

**American College of Radiology
ACR Appropriateness Criteria®
Chylothorax Treatment Planning**

Variant: 1 Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

Procedure	Appropriateness Category	Relative Radiation Level
Lymphangiography chest abdomen pelvis	Usually Appropriate	⊕⊕⊕
Lymphangiography chest and abdomen	Usually Appropriate	⊕⊕⊕⊕
MR lymphangiography chest abdomen pelvis	Usually Appropriate	○
MR lymphangiography chest and abdomen	Usually Appropriate	○
Radiography chest	May Be Appropriate	⊕
Radiography chest abdomen pelvis	May Be Appropriate	⊕⊕⊕
MRI chest without and with IV contrast	May Be Appropriate	○
MRI chest without IV contrast	May Be Appropriate	○
Lymphoscintigraphy chest abdomen pelvis	May Be Appropriate	⊕⊕
Lymphoscintigraphy chest and abdomen	May Be Appropriate	⊕⊕
CT chest with IV contrast	May Be Appropriate	⊕⊕⊕⊕
CT chest without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕
CT chest without IV contrast	May Be Appropriate	⊕⊕⊕⊕
CT chest abdomen pelvis with IV contrast	May Be Appropriate	⊕⊕⊕⊕⊕
CT chest abdomen pelvis without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕⊕
CT chest abdomen pelvis without IV contrast	May Be Appropriate	⊕⊕⊕⊕⊕
US chest	Usually Not Appropriate	○
US chest abdomen pelvis	Usually Not Appropriate	○

Variant: 2 Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

Procedure	Appropriateness Category	Relative Radiation Level
Lymphangiography chest abdomen pelvis	Usually Appropriate	⊕⊕⊕
Lymphangiography chest and abdomen	Usually Appropriate	⊕⊕⊕⊕
MR lymphangiography chest abdomen pelvis	Usually Appropriate	○
MR lymphangiography chest and abdomen	Usually Appropriate	○
Radiography chest	May Be Appropriate	⊕
Radiography chest abdomen pelvis	May Be Appropriate	⊕⊕⊕
MRI chest without and with IV contrast	May Be Appropriate	○
MRI chest without IV contrast	May Be Appropriate	○
Lymphoscintigraphy chest abdomen pelvis	May Be Appropriate	⊕⊕
Lymphoscintigraphy chest and abdomen	May Be Appropriate	⊕⊕
CT chest with IV contrast	May Be Appropriate	⊕⊕⊕⊕
CT chest without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕
CT chest abdomen pelvis with IV contrast	May Be Appropriate	⊕⊕⊕⊕⊕
CT chest abdomen pelvis without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕⊕

CT chest abdomen pelvis without IV contrast	May Be Appropriate	
US chest	Usually Not Appropriate	
US chest abdomen pelvis	Usually Not Appropriate	
CT chest without IV contrast	Usually Not Appropriate	

Panel Members

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Summary of Literature Review

Introduction/Background

Chyle is primarily formed in the intestines and is composed of proteins, lipids, electrolytes, and lymphocytes. A chylous pleural effusion, or chylothorax, is a highly morbid condition defined by the presence of chyle within the pleural space. A chronic chyle leak results in metabolic abnormalities, respiratory compromise, immunosuppression, malnutrition, and even death [1-3]. Chylothoraces can be categorized etiologically as traumatic or nontraumatic. Collectively, the incidence of chylothorax is approximately 1 per 6,000 admissions [1]. Historically, nontraumatic etiologies accounted for up to 72% of cases. Most recently, the largest study reports that traumatic etiologies account for 54% of cases [1,4-7]. The discrepancy may reflect the growth in thoracic oncologic resections or specific referral patterns.

Diagnosis: Chylothorax most commonly presents with dyspnea, although chest pain, fever, and fatigue may also occur. Chyle is odorless, alkaline, sterile, and milky in appearance, although the appearance may vary based on the nutritional status of the patient. Increasing fatty intake increases the volume and can change the color of the fluid and has been described for the diagnosis of a chyle leak. The hallmark of chylous effusion is the presence of chylomicrons in the fluid. Objective diagnostic criteria include a pleural fluid triglyceride level >110 mg/dL and a ratio of pleural fluid to serum triglyceride level of >1.0 . A ratio of pleural fluid to serum cholesterol level of <1.0 distinguishes chylothorax from cholesterol pleural effusions, which may present similarly [2,3].

Management: The diagnosis is confirmed by draining the fluid for studies; this is also palliative. After replacing fluid and protein losses, a decision about conservative versus invasive therapies can be made. If the chylothorax reaccumulates, treatment is guided by daily outputs, with higher outputs warranting a more aggressive approach [2,4,8-11].

