



**European  
Reference  
Network**

for rare or low prevalence  
complex diseases



**Network**

Vascular Diseases  
(VASCERN)

VASCERN DO'S AND DON'TS FACTSHEETS  
FOR RARE VASCULAR DISEASE PATIENTS  
FACING FREQUENT SITUATIONS

# Pediatric and Primary Lymphedema

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# VASCERN

VASCERN, the European Reference Network on Rare Multisystemic Vascular Diseases, is dedicated to gathering the best expertise in Europe in order to provide accessible cross-border healthcare to patients with rare vascular diseases (an estimated 1.3 million concerned). These include arterial disease (affecting aorta to small arteries), arterio-venous anomalies, venous malformations, and lymphatic diseases.

VASCERN currently consists of 31 highly specialised multidisciplinary Healthcare Providers (HCPs) from 11 EU Member States and of various European Patient Organisations and is coordinated in Paris, France.

Through our 5 Rare Disease Working Groups (RDWGs) as well as several thematic WGs and the ePAG – European Patient Advocacy Group, we aim to improve care, promote best practices and guidelines, reinforce research, empower patients, provide training for healthcare professionals and realise the full potential of European cooperation for specialised healthcare by exploiting the latest innovations in medical science and health technologies.

More information available at: <https://vascern.eu>

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# Abbreviations

**NSAIDs:** Nonsteroidal Anti-Inflammatory Drugs

**PPL-WG:** Pediatric and Primary Lymphedema Working Group

**BMI:** Body Mass Index

# Introduction

These factsheets have been written and reviewed by the experts of the VASCERN PPL-WG. The PPL-WG agrees with the recommendations but wishes to emphasize that these are recommendations made by consensus at expert level. We would recommend that these factsheets be used as a guide to implement locally agreed policies.

These factsheets are meant for patients as well as for caregivers. Implementing these recommendations should go hand in hand with strategies to educate patients about medical situations where specific care is required and about relevant symptoms and how to act when they occur.

# Diagnosing Primary Lymphedema



### WHAT IS RECOMMENDED

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- Consider the diagnosis in any newborn or child with swelling of one or more limbs, chylothoraces/pleural effusions or ascites, for which no other obvious explanation is present.
- Refer children suspected of having lymphedema to an expert centre for inter(multi)- disciplinary analysis (follow the guidelines).
- Include parents in the diagnostic process if the lymphedema is present in a child.
- Refer adults with lymphedema to an expert clinic for inter(multi)-disciplinary analysis if lymphedema is present in one or more limbs and no obvious other explanation is present, especially if other congenital anomalies or disorders are present.



### WHAT YOU SHOULD NOT DO

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- Perform routine genetic testing, lymphoscintigraphy and blood examinations without dedicated lympho-vascular expertise.

# Inter/(Multi)-disciplinary treatment

There exist various non-surgical treatment options for lymphedema and a few surgical options. These can be divided into reconstructive and reductive methods. All surgical treatments should be performed in a dedicated expert center with interdisciplinary cooperation and embedded in a non-operative treatment protocol.



### WHAT IS RECOMMENDED

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- After confirming the diagnosis of primary lymphedema, an interdisciplinary treatment protocol should be created for the individual patient.
- Treatment of lymphedema is divided into 2 stages: initial and maintenance treatment phases: ensure that the patient (or the parents) is aware of this.
- The treatment protocol is monitored by the lymphedema therapists.
- Compression technology is the cornerstone in both stages of treatment.
- Swelling of the toes can be treated with compression and podiatry.
- The patient and/or the parents should pay attention to skin care, toenail problems and be vigilant for signs of cellulitis.
- Monitoring of the patient's weight and volume / circumference, with clinimetrics and photography, are part of the treatment.



### WHAT YOU SHOULD NOT DO

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- Wait and see without follow-up.
- Surgery for lymphedema without an interdisciplinary team approach.
- Reconstructive surgery in patients with primary lymphedema.

# Follow-up for patients with primary lymphedema



### WHAT IS RECOMMENDED

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- Children and adults with primary lymphedema should be followed regularly in an expert centre for individuals with primary lymphedema.
- Follow-up for children and adults with primary lymphedema should be provided using the standard practice, but this standard practice may need to be adapted depending on the nature of the primary lymphedema, due to potential differences in their response to management and the possible development of other associated health problems.
- The expert centres should organize a network of care and work with the local healthcare provider and have an open door policy.
- Patients should be pro-active and be encouraged to monitor their own lymphedema (self-management).



