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Symposium Highlight

IMAGING AND INTERVENTIONAL MANAGEMENT OF LYMPHATIC DISORDERS

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ABSTRACT

Lymphatic flow disorders are reviewed, and a classification based on magnetic resonance lymphography findings outlined. Examples of successful interventional management based on this classification are provided.

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Keywords: Lymphatics, thoracic duct, lymphatic imaging, interventional management, lymphatic flow disorders, classification

Lymphatic dysfunction and abnormalities in the lymphatic system affect millions of people throughout the world causing debilitating symptoms, impairing their quality of life, and in certain cases shortening their life as well. Recognition and characterization of lymphatic disorders has become increasingly more common with the development of multicompartment access techniques for imaging the lymphatic system with MR Lymphangiography. With the more readily available ability to visualize the lymphatic system, there has also been an increased interest in the medical community to gain the technical experience to perform these newer techniques to image the central lymphatic system as well as to perform percutaneous and surgical interventions on the lymphatic system.

LYMPHATIC FLOW DISORDERS: BACKGROUND, CLASSIFICATION, AND IMAGING INTERVENTIONS

Advanced imaging and interventional techniques are critical for guiding management and recommendations for complex lymphatic disorders. There are several imaging techniques that are essential for diagnosing and planning interventions for lymphatic disorders. This comprehensive overview encompasses diagnostic methods, interventional procedures, and their applications in various lymphatic conditions.

The gold standard to image the central lymphatic system is dynamic contrast magnetic resonance lymphangiography (DCMRL), in which inguinal lymph nodes, liver lymphatics, and mesenteric lymphatics are accessed under ultrasound guidance in the cardiac catheterization lab, using a 25- gauge spinal needle (1,2). Patients are then transferred into an adjacent MRI suite equipped with a 1.5-T magnet (Siemens Healthineers transferred into an adjacent MRI suite equipped with a 1.5-T

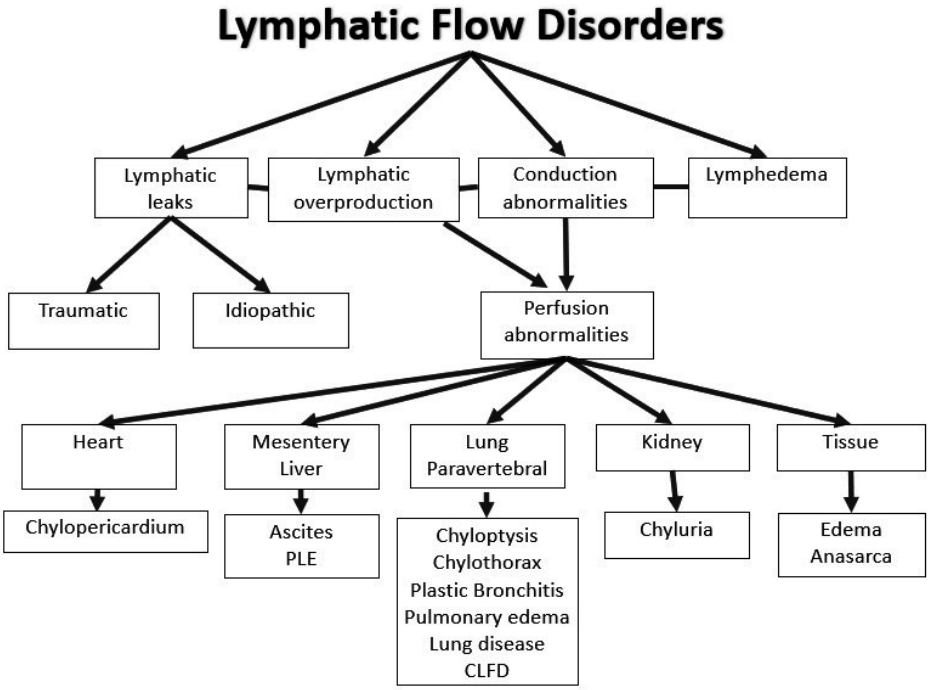

Fig. 1. *Classification flow chart for central lymphatic flow disorders and their clinical presentation.*

adjacent MRI suite equipped with a 1.5-T magnet (Siemens Healthineers MAGNETOM Avanto). Heavy T2-weighted MRI lymphatic imaging using a respiratory navigated and cardiac-gated three-dimensional turbo spin echo sequence is completed. For DCMRL, a weight-based dose of undiluted Gadobutrol (Gadavist, Bayer Healthcare) at a dose of 0.1-0.2 mmol/kg injected by hand at a rate of 1-2 mL/min into each inguinal lymph node, liver lymphatics, and mesenteric lymphatics. Imaging analysis is done by pediatric radiologists that are trained in lymphatic imaging. These sequences as well as imaging parameters can be found outlined by Dori et al (3).