Conservative measures include management of the underlying disease, thoracentesis, and dietary modifications such as total parenteral nutrition or a nonfat diet to reduce production of chyle and consequently flow through the thoracic duct. Adjunctive therapy may include somatostatin, etilefrine, or nitric oxide, with the underlying etiology determining the efficacy, although the evidence remains scarce. The success of conservative therapy approaches 50% in nonmalignant

etiologies but is only minimally beneficial in neoplastic etiologies [2,8,11].

Exact criteria for the implementation of invasive treatment are not well defined, but several authors advocate its use if conservative treatment has not resolved the chylothorax after 2 weeks or in higher-output chylothoraces. Invasive treatments include surgical thoracic duct ligation, pleurodesis, and thoracic duct embolization (TDE) [2,4,8-11]. Less commonly, tunneled drains or pleural shunt procedures are performed, although prolonged drainage is not recommended as a long-term option because of an increased risk of complications [12,13]. In chylothorax related to underlying malignancy, indwelling pleural catheters have been used without significant increase in infection or albumin levels [14,15]. Although the technical success of direct surgical ligation is high, these debilitated patients are at an increased risk for postoperative adhesions, infection, and poor wound healing. Reported postoperative mortality rates for patients who have failed conservative management range from 4.5% to as high as 50% [2,4,9,10].

TDE is a percutaneous alternative to thoracic duct ligation. TDE allows for direct embolization (type I) or needle disruption of the thoracic duct (type II). Whereas the former directly treats the focus of injury, the latter is purported to create a controlled leak and inflammatory reaction in the retroperitoneum, which collateralizes and diverts flow from the thoracic duct. Over several successive publications, Cope et al [16,17] defined the technique and reported its feasibility. The initial series of 42 patients by Cope and Kaiser [18] revealed effective percutaneous treatment in >70% of cases. In 109 patients with traumatic thoracic duct leak, Itkin et al [5] reported 90% clinical resolution postembolization and 72% clinical resolution of the chyle leak with thoracic duct disruption. A subsequent recent study by Gurevich et al [7] reported an overall clinical success rate of 97% after TDE for nontraumatic chylous effusions in 31 patients. Pamarthi et al [6] reported an 85% technical success rate and a 64% clinical success rate in 105 patients with all-cause chylous leaks. Additional series have yielded similar results [19-22]. A recent published study of 355 patients by Pan et al [23] reported an 88.5% technical success and a 61.6% clinical success rate for therapeutic lymphangiography alone with all cause leaks. Collectively, TDE has higher clinical success treating traumatic compared with nontraumatic chyle leaks and when compared with thoracic duct disruption [8,9,11,24]. Overall, acute complications associated with TDE are minor and generally self-limited and are estimated at 2% to 6% [5-7,25]. Long-term complications may be seen in up to 14% of patients and may include leg swelling, abdominal swelling, or chronic diarrhea [26].

Special Imaging Considerations

MR Lymphangiography Chest and Abdomen: MR lymphangiography chest and abdomen with intralymphatic contrast involves the use of gadolinium-based contrast material injection within the groin lymph nodes (intranodal) or in the web spaces between toes. Following the contrast material injection, patients are imaged with MRI. High image quality of lymph nodes, central lymphatics, and flow patterns within the lymphatics has been described, with more recent studies further demonstrating the characteristics and outcomes of MR lymphangiography with intranodal or transpedal gadolinium-based contrast [27-33]. Visualization of the cisterna chyli, thoracic duct, and tributary lymphatic vessels with MRI was described in healthy volunteers as early as 1999 [34]. Initial MR lymphangiography technique involved unenhanced thin-collimated axial and coronal sequences similar to MR cholangiopancreatography. Further refinements of sequences, particularly heavily T2-weighted sequences with and without intravenous (IV) contrast and fat suppression, pre- and postcontrast fat-saturated T1, and black-blood fat-saturated turbo spin-echo T2,

combined with 3-D techniques, maximum-intensity projections, and higher magnetic fields, increased the reliability and quality of MR lymphangiography [35-40]. These heavily T2-weighted sequences, however, are susceptible to artifacts secondary to movement (breathing, pulsing artery, and heart contractions), proximity to surgical lines and drains, and air within the lung. A new acquisition technique, controlled aliasing in parallel imaging results in higher acceleration (CAIPIRINHA), has also contributed to the improved image quality of these heavily T2-weighted sequences in noncontrast MR lymphangiography because it does not require the patient to perform breath-holds [40-42]. Lastly, this technique is often performed immediately after ingestion of a high-fat-content food, such as olive oil, to improve lymphatic flow and visualization [40-42].