### WHAT YOU SHOULD NOT DO

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- Fail to follow-up the patient.



# Physical activity and lymphedema

Physical activity is very important in the management of lymphedema. During the maintenance phase, exercise in combination with compression is recommended. No type of physical activity is contraindicated in children with lymphedema. Regular physical activity is beneficial for both the affected limb and for general health.

An increase in limb volume during exercise is normal and transient and should not result in the cessation of physical activity.



### WHAT IS RECOMMENDED

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- There are no physical activity or sport restrictions. Let the children play.
- Physical activities in combination with compression will stimulate lymph flow and reduce swelling.
- Exercise is important to reduce weight (if they are overweight) or keep weight under control.
- Exercise is important to prevent muscle wasting.
- Wear compression garments during exercise if tolerated. If not tolerated, remove compression garments during exercise but put them back on afterwards or wear a lighter compression garment during exercise.
- Be aware of the daily recommended physical activity level for adults (10,000-15,000 steps a day).



### WHAT YOU SHOULD NOT DO

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- Take more rest than usual as part of lymphedema treatment.
- Sleep in a chair / recliner chair with legs downwards.
- Perform activities that may harm the skin without proper protection.
- Refrain from physical activity because of lymphedema.
- Restrict physical activities associated with daily life.

# Weight and lymphedema

Obesity is the most important risk factor for the worsening of lymphedema. Weight loss to the normal range of weight and BMI will result in a reduced severity of the lymphedema.



### WHAT IS RECOMMENDED

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- Have a normal healthy diet.
- If intestinal lymphangiectasia, chylothoraces, chylous reflux and/or chylous ascites are present, a medium chain triglyceride (MCT), high protein diet should be considered.
- Aim for a healthy weight (BMI 19-24).
- Measure weight every 3-6 months.



### WHAT YOU SHOULD NOT DO

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- Allow obesity to develop (BMI > 29).

# Cellulitis/erysipelas

Children and adults with lymphatic impairment have a much higher risk of developing cellulitis/erysipelas.

The clinical signs of erysipelas are: high fever (39-40°C) of sudden onset, with rigors, a 'flu-like' or unwell feeling, followed by redness, warmth, and increased volume of the affected limb. Sometimes additional treatment is necessary, depending on the diagnosis.



### WHAT IS RECOMMENDED

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- Awareness of the first signs and symptoms of cellulitis.
- Prompt treatment of cellulitis / erysipelas with systemic antibiotics.
- Consider hospitalization in case of a young child and/or marked illness.
- Proper skincare and good compression treatment.
- Prompt treatment of interdigital fungal infections (Athletes foot) and eczema.
- Take preventive measures such as good hygiene following injury or insect bite: clean with soap and water and disinfect using topical antiseptic cream.
- Provide antibiotics for the patient to use if they feel that they are starting to develop cellulitis.
- Consider preventive long-term use of antibiotics in children/adults with recurrent cellulitis (2≥ episodes per year).



### WHAT YOU SHOULD NOT DO

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- Ignore the increased swelling.
- Stop wearing garments or low-stretch bandages. The patient may need to take off their compression materials for the first 2 days because of pain and/or discomfort in the acute phase, but they should be reintroduced as soon as possible or the lymphedema will deteriorate.
- Take antibiotics without clear signs of cellulitis/erysipelas.

# Genetic testing for Primary Lymphedema

Genetic testing is a specialised test, which can clarify a genetic cause of primary lymphedema. The indication for testing and the type of test may vary.



### WHAT IS RECOMMENDED

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- Every family with one or more children or adults with primary lymphedema should be referred for genetic counseling.
- Gene testing should be performed only in the context of evaluation by a inter(multi)disciplinary clinic, in which a clinical geneticist participates.



### WHAT YOU SHOULD NOT DO

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- Perform routine genetic testing without expertise in lympho-vascular medicine or clinical genetics and without patient consent.

# Pregnancy and Lymphedema

Pregnancy can increase lymphatic filtration and may result in aggravation of the lymphedema. However, this is usually reversible.

Consider the risk of an offspring or sibling of an affected individual inheriting the same condition.



