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Post-Operative Chylothorax in Patients With Congenital Heart Disease

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ABSTRACT

BACKGROUND Post-operative chylothorax in patients with congenital heart disease is a challenging problem with substantial morbidity and mortality. Currently, the etiology of chylothorax is poorly understood and treatment options are limited.

OBJECTIVES This study aimed to report lymphatic imaging findings, determine the mechanism of chylothorax after cardiac surgery, and analyze the outcomes of lymphatic embolization.

METHODS We conducted a retrospective review of 25 patients with congenital heart disease and post-operative chylothorax who presented for lymphatic imaging and intervention between July 2012 and August 2016.

RESULTS Based on dynamic contrast-enhanced magnetic resonance lymphangiography and intranodal lymphangiography, we identified 3 distinct etiologies of chylothorax: 2 patients (8%) with traumatic leak from a thoracic duct (TD) branch, 14 patients (56%) with pulmonary lymphatic perfusion syndrome (PLPS), and 9 patients (36%) with central lymphatic flow disorder (CLFD), the latter defined as abnormal central lymphatic flow, effusions in more than 1 compartment, and dermal backflow. Patients with traumatic leak and PLPS were combined into 1 group of 16 patients without CLFD, of whom 14 (88%) had an intact TD. Sixteen patients underwent lymphatic intervention, including complete TD embolization. All 16 patients had resolution of chylothorax, with a median of 7.5 days from intervention to chest tube removal and 15 days from intervention to discharge. The 9 patients with CLFD were considered a separate group, of whom 3 (33%) had an intact TD. Seven patients underwent lymphatic intervention but none survived.

CONCLUSIONS Most patients in this study had nontraumatic chylothorax and dynamic contrast-enhanced magnetic resonance lymphangiography was essential to determine etiology. Lymphatic embolization was successful in patients with traumatic leak and PLPS and, thus, should be considered first-line treatment. Interventions in patients with CLFD were not successful to resolve chylothorax and alternate approaches need to be developed. (J Am Coll Cardiol 2017;69:2410–22) © 2017 by the American College of Cardiology Foundation.

Post-operative chylothorax in patients with congenital heart disease (CHD) is a challenging clinical problem with substantial morbidity and mortality (1). The incidence of chylothorax after cardiothoracic surgery has been reported between 2% and 5% (2). A recent analysis of the Pediatric Health Information System (PHIS) database found that the overall incidence of chylothorax in pediatric patients after congenital heart surgery or heart transplantation was 2.8%, with an increased incidence from 2.0% in 2004 to 3.7% in 2011 (3). In this PHIS cohort, the procedure codes associated with the highest incidence of chylothorax were cavopulmonary anastomoses (Glenn and Fontan surgeries), repair of transposition of the great arteries, and heart transplantation. Additionally, the development of chylothorax was associated with a significantly longer length of hospital stay ($p < 0.0001$), increased risk of

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in-hospital mortality (odds ratio: 2.13), and higher cost of hospitalization ($p < 0.0001$).

Multiple diagnostic algorithms for chylothorax have been developed and theories have been proposed to suggest the etiologies of traumatic and nontraumatic chylothorax (4). The cause of traumatic chylothorax is direct injury or surgical laceration of the central thoracic duct (TD) or 1 of its lymphatic tributaries. The causes of nontraumatic chylothorax have been reported to include lymphatic malformations, malignancy (e.g., lymphoma), infection (e.g., tuberculosis), extension from chylous ascites, and congenital syndromes (e.g., Down, Noonan, or Turner syndromes) (5).

SEE PAGE 2423

Management of chylothorax can be challenging and includes both conservative and interventional treatments. The goal of the conservative approach is to reduce intestinal lymphatic flow through dietary modifications (such as a low-fat diet or total parenteral nutrition) and medications (e.g., octreotide or somatostatin) (6,7). If conservative management fails, then surgical procedures such as TD ligation, pleurodesis, and pleuroperitoneal shunts are considered (8,9). The PHIS analysis reported that TD ligation or pleurodesis was performed on patients a median of 18 days (interquartile range [IQR]: 7 to 28 days) after the cardiac procedure and patients were discharged from the hospital a median of 22 days (IQR: 10 to 47 days) after surgical treatment of chylothorax (3). More recently, percutaneous TD embolization has emerged as a minimally invasive alternative for the treatment of chylothorax (10,11).

One of the difficulties in determining the etiology of chylothorax has been the lack of methods to image the central lymphatic system. Dynamic contrast-enhanced magnetic resonance lymphangiography (DCMRL) is a new imaging technique that uses an intranodal injection of gadolinium-based contrast agents to visualize the anatomy and flow characteristics of the central lymphatic system, with good spatial and temporal resolution (12,13). DCMRL recently showed abnormal pulmonary lymphatic flow from the TD toward the lung parenchyma and/or lymphatic perfusion of the mediastinum in patients with single ventricle physiology and plastic bronchitis; this was termed pulmonary lymphatic perfusion syndrome (PLPS) (14). Percutaneous embolization of this abnormal lymphatic flow resulted in symptom resolution for the majority of patients.

For several years, our institutional approach to post-operative chylothorax has been to perform magnetic resonance lymphangiography on all patients before any lymphatic intervention or procedure in order to define the lymphatic anatomy and determine the mechanism of chylothorax. The objective of this study was to report the lymphatic imaging findings and outcomes of percutaneous lymphatic embolization for the treatment of chylothorax in patients with CHD.

ABBREVIATIONS AND ACRONYMS

CHD = congenital heart disease

CLFD = central lymphatic flow disorder

IQR = interquartile range

MRI = magnetic resonance imaging

PLPS = pulmonary lymphatic perfusion syndrome

TD = thoracic duct

METHODS

This study is a retrospective analysis of patients with CHD and post-operative chylothorax who presented to our institution for lymphatic imaging and intervention between July 2012 and August 2016. Permission from our institutional review board was obtained before study initiation.

Data collection included patient demographics, cardiac diagnoses, surgical histories, prior therapies, weight-adjusted volume of chest tube drainage, imaging findings, results of lymphatic interventions, and the clinical course post-intervention. The diagnosis of chylothorax was established by the presence of a high percentage of lymphocytes (>70%) and/or a high concentration of triglycerides (in patients on a regular fat-containing diet) in the pleural fluid. In some cases, a high-fat food challenge test was performed and, if the concentration of triglycerides increased, then the diagnosis of chylothorax was confirmed.

The primary endpoints of the study were resolution of chylothorax and patient survival. The secondary endpoints were the weight-adjusted volume of chest tube drainage during the 7 days before intervention compared to the 7 days after intervention and the duration of chylothorax before intervention compared to the duration of chylothorax post-intervention (with resolution of chylothorax defined as removal of all chest tubes).

LYMPHATIC IMAGING AND INTERVENTION. Imaging and interventions were performed in an XMR suite that combines a magnetic resonance image (MRI) scanner with a cardiac catheterization laboratory. All procedures were performed under general anesthesia. Patients initially underwent DCMRL using the technique previously described by Dori et al. (14). Briefly, the inguinal lymph nodes were first accessed under ultrasound guidance in the catheterization laboratory, using a 25-gauge spinal needle. A small amount of water-soluble iodinated contrast agent was injected to confirm the position of the needle inside