Discussion of Procedures by Variant

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

Traumatic chylothoraces are a result of direct injury to thoracic lymphatics. Iatrogenic traumatic chylothorax complicates up to 3.9% of general thoracic surgeries resections [1,2,4-7,43]. Lung cancer resections, cardiovascular surgeries, and spinal surgeries can also be complicated by chylothorax, although at a lesser rate. Noniatrogenic causes of traumatic chylothorax include penetrating trauma, fracture-dislocation of the spine, and hyperflexion injuries [1,6,7]. Generally, the causative etiology is known in the traumatic setting. Imaging a patient with a known traumatic chylothorax serves only to confirm the diagnosis and assist in therapeutic planning.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

A. CT Chest With IV Contrast

CT imaging is able to visualize portions of the lymphatic system but provides less anatomic detail than MRI [36,44,45]. If the etiology is known, CT of the chest with IV contrast has little value, in that it does not help guide therapy directed at chylothorax in most cases. No evidence is present to suggest a role for IV contrast. However, CT chest with IV contrast may be useful for treatment planning in certain clinical situations such as patients with connective tissue disease, Marfan disease with an aortic pathology, celiac artery aneurysm, or other anatomic abnormalities and in postoperative patients.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

B. CT Chest Without and With IV Contrast

CT imaging is able to visualize portions of the lymphatic system but provides less anatomic detail than MRI [36,44,45]. Studies with 1 mm collimation and multiplanar reformation were able to identify the thoracic duct and cisterna chyli in nearly 100% of CT scans with normal anatomy [46]. If the etiology is known, CT of the chest with and without IV contrast has little value, in that it does not help guide therapy directed at chylothorax in most cases. No evidence is present to suggest a role for IV contrast. However, CT chest with IV contrast may be useful for treatment planning in certain clinical situations such as patients with connective tissue disease, Marfan disease with an aortic pathology, celiac artery aneurysm, or other anatomic abnormalities and in postoperative patients.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

C. CT Chest Without IV Contrast

CT imaging is able to visualize portions of the lymphatic system but provides less anatomic detail than MRI [36,44,45]. One series showed that the combination of CT and unilateral pedal lymphangiography was able to identify the cause and locate the leak in 75% of idiopathic chylothoraces after failure of thoracic duct ligation [47]. Moreover, in this series of 24 patients, the lack of thoracic duct leakage was managed with nonoperative therapy with higher success rates [47]. Older studies noted that noncontrast CT visualizes the cisterna chyli in 1.7% of cases and could differentiate this from adjacent anatomy by its low attenuation, continuity with the thoracic duct, and tubular nature [45]. At least some portion of the thoracic duct was visualized in 55% of patients in a different series [44]. If the etiology is known, CT of the chest without IV contrast has little value, in that it does not help guide therapy directed at chylothorax in most cases.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

D. CT Chest Abdomen Pelvis With IV Contrast

CT imaging is able to visualize portions of the lymphatic system but provides less anatomic detail than MRI [36,44,45]. If the etiology is known, CT of the chest with IV contrast has little value, in that it does not help guide therapy directed at chylothorax in most cases. No evidence is present to suggest a role for IV contrast. However, CT chest with IV contrast may be useful for treatment planning in certain clinical situations such as patients with connective tissue disease, Marfan's disease with an aortic pathology, celiac artery aneurysm, or other anatomic abnormalities and in postoperative patients. For treatment planning purposes, it may also be helpful and advisable to image the abdomen and pelvis in addition to the chest for further anatomical evaluation.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

E. CT Chest Abdomen Pelvis Without and With IV Contrast

CT imaging is able to visualize portions of the lymphatic system but provides less anatomic detail than MRI [36,44,45]. Studies with 1 mm collimation and multiplanar reformation were able to identify the thoracic duct and cisterna chyli in nearly 100% of CT scans with normal anatomy [46]. If the etiology is known, CT of the chest with and without IV contrast has little value, in that it does not help guide therapy directed at chylothorax in most cases. No evidence is present to suggest a role for IV contrast. However, CT chest with IV contrast may be useful for treatment planning in certain clinical situations such as patients with connective tissue disease, Marfan's disease with an aortic pathology, celiac artery aneurysm, or other anatomic abnormalities and in postoperative patients. For treatment planning purposes, it may be helpful and advisable to image the abdomen and pelvis in addition to the chest for further anatomical evaluation.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

F. CT Chest Abdomen Pelvis Without IV Contrast

CT imaging is able to visualize portions of the lymphatic system but provides less anatomic detail than MRI [36,44,45]. One series showed that the combination of CT and unilateral pedal lymphangiography was able to identify the cause and locate the leak in 75% of idiopathic chylothoraces after failure of thoracic duct ligation [47]. Moreover, in this series of 24 patients, the lack of thoracic duct leakage was managed with nonoperative therapy with higher success rates [47]. Older studies noted that noncontrast CT visualizes the cisterna chyli in 1.7% of cases and could differentiate this from adjacent anatomy by its low attenuation, continuity with the thoracic duct, and tubular nature [45]. At least some portion of the thoracic duct was visualized in 55% of

patients in a different series [44]. If the etiology is known, CT of the chest without IV contrast has little value, in that it does not help guide therapy directed at chylothorax in most cases. For treatment planning purposes, it is advisable to image the abdomen and pelvis in addition to the chest and may be helpful.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

G. Lymphangiography Chest and Abdomen

Conventional lymphangiography of the chest and abdomen can provide a visualization of lymph nodes, lymphatic vessels, cisterna chyli, the thoracic duct, and sites of injury [16,48,49].

