities of taping children?*

Taping children of the age of two to three years old with lymphedema of lower extremities is particularly important. Each base is attached to a rear of a foot and each strip (excluding little toe) will not pass through the interdigital spaces, as with lymphatic drainage technique, and on each finger through a nail plate and displayed on a foot. This helps, first, to improve the lymph drainage of fingers themselves, and secondly, it helps, especially on a thumb, to prevent the development of the problem of ingrown or incorrectly growing nails - a fairly common problem in small patients with lymphatic edema of a foot.

Another technique that is successfully used in children is taping of the base of toes. To do this, the tape square is cut in the form of half H letter, that is, having two lines and a base. The base is stuck to the base of a toe, and the legs are stuck with the proximal phalanx into the girth. This contributes to a better transport of lymph in this area (which is difficult to affect during the imposition of compression banadage). In addition, the band is used on the top of the tape, which allows a more even distribution of pressure.

*Taping can be used in various fields of medicine. With regard to the use of tapes in patients with lymphedema, this may be useful in combination with complex physical decongestion therapy (CPT), which includes manual lymph drainage, compression bandage, exercise therapy and skin care. Taping prolongs and intensifies the effect of manual lymph drainage, directing the lymph flow in the right direction, which will improve the result from the treatment. It should also be remembered that tapping is an auxiliary technique for the majority of patients with lymphedema, and its use should be agreed with the treating physician-lymphologist.*

Tape, carved in the form of half H letter, is stuck to the proximal phalanx of a toe, the base is stuck to the base of a finger

# “Every person is a unique integral and consistent organism, full of causal relationships”

Stella Arbitmann is a medical doctor in the world’s leading lymphological clinic.

Foeldi Klinik is the world's leading clinic for the treatment of lymphatic vessels. In order to make some points clear and learn how to treat lymphedema in Europe, we have talked with a doctor from Foeldi Klinik - Stella Arbitman.

**- Glad to see you, dr. Arbitman! Could you tell us, please, about yourself. And why did you become a lymphologist?**

- After graduating with honors from the school, I realized that my future profession should be connected with working with people. Therefore, I decided to become a doctor, and I have never regretted about my decision in the future.

I graduated from Leningrad First Medical Institute as a therapist. Today, this is the St. Petersburg First State Medical University named after academician I.P. Pavlova.

**- How had your further medical career been developing and what is especially interesting for us is how you found yourself in lymphology?**

- I accumulated great medical experience in various fields of medicine, working as the head of the department of one of the clinics in Leningrad. Moreover, I was always interested in complex clinical cases with severe diagnosis and non-trivial methods of treatment.

After moving to Germany in 1991, I was able to continue my medical practice, passed the exam, received a medical certificate of a specialist (Facharzt Allgemeinmedizin) and began a search for a hospital where patients with unusual clinical cases can be found. I got to Foeldi clinic by accident and did not plan to stay there long, but the profile of the clinic and the approach to treatment of the patients completely convinced me, because it corresponded to my ideas about good medicine. And I have been working for many years there.

Disease of the lymphatic system, as a rule, is a consequence of other clinical disorders in the body. Work in the field of lymphology allows you to meet with very diverse diseases; some of them are unique and almost never occur in ordinary hospitals. Here I was able to satisfy my desire to help patients with complex clinical cases.

A very important aspect for me and, of course, for the patient, is that Foeldi Klinik treats not the patient's illness, but takes into account the whole complex of his/her problems. In addition, lymphology is an insufficiently studied and very interesting area of medicine. So, once I got into lymphology, I stayed there for almost 16 years.

**- Stella Efimovna, now the question to you as an expert in the field of lymphedema treatment: is the complex physical decongestant therapy (CFTT) a standard for the treatment of lymphedema at the moment?**

- Yes, it is. It is the standard for the treatment of lymphedema.

**- What do you think about the prospects in the treatment of lymphedema? Are there any new studies in lymphology on this subject?**

- I do not think that in the near future some other methods will become the world standard for the treatment of lymphedema. Of course, science does not remain unchanged. Now intensive studies of genetic factors causing primary lymphedema are conducted. This means that probably in the future we will be able to live in the world when it will be possible to recognize and prevent the development of lymphedema at the genetic level. Those are the prospects. The second very interesting direction is the discovery of vascular growth factors, i.e. substances that cause the growth of new lymphatic vessels. An experimental treatment of a small group of patients with secondary lymphedema of hands is already under way after surgical treatment of breast cancer.

That means that perhaps in the future we will be able to treat the cause of lymphedema and, thus, achieve a perfect cure, but this issue is still at the level of scientific research.