## WHAT IS RECOMMENDED

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- Standard monitoring of pregnancy and focus on lymphedema.
- Contact the patient's center of expertise for genetic counseling for risk of lymphatic problems in the fetus and the option of prenatal diagnosis.
- More frequent assessments by the lymphedema therapists. They may need extra help with bandaging or compression garment fitting.
- Continue compression and bandage treatment as long as possible but adapt to the patient's needs (e.g. use thigh length stockings instead of tights).
- Encourage control of the weight gain.
- In the event of a sudden increase in volume, a venous Doppler ultrasound of the lower limbs is indicated to exclude an underlying deep vein thrombosis (rare).
- Consider the guidance of an obstetrician during pregnancy, including advice for delivery.
- If the patient is on prophylactic antibiotics, ensure that they are not harmful to the fetus.



## WHAT YOU SHOULD NOT DO

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- Stop all compression treatments.
- Ignore a sudden increase in swelling.
- Start anticoagulants because of the lymphedema swelling alone.

# Compression treatment

Compression technology is the cornerstone in lymphedema treatment. There are dedicated modalities for both the initial and maintenance treatment phases. Combinations of various products can be very helpful to the patient.



### WHAT IS RECOMMENDED

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- Measure the affected limb to provide 'made to measure' garments.
- Use various compression technologies and teach the patient how to use them.
- Application of compression by the patient or the parent can be learned easily in most cases.
- Compression treatment is always individualized for each patient.
- Wear flat-knitted garments (Circular knit garments are often ineffective).
- Dedicated use of various compression technologies is possible for all ages.



### WHAT YOU SHOULD NOT DO

---

- Ignore the lymphedema.
- Withdraw compression without proper monitoring.

# Medication and lymphedema

No medication is formally contraindicated. However, some medications may aggravate the lymphedema so consider alternative treatments when available.



### WHAT IS RECOMMENDED

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- Consider the risks and benefits when medication is provided which can lead to more swelling. (eg calcium channel blockers).
- Be aware that the following drugs may aggravate the swelling: calcium channel blockers, corticosteroids, NSAIDs, sex hormones and related compounds, pregabalin, docetaxel, zoledronic acid, and sirolimus.
- In case of doubt: contact the centre of expertise responsible for the care of the patient.



### WHAT YOU SHOULD NOT DO

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- Ignore any increase in the degree of swelling.
- Ignore a worsening of the fit of the compression garment.
- Use diuretics to treat the lymphedema.

# Operative intervention in a lymphedematous limb

Sometimes an operation is indicated in a patient with lymphedema. Precautions should be considered especially when a operation is performed on a lymphedematous limb, even for conditions which are not directly related to the lymphedema (e.g. varicose veins, hydrocele).



### WHAT IS RECOMMENDED

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- Discuss the risks and benefits with the centre of expertise responsible for the patient.
- Consider consultation with a specialist in lymphatic diseases prior to non-lymphatic surgery.
- Choose the direction of the incision to minimize the damage to the lymph vessels (e.g. hydrocele operation by a midline scrotal incision instead of a groin incision).
- Post-operatively, ensure proper compression treatment with bandages if there is a hematoma or extra swelling.
- If there is no extra swelling, continue wearing compression garments.
- A team of specialists in lymphatic diseases should perform surgery for lymphedema.
- Measure the swelling/volume /circumference pre-and post-operatively.



### WHAT YOU SHOULD NOT DO

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- Discontinue the routine lymphedema treatment (e.g. compression garments) when the patient undergoes surgery.
- In general, operations for lymphedema are not indicated for patients under the age of 18 years.



# Anesthesia

Anesthesia does not pose a particular problem in patients with lymphedema.



### WHAT IS RECOMMENDED

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- No specific recommendations related to the management of a patient with lymphedema.
- If necessary, blood samples, infusions and blood pressure can be performed on the affected limb. However due to the volume of the limb, which can make these procedures more difficult, the use of an unaffected limb is preferred if possible.



### WHAT YOU SHOULD NOT DO

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- No specific recommendations.

# Care for patient with traumatic injuries



### WHAT IS RECOMMENDED

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- Consider extra compression to the swelling of the limb (including the fingers/toes).
- Monitor the swelling.
- Be aware of the increased risk of cellulitis and consider antibiotic prophylaxis.



### WHAT YOU SHOULD NOT DO

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- Forget the routine lymphedema treatment (e.g. compression garments).

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