Lymphangiography alone has been shown to be therapeutic in a sizable percentage of patients, irrespective of attempts at TDE or disruption, with efficacy reported to be 61.6% in a recent study of 355 patients [23,50-54]. Additionally, therapeutic lymphangiography had a similar rate of chylothorax resolution compared with lymphangiography followed by TDE (71.4% versus 90.5%) [55]. When performed as a prelude to TDE, the combination is particularly effective in treating traumatic chylothorax, with technical and clinical success rates approaching 90% [5-9,11,19-22,25,56].

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

H. Lymphangiography Chest Abdomen Pelvis

Conventional lymphangiography of the chest, abdomen, and pelvis can provide an even more complete visualization of lymph nodes, lymphatic vessels, cisterna chyli, the thoracic duct, and sites of injury than just lymphangiography of the chest and abdomen [16,48,49]. Lymphangiography alone has been shown to be therapeutic in a sizable percentage of patients, irrespective of attempts at TDE or disruption, with efficacy reported to be 61.6% in a recent study of 355 patients [23,50-54]. Additionally, therapeutic lymphangiography had a similar rate of chylothorax resolution compared with lymphangiography followed by TDE (71.4% versus 90.5%) [55]. When performed as a prelude to TDE, the combination is particularly effective in treating traumatic chylothorax, with technical and clinical success rates approaching 90% [5-9,11,19-22,25,56].

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

I. Lymphoscintigraphy Chest and Abdomen

Nuclear lymphoscintigraphy of the chest and abdomen can be used to confirm a lymphatic leak and identify the site, particularly if used with 3-D single-photon emission CT/CT techniques that has a sensitivity of 88% and a specificity of 100%, but little evidence is present to support its routine use [57-60]. Moreover, this adds little to the clinical care of a patient, because the traumatic etiology is usually known and any information gained would be redundant if conventional lymphangiography was performed as part of TDE.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

J. Lymphoscintigraphy Chest Abdomen Pelvis

Nuclear lymphoscintigraphy of the chest, abdomen, and pelvis can be used to confirm a lymphatic leak and identify the site, particularly if used with 3-D single-photon emission CT/CT techniques that has a sensitivity of 88% and a specificity of 100%. Although little evidence is present to support its routine use, this procedure may still be helpful in this clinical scenario if the traumatic leak is known and may still help localize the leak [57-60].

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

K. MR Lymphangiography Chest and Abdomen

MR lymphangiography chest and abdomen with intranodal gadolinium injection and dynamic acquisition of MR images has led to a new technique, dynamic contrast-enhanced MR lymphangiography (DCMRL). Compared with noncontrast MRI, DCMRL yields high signal intensity within the lymphatic vessels. Unlike conventional lymphangiography, DCMRL provides 3-D dynamic volumetric lymphatic flow data over time that aid in identifying lymphatic leaks [27,31]. MR lymphangiography with transpedal gadolinium injection has been shown to be successful in visualizing lymphatic anatomy, including variations or lymphatic pathologies in 23 of 25 (92%) patients [30]. This technique is a minimally invasive imaging technique that can be performed easily in a postoperative patient with suspected chyle leak to aid in interventional planning.

Initial studies showed that MR lymphangiography without contrast can visualize the lymphatic system [37]. More recent studies have demonstrated that the CAIPIRINHA acquisition technique in heavily T2-weighted sequences and oral administration of high-fat foods (olive oil) have significantly improved the image quality of noncontrast MR lymphangiography [40-42]. Although noncontrast MR lymphangiography has limited resolution in imaging the central lymphatic system and is poor at depicting anatomic details, it is highly effective in detecting postoperative leakage, with a 100% sensitivity and 97.1% specificity [40,41]. This technique is a noninvasive imaging technique that can be performed easily in a postoperative patient with suspected chyle leak before intervention.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

L. MR Lymphangiography Chest Abdomen Pelvis

MR lymphangiography chest, abdomen, and pelvis with intranodal gadolinium injection and dynamic acquisition of MR images has led to a new technique, DCMRL. Compared with noncontrast MRI, DCMRL yields high signal intensity within the lymphatic vessels. Unlike conventional lymphangiography, DCMRL provides 3-D dynamic volumetric lymphatic flow data over time that aid in identifying lymphatic leaks [27,31]. MR lymphangiography with transpedal gadolinium injection has been shown to be successful in visualizing lymphatic anatomy, including variations or lymphatic pathologies in 23 of 25 (92%) patients [30]. This technique is a minimally invasive imaging technique that can be performed easily in a postoperative patient with suspected chyle leak to aid in interventional planning.