**- If the cause is a malformation of the lymphatic system, and, for example, it is possible to force new lymph vessels to grow (activate lymphangiogenesis), then what about patients with the second and third stage of lymphedema? There are already changes in tissues, subcutaneous fat...**

- In my opinion, for complex physical anti-edema therapy will become our nearest future. Perhaps, other methods will appear, but for today, for example, we do not know how to prevent the appearance and development of primary lymphedema. When we will be able to prevent it at the genetic level, that is, in utero, there will be a revolution in this area. Let

**St. Petersburg First State Medical University named after academician I.P. Pavlova**

Foeldi Klinik, Hinterzarten, Germany

us wait. So far, this is only a theory.

If we are talking about cancer, then a lot has been done, including in the field of its prevention. In the treatment of advanced cases of lymphedema, surgical treatment will be used, but the standard will remain the same (CFPT-Ed. Ed.). Now more operations are done with secondary lymphedema than before, and some additional treatment methods appear, but they are not standard, as surgical treatment is indicated only for a certain group of patients with lymphedema. I very much doubt that the standard of treatment will change in the nearest future.

**- So, now, the main directions for science to move in terms of treating lymphatic edema are either their prevention or treatment at the earliest stages?**

- Yes, right. First, we are talking about prevention, if we talk about secondary lymphedema. Primary lymphedema is more difficult issue, it is necessary to understand how it happened, what the reason is... There is a lot of new, it is very interesting.

**- Please, tell our readers about your first meeting with a patient. How does your conversation start?**

- Each patient, each person is a unique holistic single organism full of causal relationships. From the doctor's point of view, the most reasonable approach is to consider a patient as a single interconnected organism. When I communicate with a patient, I want to learn everything about his/her medical history.

**- In addition to lymphology, what other areas of medicine are most interesting to you?**

- Cardiology and psychiatry (is not it an interesting combination?), but still I am interested in lymphology most of all.

**- And what is the most interesting issue in lymphology?**

- I am interested in vascular malformations, especially the combination of malformations of the blood and lymph vessels, in particular - the combination of venous and lymphatic malformations, expressed lymphatic malformation is also on the list, lymphangiomatosis, i.e., so-called complex malformations.

**- Do vascular malformations include reconstructive surgery?**

- Yes, there is a comprehensive treatment, it is interesting. A pure vascular malformation - arterial or venous - is not our profile. However, in reality, they almost all come with lymphedema. Patients with lymphangiomatosis are very interesting - they have peripheral and central edema, and ascites, and pleural effusions. There are cases of a combination of primary lymphedema, venous malformation and effusions in different cavities, so-called Klippel-Trenone syndrome, or there is no venous malformation, but there are effusions in conjunction with peripheral lymphedema. Therapy of patients with complex vascular malformations (complex angiodysplasia) is a complex medical problem. Since each patient with a vascular development disorder is unique in its own way, there is no standard treatment guidelines. The therapy of each patient is individual. This is a rare pathology, the number of specialists with experience in diagnosis and treatment in this area is limited. It is necessary to attract doctors of different specialization, coordinate strategies, exchange information. In addition, therapy of such diseases is long-term, correction of strategy and treatment methods is necessary in connection with the change in symptoms as the disease develops.

**- Stella Efimovna, as far as we know, do you still teach at school?**

- Yes, I am an assistant professor of Foeldi Schule and I teach lymphology in the school for the training of physiotherapists.

S. Arbitman and I. Makarov on the 43rd European lymphological Congress in Stuttgart, 2017.

**- What other medical activities do you do?**

- I constantly work in the hospital, I treat patients. At the university clinic in the city of Freiburg, a monthly consultation of physicians of different specialties is held to discuss methods for treating patients with malformations. Professor Földi and I are regular participants in these meetings.

**- Many thanks, dr. Arbitman, for an interesting interview. Our readers will be very interested to know the opinion of a doctor from Foeldi Klinik on the treatment of lymphoedema of various etiologies. Until next time!**

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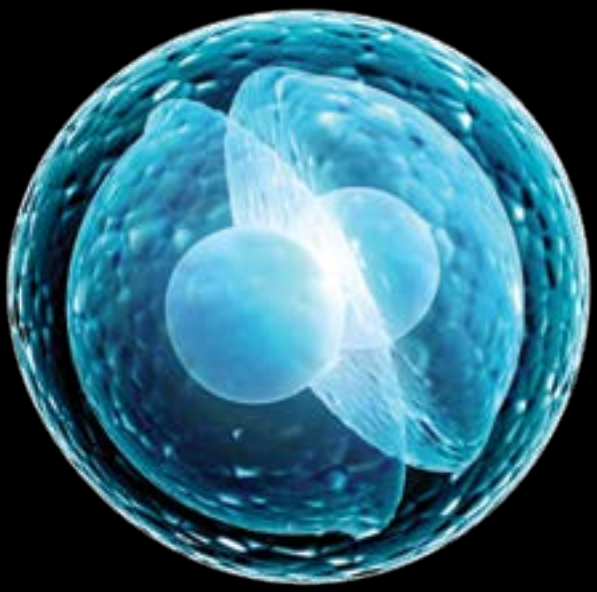

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