General Patient Pathway for Pediatric and Primary Lymphedema

Final Approved Patient Pathway by the Pediatric and Primary Lymphedema (PPL) Working Group – Version 1 - 08/11/2019

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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 08/11/2019 and will be further validated and adjusted as needed.

• Responsibility for care of individual patients remains with the treating physician.
Start

Swelling? Is this lymphedema?

Clinical evaluation including:
- Family history and travel history
- Examination (stemmer sign/ pitting)
- Renal function and albumin level
- Urine for albumin
- Venous doppler/duplex
- Abdominal ultrasound

REFER if:
- Congenital or chronic (>3/12)
- Family history of swelling
- Genital swelling
- Chylous effusions
- Syndromic
- Recurrent cellulitis

Initial
- Compression (garments, bandages)
- Exercise
- Skin care
- Management of cellulitis
- Patient/Guardian education for self-management

Maintenance
- Compression (garments, bandages)
- Exercise
- Skin care
- Prevention of cellulitis

Consider
- Weight management
- MLD
- Diet (e.g. MCT)
- Psychological support
- Referral to other specialties
- Pharmacology (e.g. sirolimus, prophylactic antibiotics)
- Surgery

Genetic testing (if appropriate)
Refer to genetic specialist

Management and treatment

Follow-up
At specialist and local centers

Clinical diagnosis

Multidisciplinary clinical evaluation
ERN PPL-WG

Investigations may include:
- Limb volume
- QoL
- Physical clinimetrics
- Tissue dielectric constant / bioimpedance
- Baseline bloods: albumin, full blood count, immunoglobulin
- Stool, alpha 1 antitrypsin
- Imaging:
  - Lymphoscintigraphy
  - MRI
  - MRL
- Intraneural Lymphangiography
- Ultrasound - venous, soft tissue
- ICG
- Chest X-ray

History and examination to include:
- Age of onset
- Distribution
- Cellulitis
- Systemic involvement
- Warts
- Skin problems
- Segmental overgrowth
- Family history
- Associated problems
- Venous incompetence
- Previous surgery

LEGEND:
- Clinical evaluation
- Investigations
- Treatment
- Genetics

MULTIDISCIPLINARY EXPERT CENTER in cooperation with local center
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 consists of 30 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.

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