Initial studies showed that MR lymphangiography without contrast can visualize the lymphatic system [37]. More recent studies have demonstrated that the CAIPIRINHA acquisition technique in heavily T2-weighted sequences and oral administration of high-fat foods (olive oil) have significantly improved the image quality of noncontrast MR lymphangiography [40-42]. Although noncontrast MR lymphangiography has limited resolution in imaging the central lymphatic system and is poor at depicting anatomic details, it is highly effective in detecting postoperative leakage, with a 100% sensitivity and 97.1% specificity [40,41]. This technique is a noninvasive imaging technique that can be performed easily in a postoperative patient with suspected chyle leak before intervention.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

M. MRI Chest Without and With IV Contrast

The diagnostic benefit of MRI is negated in the setting of traumatic chylothoraces. However, the ability of MRI to map the lymphatic system can be of benefit in select cases in which identifying the cisterna chyli and/or thoracic duct is difficult or conventional lymphangiography is unsuccessful [61-64].

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

N. MRI Chest Without IV Contrast

The diagnostic benefit of MRI is negated in the setting of traumatic chylothoraces. However, the ability of MRI to map the lymphatic system can be of benefit in select cases in which identifying the cisterna chyli and/or thoracic duct is difficult or conventional lymphangiography is unsuccessful [61-64]. A recent study of 29 patients investigated the use of MRI in characterizing chylous versus nonchylous effusions using multipoint Dixon fat quantification that yielded a sensitivity of 82% and a specificity of 100% [65]. MRI without IV contrast can provide useful information; however, newer techniques such as MR lymphangiography with or without IV contrast provide higher resolution and more valuable lymphatic information.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

O. Radiography Chest

In the setting of a traumatic injury to the thoracic duct, most commonly postoperative or mechanical trauma, chest radiographs can confirm the presence of pleural fluid and lateralize the process and are routinely acquired in the daily evaluation of supportive lines and tubes. Although this technique cannot reliably characterize the type of effusion and is of little diagnostic value [66,67], it can be used as a rapid and simple first test for treatment planning in determining laterality in a postoperative patient.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

P. Radiography Chest Abdomen Pelvis

In the setting of a traumatic injury to the thoracic duct, most commonly postoperative or mechanical trauma, chest radiographs can confirm the presence of pleural fluid and lateralize the process and are routinely acquired in the daily evaluation of supportive lines and tubes. Radiography cannot reliably characterize the type of effusion and is of little diagnostic value [66,67]. There is no evidence supporting the addition of abdominal and pelvic radiographs in the treatment planning of chylothorax.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

Q. US Chest

Ultrasound (US) reliably detects pleural fluid but cannot definitively discriminate between the types of pleural effusion and provides minimal additional information to narrow the differential diagnosis [68]. US can be helpful in the guidance of thoracentesis. However, US has little, if any, diagnostic role in the setting of a known traumatic chylothorax.

Variant 1: Adult or child. Chylothorax: traumatic or iatrogenic etiology. Pretreatment planning.

R. US Chest Abdomen Pelvis

US reliably detects pleural fluid but cannot definitively discriminate between the types of pleural effusion and provides minimal additional information to narrow the differential diagnosis [68]. US can be helpful in the guidance of thoracentesis and intranodal injection during conventional and MR lymphangiography [31,69]. However, US has little, if any, diagnostic role in the setting of a known traumatic chylothorax.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

Nontraumatic chylothorax accounts for approximately 46% of chylothoraces and can be subcategorized as resulting from malignancy, because it occurs in 18% of all chylothoraces, or nonmalignant etiologies, which account for 28% of all chylothoraces [1,2,6,7]. Of the malignant etiologies, lymphoma is the leading cause, accounting for 75% of all malignant chylothoraces. Nonmalignant, nontraumatic chylothorax has been described in lymphangioleiomyomatosis, sarcoidosis, cirrhosis, heart failure, nephrotic syndrome, venous thrombosis, filariasis, venolymphatic malformations, and a variety of other congenital, idiopathic, and systemic diseases. Approximately 9% of all chylous effusions are idiopathic [1,2,6,7]. Imaging a patient with either a nontraumatic chylothorax or a chylothorax of unknown etiology serves to narrow the differential diagnosis, further characterize the underlying cause and its severity, and assist in treatment planning.

