



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



## https://vascern.eu Lymphatic malformation: Management and treatment (Diffuse and extensive LM) Part B Diffuse and extensive lesions, MRI to evaluate relation to vital verified deep extension or fluid structures and organs and prior to /lymph effusions in deep cavities on surgery or sclerotherapy **MRI** Generalized Lymphatic Anomaly (GLA) Gorham stout disease (GSD), Channel type LM, Kaposiform Lymphangiomatosis LMs part of the central conducting lymphatic Mixed- and macro cystic LM not limited to GLA phenotype with bony (KLA) vessels associated with lymphatic shunts or the subcutaneous tissue with deep involvement GLA phenotype associated with expansion, lymphatic/chylus leak into pleura, chylus reflux coagulation disorder such as pericardium or the peritoneal cavity Kasabach Merritt Phenomenon (KMP) CT for investigation of bony involvement 1) Blood work, with haemostatic/coagulation evaluation 2) Consider MRI lymphangiography to evaluate lymphatic shunts and chylous reflux **Always** Somatic PIK3CA/ germline ARAF/ somatic NRAS mutation? individualized treatments tailored to reduce risk of life threatening Treatment options are: complications, 1) Dietary adjustments to reduce chylus circulation improving life 2) Watchful waiting if no symptoms and no risk of complications to vital functions if sudden swelling occurs. quality including 3) Sclerotherapy if no risk of complications to vital structures, first line of treatment in macrocysts including limited bone cysts. prevention of 4) Consider surgery when indication is to protect vital functions if sclerotherapy requires several sessions with high risk of prolonged intensive care. psychosocial 5) Consider lymphangiography with embolization in case of lymph flow obstruction or lymphatic leakage or chylus reflux impairment. 6) Low molecular weight heparin if haemostatic disorder and recurrent bleedings 7) Consider Sirolimus, if surgery or sclerotherapy is not possible or safe. Clinical Associated Investigations Particular cases **Treatment** LEGEND: evaluation