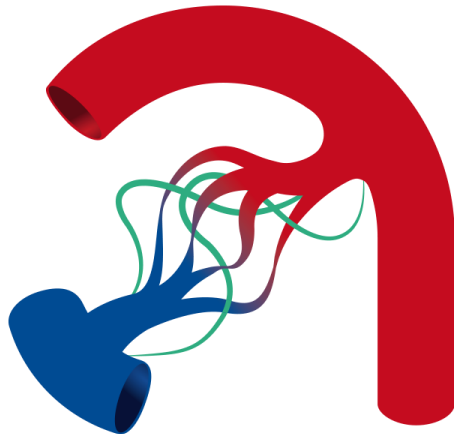




**European
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for rare or low prevalence
complex diseases

 **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 2 - 21/06/2021 by VASCERN PPL working group Members:

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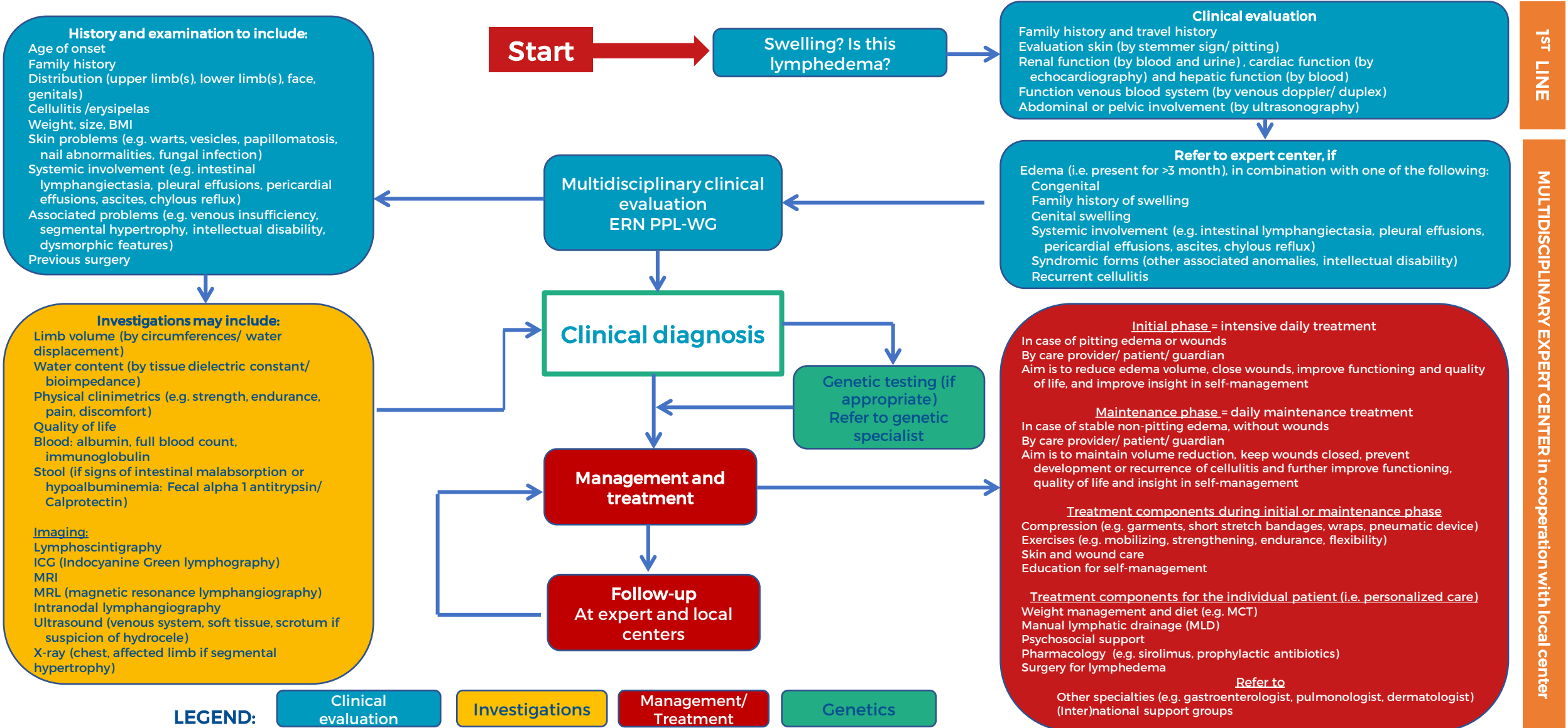
Updated from: Patient Pathway by the Pediatric and Primary Lymphedema (PPL) Working Group – Version 1 - 08/11/2019 by Robert Damstra, Florence Belva, Nele Devoogdt, Janine Dickinson, Guido Giacalone, Kristiana Gordon, Peter Hall, Pernille Henriksen, Heli Kavola, Vaughan Keeley, Manuela Lourenço Marques, 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 21/06/2021 and will be further validated and adjusted as needed.
- Responsibility for care of individual patients remains with the treating physician.



General Patient Pathway for Pediatric and Primary Lymphedema (v2 - 21/06/2021)



History and examination to include:

- Age of onset
- Family history
- Distribution (upper limb(s), lower limb(s), face, genitals)
- 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, dysmorphic features)
- Previous surgery

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: Fecal alpha 1 antitrypsin/ Calprotectin)

Imaging:

- Lymphoscintigraphy
- ICC (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)

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

Clinical diagnosis

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
- Pharmacology (e.g. sirolimus, prophylactic antibiotics)
- Surgery for lymphedema

Refer to

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



European
Reference
Network

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

VASCERN currently gathers 31 expert teams from 26 highly specialized multidisciplinary HCPs, plus 7 Affiliated Partner centers, from 16 EU Member States, as well as 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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Co-funded by
the Health Programme
of the European Union