Most patients with nontraumatic chylothoraces or chylothoraces of unknown etiologies present with acute respiratory illness (ARI), which consists of one or more of the following: cough, sputum production, chest pain, or dyspnea (with or without fever). The evaluation of ARI has been addressed by the ACR, and the imaging evaluation includes chest radiography and chest CT [70,71]. The consistent finding of chylothorax on initial imaging is the presence of a pleural effusion, which is a common medical problem with >50 recognized causes [72]. Pleural fluid studies are necessary for definitive diagnosis and to narrow the etiology of chylothorax.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

A. CT Chest With IV Contrast

Although CT imaging is inferior to MRI in visualizing lymphatics, it is a highly sensitive and specific examination to narrow a broader differential diagnosis of thoracic and abdominal pathology and is a rapid examination that is easily tolerated by a supine patient [36,70,71]. The addition of IV contrast accurately defines vascular and mediastinal structures and provides information on enhancement characteristics, which is a consideration when the etiology of chylothorax is unknown.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

B. CT Chest Without and With IV Contrast

Although CT imaging is inferior to MRI in visualizing lymphatics, it is a highly sensitive and specific examination to narrow a broader differential diagnosis of thoracic and abdominal pathology and is a rapid examination that is easily tolerated by a supine patient [36,70,71]. Moreover, studies with 1 mm collimation and multiplanar reformation were able to identify the thoracic duct and cisterna chyli in nearly 100% of CT scans with normal anatomy [46]. The addition of IV contrast accurately defines vascular and mediastinal structures and provides information on enhancement characteristics, which is a consideration when the etiology of chylothorax is unknown.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.**C. CT Chest Without IV Contrast**

One series showed that the combination of CT and unilateral pedal lymphangiography was able to identify the cause and locate the leak in 75% of idiopathic chylothoraces after failure of thoracic duct ligation [47]. Moreover, in this series of 24 patients, the lack of thoracic duct leakage was managed with nonoperative therapy with higher success rates [47]. Older studies noted that noncontrast CT visualizes the cisterna chyli in 1.7% of cases and could differentiate this from adjacent anatomy by its low attenuation, continuity with the thoracic duct, and tubular nature [45]. At least some portion of the thoracic duct was visualized in 55% of patients in a different series [44]. Although CT imaging is inferior to MRI in visualizing lymphatics, it is a highly sensitive and specific examination to narrow a broader differential diagnosis of thoracic and abdominal pathology and is a rapid examination that is easily tolerated by a supine patient [36,70,71]. However, CT imaging without contrast alone may not be as helpful in this clinical scenario.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.**D. CT Chest Abdomen Pelvis With IV Contrast**

Although CT imaging is inferior to MRI in visualizing lymphatics, it is a highly sensitive and specific examination to narrow a broader differential diagnosis of thoracic and abdominal pathology and is a rapid examination that is easily tolerated by a supine patient [36,70,71]. The addition of IV contrast accurately defines vascular and mediastinal structures and provides information on enhancement characteristics, which is a consideration when the etiology of chylothorax is unknown. For treatment planning purposes, it is advisable to image the abdomen and pelvis in addition to CT chest with IV contrast and may be helpful in cases such as malignancy.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.**E. CT Chest Abdomen Pelvis Without and With IV Contrast**

Although CT imaging is inferior to MRI in visualizing lymphatics, it is a highly sensitive and specific examination to narrow a broader differential diagnosis of thoracic and abdominal pathology and is a rapid examination that is easily tolerated by a supine patient [36,70,71]. Moreover, studies with 1 mm collimation and multiplanar reformation were able to identify the thoracic duct and cisterna chyli in nearly 100% of CT scans with normal anatomy [46]. The addition of IV contrast accurately defines vascular and mediastinal structures and provides information on enhancement characteristics, which is a consideration when the etiology of chylothorax is unknown. For treatment planning purposes, it is advisable to image the abdomen and pelvis in addition to the chest with IV contrast and may be helpful in cases such as malignancy.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.**F. CT Chest Abdomen Pelvis Without IV Contrast**

One series showed that the combination of CT and unilateral pedal lymphangiography was able to identify the cause and locate the leak in 75% of idiopathic chylothoraces after failure of thoracic duct ligation [47]. Moreover, in this series of 24 patients, the lack of thoracic duct leakage was managed with nonoperative therapy with higher success rates [47]. Older studies noted that noncontrast CT visualizes the cisterna chyli in 1.7% of cases and could differentiate this from adjacent anatomy by its low attenuation, continuity with the thoracic duct, and tubular nature [45].