Another imaging technique that is utilized in conjunction with DCMRL is thoracic duct outlet patency. This is performed with US contrast via intranodal lymphangiography to assess the patency of the thoracic duct outlet using ultrasound contrast agents (4). Direct lymphangiography is utilized to map the central lymphatic system in cases where MRI cannot be utilized. Access to the central lymphatic system is accomplished via transabdominal access into the cisterna chyli where contrast is injected under fluoroscopy.

Lymphatic flow disorders can be categorized into four major categories based on whether the lymphatic dysfunction is secondary to a lymphatic leak, a lymphatic overproduction disorder, a lymphatic conduction disorder, or lymphedema. The lymphatic leak category includes traumatic lymphatic leaks and idiopathic leaks. There are overproduction and conduction abnormalities that result in organ-specific perfusion and therefore abnormalities including: heart (chylopericardium); liver and mesentery (protein losing enteropathy and/or ascites); kidneys (chyluria); lungs (pleural effusions, plastic bronchitis, chyloptysis, pulmonary edema, lung disease); tissue (edema, anasarca) *(Fig. 1)*.

There are different percutaneous and surgical interventional techniques to address the various lymphatic dysfunction(s) based upon the lymphatic imaging findings (5,6). Percutaneous embolization involves methods

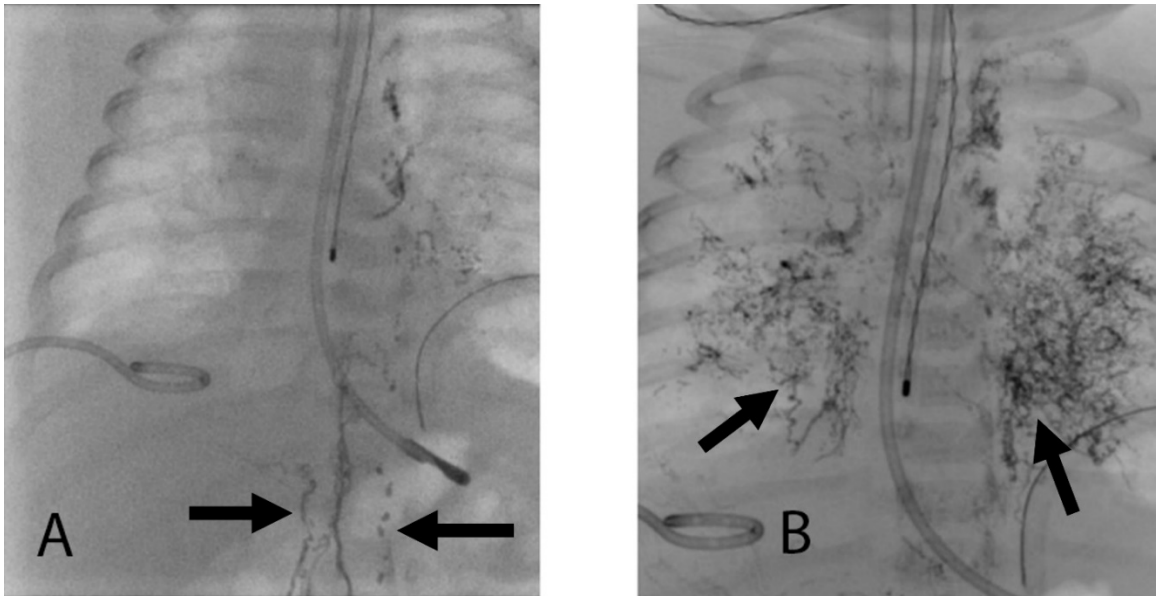

Fig. 2. *Neonatal Chylothorax Treatment with Lipiodol. A) Arrows depict flow of lipiodol up the central lymphatics following injection into the bilateral inguinal lymph nodes. B) Arrows highlight lipiodol after a delay period demonstrating transit out of the central system and into the abnormal pulmonary lymphatics. Images courtesy of Dr. Yoav Dori.*

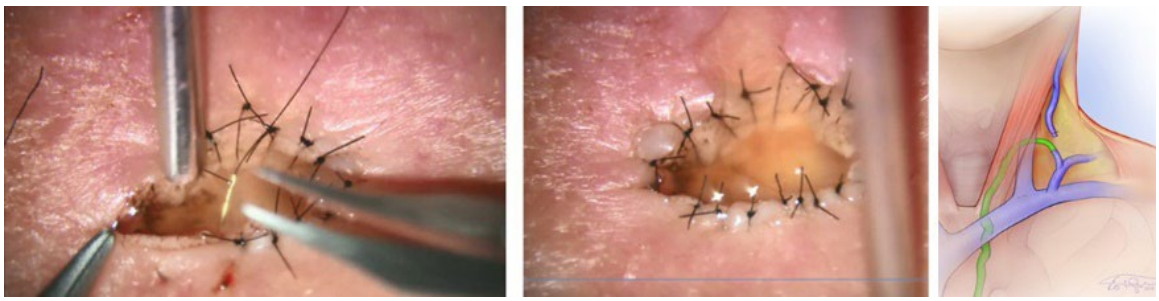

Fig. 3. *(left and middle) Intra-operative photograph of a lymphocutaneous anastomosis (LCA). (right) Diagram depicting a lymphovenous anastomosis (LVA) of the thoracic duct. Left and middle courtesy of Dr. Pablo Laje.*

