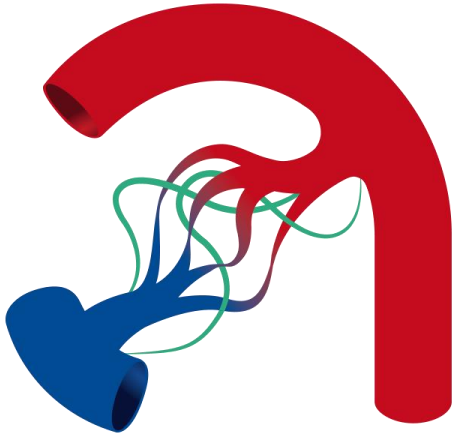
European Reference Network

for rare or low prevalence complex diseases

Network Vascular Diseases (VASCERN)



General Patient Pathway for Pediatric and Primary Lymphedema

Final Approved Patient Pathway by the Pediatric and Primary Lymphedema (PPL) Working Group – Version 3 - 13/04/2023 by VASCERN PPL working group Members:

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Updated from: Patient Pathway by the Pediatric and Primary Lymphedema (PPL) Working Group – Version 2- 21/06/2021 by Robert Damstra, Florence Belva, Nele Devoogdt, Janine Dickinson, Guido Giacalone, Kristiana Gordon, Peter Hall, Pernille Henriksen, Heli Kavola, Vaughan Keeley, Manuela Lourenço Marques, Michael Oberlin, Katie Riches, Tanja RuČigaj, Sinikka Suominen, Sarah Thomis, Stéphane Vignes, Kirsten van Duinen, Malou van Zanten, Sahar Mansour

Disclaimer

- This document is an opinion statement reflecting strategies put forward by experts and patient representatives involved in the Pediatric and Primary Lymphedema (PPL) Rare Disease Working Group of VASCERN.
- This pathway is issued on 13/04/2023 and will be further validated and adjusted as needed.
- Responsibility for care of individual patients remains with the treating physician.

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Gathering the best expertise in Europe to provide accessible cross-border healthcare to patients with rare vascular diseases

https://vascern.eu

General Patient Pathway for Pediatric and Primary Lymphedema (V3 - 13/04/2023)

1ST LINE

MULTIDISCIPLINARY EXPERT CENTER in cooperation with local center

Start

Swelling? Is this lymphedema?

Clinical evaluation
Family history and travel history
Evaluation skin (by stemmer sign/ pitting)
Renal function (by blood and urine) , cardiac function (by echocardiography) and hepatic function (by blood)
Function venous blood system (by venous doppler/ duplex)
Abdominal or pelvic involvement (by ultrasonography)

Refer to expert center, if
Edema (i.e. present for >3 month), in combination with one of the following:
- Congenital
- Family history of swelling
- Genital swelling
- Systemic involvement (e.g. **intestinal lymphangiectasia**, pleural effusions, pericardial effusions, ascites, chylous reflux)
- Syndromic forms (other associated anomalies, intellectual disability)
- Recurrent cellulitis

Multidisciplinary clinical evaluation
ERN PPL-WG

History and examination to include:
Age of onset
Family history
Distribution (upper limb(s), lower limb(s), face, genital)
Cellulitis /erysipelas
Weight, size, BMI
Skin problems (e.g. warts, vesicles, papillomatosis, nail abnormalities, fungal infection)
Systemic involvement (e.g. intestinal lymphangiectasia, pleural effusions, pericardial effusions, ascites, chylous reflux)
Associated problems (e.g. venous insufficiency, segmental hypertrophy, intellectual disability, dismorphic features)
Previous surgery

Clinical diagnosis

Investigations may include:
Limb volume (by circumferences/ water displacement)
Water content (by tissue dielectric constant/ bioimpedance)
Physical clinimetrics (e.g. strength, endurance, pain, discomfort)
Quality of life
Blood: albumin, full blood count, immunoglobulin
Stool (if signs of intestinal malabsorption or hypoalbuminemia: Faecal alpha 1 antitrypsin/ Calprotectin)

Imaging:
Lymphoscintigraphy
ICG (Indocyanine Green lymphography)
MRI
MRL (magnetic resonance lymphangiography)
Intranodal lymphangiography
Ultrasound (venous system, soft tissue, scrotum if suspicion of hydrocele)
X-ray (chest, affected limb if segmental hypertrophy)

Genetic testing (if appropriate)
Refer to genetic specialist

Management and treatment

Follow-up
At expert and local centers

Initial phase = intensive daily treatment
In case of pitting edema or wounds
By care provider/ patient/ guardian
Aim is to reduce edema volume, close wounds, improve functioning and quality of life, and improve insight in self-management

Maintenance phase = daily maintenance treatment
In case of stable non-pitting edema, without wounds
By care provider/ patient/ guardian
Aim is to maintain volume reduction, keep wounds closed, prevent development or recurrence of cellulitis and further improve functioning, quality of life and insight in self-management

Treatment components during initial or maintenance phase
Compression (e.g. garments, short stretch bandages, wraps, pneumatic device)
Exercises (e.g. mobilizing, strengthening, endurance, flexibility)
Skin and wound care
Education for self-management

Treatment components for the individual patient (i.e. personalized care)
Weight management and diet (e.g. MCT)
Manual lymphatic drainage (MLD
Psychosocial support
Medical therapy (e.g. sirolimus, prophylactic antibiotics)
Surgery for lymphedema

Refer to
Other specialties (e.g. gastroenterologist, pulmonologist, dermatologist)
(Inter)national support groups

LEGEND: Clinical evaluation | Investigations | Management / Treatment | Genetics

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VASCERN

Gathering the best expertise in Europe
to provide accessible cross-border healthcare
to patients with rare vascular diseases

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 diseases (affecting aorta to small arteries), arterio-venous anomalies, vascular malformations, and lymphatic diseases.

VASCERN currently gathers 48 expert teams from 39 highly specialized multidisciplinary HCPs, plus 6 additional Affiliated Partner centers, coming from 19 EU Member States, as well as various European Patient Organisations, and is coordinated in Paris, France.

Through our 6 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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