At least some portion of the thoracic duct was visualized in 55% of patients in a different series [44]. Although CT imaging is inferior to MRI in visualizing lymphatics, it is a highly sensitive and specific examination to narrow a broader differential diagnosis of thoracic and abdominal pathology and is a rapid examination that is easily tolerated by a supine patient [36,70,71].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

G. Lymphangiography Chest and Abdomen

Conventional lymphangiography can provide a visualization of lymph nodes, lymphatic vessels, cistern a chyli, and the thoracic duct and for detection of lymphatic leakage [16,48,49]. In a nontraumatic or idiopathic chylothorax, conventional lymphangiography may help diagnose lymphatic vessel diseases and anatomic abnormalities and prevent unnecessary procedures. However, compared with traumatic chylothorax and particularly in the setting of a systemic disease, conventional lymphangiography does not always elucidate the underlying etiology. Patients presenting with nontraumatic leaks found during lymphangiography followed by TDE had a clinical success rate of 82% [21]. Furthermore, a more recent study by Gurevich et al [7] reported an overall clinical success rate of 97% after TDE for nontraumatic chylous effusions in 31 patients. Lastly, additional studies have shown the ineffectiveness of TDE in central lymphatic flow disorder (CLFD) and its associated increased of mortality [32,73].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

H. Lymphangiography Chest Abdomen Pelvis

Conventional lymphangiography of the chest, abdomen, and pelvis can provide a visualization of lymph nodes, lymphatic vessels, cisterna chyli, and the thoracic duct and for detection of lymphatic leakage [16,48,49]. In a nontraumatic or idiopathic chylothorax, conventional lymphangiography may help diagnose lymphatic vessel diseases and anatomic abnormalities and prevent unnecessary procedures. However, compared with traumatic chylothorax and particularly in the setting of a systemic disease, conventional lymphangiography does not always elucidate the underlying etiology. Patients presenting with nontraumatic leaks found during lymphangiography followed by TDE had a clinical success rate of 82% [21]. Furthermore, a more recent study by Gurevich et al [7] reported an overall clinical success rate of 97% after TDE for nontraumatic chylous effusions in 31 patients. Lastly, additional studies have shown the ineffectiveness of TDE in CLFD and its associated increased of mortality [32,73].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

I. Lymphoscintigraphy Chest and Abdomen

Nuclear lymphoscintigraphy has only a few reports showing that it is able to localize the site of chylous leak, particularly if used with 3-D single-photon emission CT/CT techniques that have a sensitivity of 88% and a specificity of 100% [57-60]. However, the localizing information gained would be redundant if conventional lymphangiography was performed as part of TDE.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

J. Lymphoscintigraphy Chest Abdomen Pelvis

Nuclear lymphoscintigraphy of the chest, abdomen, and pelvis has only a few reports showing that it is able to localize the site of chylous leak, particularly if used with 3-D single-photon emission

CT/CT techniques that have a sensitivity of 88% and a specificity of 100% [57-60]. However, the localizing information gained would be redundant if conventional lymphangiography was performed as part of TDE.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

K. MR Lymphangiography Chest and Abdomen

MR lymphangiography with intranodal gadolinium injection and dynamic acquisition of MR images has led to a new technique, DCMRL. Compared with noncontrast MRI, DCMRL yields high signal intensity within the lymphatic vessels. Unlike conventional lymphangiography, DCMRL provides 3-D dynamic volumetric lymphatic flow data over time that aid in identifying the location of lymphatic pathologies [27,31]. DCMRL is key in determining the etiology of nontraumatic chylothorax and the associated lymphatic flow abnormalities, such as CLFD and pulmonary lymphatic perfusion syndrome, because lymphatic embolization was shown to be successful in resolving chylothorax in patients with pulmonary lymphatic perfusion syndrome but not in CLFD [32]. MR lymphangiography with transpedal gadolinium injection has been shown to be successful in visualizing lymphatic anatomy, including variations or lymphatic pathologies in 23 out of 25 (92%) patients [30]. More recently, intrahepatic MR lymphangiography has been utilized to image liver lymphatics in patients with normal central lymphatic imaging, and has yielded abnormal lymphatic imaging patterns that correlate with symptoms such as chylothorax [33].

Initial studies showed that MR lymphangiography without contrast can visualize the lymphatic system [37]. More recent studies have demonstrated that the CAIPIRINHA acquisition technique in heavily T2-weighted sequences and prior oral administration of olive oil have significantly improved the image quality of noncontrast MR lymphangiography [40-42]. Although noncontrast MR lymphangiography has limited resolution in imaging the central lymphatic system and is poor at depicting anatomic details, it is highly effective in detecting postoperative leakage with a 100% sensitivity and a 97.1% specificity [40,41].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

L. MR Lymphangiography Chest Abdomen Pelvis

MR lymphangiography of the chest, abdomen, and pelvis with intranodal gadolinium injection and dynamic acquisition of MR images has led to a new technique, DCMRL. Compared with noncontrast MRI, DCMRL yields high signal intensity within the lymphatic vessels. Unlike conventional lymphangiography, DCMRL provides 3-D dynamic volumetric lymphatic flow data over time that aid in identifying the location of lymphatic pathologies [27,31]. DCMRL is key in determining the etiology of nontraumatic chylothorax and the associated lymphatic flow abnormalities, such as CLFD and pulmonary lymphatic perfusion syndrome, as lymphatic embolization was shown to be successful in resolving chylothorax in patients with pulmonary lymphatic perfusion syndrome but not in CLFD [32]. MR lymphangiography with transpedal gadolinium injection has been shown to be successful in visualizing lymphatic anatomy, including variations or lymphatic pathologies in 23 out of 25 (92%) patients [30]. More recently, intrahepatic MR lymphangiography has been used to image liver lymphatics in patients with normal central lymphatic imaging, and has yielded abnormal lymphatic imaging patterns that correlate with symptoms such as chylothorax [33].