such as selective lymphatic duct embolization (SLDE), full thoracic duct embolization (TDE), thoracic duct ligation (TDL), and lipiodol-based embolization. These embolization techniques are used to treat conditions such as chylothorax, chylopericardium, chyloptysis, and plastic bronchitis. Stent dilation or exclusion is performed to maintain or block lymphatic flow to a certain area. Balloon occlusion or dilation modulates lymphatic channels, while thoracic duct externalization and drainage are used to palliate symptoms associated with multicompartment lymphatic failure. These cases typically require temporary stabilization of the lymphatic system and the affected organs/tissues with external lymphatic drainage to decrease anasarca/edema. Manual occlusion and manipulation are direct intervention techniques to control lymphatic flow that can be utilized as well in the interventional suite. Treatment for neonatal chylothorax includes mostly lipiodol-based interventions *(Fig. 2)*. Congenital lymphatic flow disorder (CLFD) typically is managed with

MR lymphangiography, conservative medical management, thoracic duct externalization, and various lymphatic anastomosis surgical techniques (lymphovenous and lymphocutaneous) *(Fig. 3a &b).*

Interventions for edema and anasarca include MR lymphangiography with central and peripheral approaches for detailed imaging, conservative medical management using IVIG, albumin, diuretics, and weaning of positive pressure ventilation. Compression stockings and manual lymphatic drainage techniques with a certified lymphedema therapist is also utilized. Surgical ligation of channels with a microsurgeon or general surgeon trained in lymphatic surgeries may be performed to treat abnormal lymphatic perfusion/leakage. Hybrid surgical techniques with a microsurgeon and lymphatic interventionalist combine a thoracotomy, mechanical pleurodesis, or laparotomy with direct percutaneous embolization(s) of abnormal lymphatic leakage/channel(s) for treatment of persistent chylothoraces or chylous ascites. Cardiac interventions to improve lymphatic flow and in turn treat lymphatic dysfunction include: fenestration creation in the Fontan circulation patient, percutaneous or surgical thoracic duct decompression (TDD), embolization of collaterals and/or antegrade pulmonary blood flow, or thrombectomies aimed to relieve antegrade flow obstruction to blood or lymphatic fluid.

Multi-compartment imaging is essential for accurate diagnosis and treatment planning. SLDE is preferred, but even with SLDE there should be caution advised to avoid exacerbating lymphatic dysfunction in other compartments. The future of lymphatic management lies not only in phenotyping, but a goal of precision lymphology, which pairs the phenotyping to the genotyping for more accurate and tailored interventions. This comprehensive overview underscores the complexity, innovation and expertise that is involved in diagnosing and treating lymphatic disorders, showcasing the critical role of advanced imaging and targeted percutaneous and surgical interventions.

CONFLICT OF INTEREST AND DISCLOSURE

The author declares no competing financial interests exist.

REFERENCES

1. Biko, DM, CL Smith, HJ Otero, et al: Intrahepatic dynamic contrast MR lymphangiography: Initial experience with a new technique for the assessment of liver lymphatics. Eur. Radiol. 29 (2019), 5190–5196.
2. Dori, Y, MM Zviman, M Itkin: Dynamic contrast-enhanced MR lymphangiography: Feasibility study in swine. Radiology 273 (2014) 410-416.
3. Dori, Y: Novel lymphatic imaging techniques. Tech. Vasc. Interv. Radiol. 19 (2016) 255-261.
4. Mejia, EJ, HJ Otero, CL Smith, et al: Use of contrast-enhanced ultrasound to determine thoracic duct patency. J. Vasc. Interv. Radiol. 10 (2020), 1670-1674.
5. Pinto, E, Y Dori, C Smith, et al: Neonatal lymphatic flow disorders: impact of lymphatic imaging and interventions on outcomes. J. Perinat. 41 (2020), 494-501.
6. Dori, Y, CL Smith, AG DeWitt, et al: Intramesenteric dynamic contrast pediatric MR lymphangiography: Initial experience and comparison with intranodal and intrahepatic MR lymphangiography. Eur. Radiol. 30 (2020), 5777-5784.

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