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system [37]. More recent studies have demonstrated that the CAIPIRINHA acquisition technique in heavily T2-weighted sequences and prior oral administration of olive oil have significantly improved the image quality of noncontrast MR lymphangiography [40-42]. Although noncontrast MR lymphangiography has limited resolution in imaging the central lymphatic system and is poor at depicting anatomic details, it is highly effective in detecting postoperative leakage, with a 100% sensitivity and a 97.1% specificity [40,41].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

M. MRI Chest Without and With IV Contrast

MRI can accurately visualize lymphatic structures without IV contrast, depicting abnormal lymphatic malformations. Moreover, with the addition of IV contrast, MRI can then characterize mediastinal masses, pleural-based lesions, chest wall pathology, and etiology. However, thoracic MRI has limited utility for parenchymal lung pathology [35-39].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

N. MRI Chest Without IV Contrast

MRI accurately visualizes lymphatic structures without IV contrast, depicting abnormal lymphatic malformations. However, thoracic MRI has limited utility for parenchymal lung pathology [35-39]. A recent study of 29 patients investigated the use of MRI in characterizing chylous versus nonchylous effusions through the use of multipoint Dixon fat quantification that yielded a sensitivity of 82% and a specificity of 100% [65].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

O. Radiography Chest

Chest radiographs can reliably detect and confirm the presence of pleural fluid and lateralize the process and are routinely acquired in the daily evaluation of supportive lines and tubes. This technique cannot reliably characterize the type of effusion and is of little diagnostic value [66,67].

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

P. Radiography Chest Abdomen Pelvis

Chest radiographs can reliably detect and confirm the presence of pleural fluid and lateralize the process and are routinely acquired in the daily evaluation of supportive lines and tubes. This technique cannot reliably characterize the type of effusion and is of little diagnostic value [66,67]. However, the addition of abdomen and pelvis radiographs may help indirectly provide clues as to the etiology of the chylothorax.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

Q. US Chest

US reliably detects pleural fluid but cannot definitively discriminate between the types of pleural effusion and provides minimal additional information to narrow the differential diagnosis [68]. US can be helpful as adjunctive imaging in the guidance of thoracentesis.

Variant 2: Adult or child. Chylothorax: nontraumatic or unknown etiology. Pretreatment planning.

R. US Chest Abdomen Pelvis

US reliably detects pleural fluid but cannot definitively discriminate between the types of pleural effusion and provides minimal additional information to narrow the differential diagnosis [68]. US can be helpful as adjunctive imaging to be used in the guidance of thoracentesis and intranodal injection during conventional and MR lymphangiography [31,69].

Summary of Highlights

This is a summary of the key recommendations from the variant tables. Refer to the complete narrative document for more information.

- **Variant 1:** Lymphangiography chest abdomen pelvis, lymphangiography chest and abdomen, MR lymphangiography chest abdomen pelvis, or MR lymphangiography chest and abdomen is usually appropriate for pretreatment planning in an adult or child patient with traumatic or iatrogenic etiology for chylothorax treatment planning. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 2:** Lymphangiography chest abdomen pelvis, lymphangiography chest and abdomen, MR lymphangiography chest abdomen pelvis, or MR lymphangiography chest and abdomen is usually appropriate for pretreatment planning in an adult or child patient with nontraumatic or unknown etiology for chylothorax treatment planning. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at <https://acsearch.acr.org/list>. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at <https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria>.

Gender Equality and Inclusivity Clause

The ACR acknowledges the limitations in applying inclusive language when citing research studies that predates the use of the current understanding of language inclusive of diversity in sex, intersex, gender, and gender-diverse people. The data variables regarding sex and gender used in the cited literature will not be changed. However, this guideline will use the terminology and definitions as proposed by the National Institutes of Health.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an

		alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® [Radiation Dose Assessment Introduction](#) document.

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
0	0 mSv	0 mSv
● (radioactive symbol)	<0.1 mSv	<0.03 mSv
● (radioactive symbol) ● (radioactive symbol)	0.1-1 mSv	0.03-0.3 mSv
● (radioactive symbol) ● (radioactive symbol) ● (radioactive symbol)	1-10 mSv	0.3-3 mSv
● (radioactive symbol) ● (radioactive symbol) ● (radioactive symbol) ● (radioactive symbol)	10-30 mSv	3-10 mSv
● (radioactive symbol)	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."

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Disclaimer

The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new

equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

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