American College of Radiology
ACR Appropriateness Criteria®
Chylothorax Treatment Planning

Variant 1: **Chylothorax treatment planning: traumatic etiology.**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	8	This procedure is the initial examination to screen for pleural effusion or an alternative cause of dyspnea or chest pain.	☢
Lymphangiography chest and abdomen	8	If further evaluation and minimally invasive treatment is warranted, this procedure is the test of choice for traumatic chylothorax and can include diagnostic and therapeutic embolization.	☢☢☢
MRI chest and abdomen without IV contrast	6	This procedure is particularly helpful if lymphangiography does not delineate an abnormality.	O
MRI chest and abdomen without and with IV contrast	5	This procedure is particularly helpful if lymphangiography does not delineate an abnormality.	O
CT chest and abdomen without IV contrast	5		☢☢☢☢
CT chest and abdomen without and with IV contrast	5		☢☢☢☢
CT chest and abdomen with IV contrast	4		☢☢☢☢
Tc-99m lymphoscintigraphy chest and abdomen	4		☢☢
US chest and abdomen	4		O
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

Variant 2: **Chylothorax treatment planning: nontraumatic or unknown etiology.**

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	8	This procedure is the initial examination to screen for pleural effusion or an alternative cause of dyspnea or chest pain.	☢
Lymphangiography chest and abdomen	8	This procedure is appropriate if a concomitant minimally invasive attempt at therapy is desired.	☢ ☢ ☢
MRI chest and abdomen without IV contrast	7	This procedure is useful to visualize lymphatic vessels.	O
MRI chest and abdomen without and with IV contrast	7	This procedure is useful to visualize lymphatic vessels and exclude vascular abnormalities.	O
CT chest and abdomen with IV contrast	7	This procedure is helpful if venous thrombosis is suspected as the cause of the chylous effusion.	☢ ☢ ☢ ☢
CT chest and abdomen without IV contrast	5		☢ ☢ ☢ ☢
CT chest and abdomen without and with IV contrast	5		☢ ☢ ☢ ☢
Tc-99m lymphoscintigraphy chest and abdomen	3		☢ ☢
US chest and abdomen	3		O
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

CHYLOTHORAX TREATMENT PLANNING

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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. Chronic chyle leak results in metabolic abnormalities, respiratory compromise, immunosuppression, malnutrition, and even death [1-3]. A review of the etiology, diagnosis, and management of chylothorax is presented in addition to an evaluation of relevant imaging studies.

Etiology

Chylothoraces can be categorized etiologically as traumatic or nontraumatic. Collectively, the incidence of chylothorax is approximately 1 per 6000 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 [2,10,11].

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, in higher-output chylothoraces, and in underlying neoplastic etiologies. 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 increased risk of complications [12,13]. Although the technical success of direct surgical ligation is high, these debilitated patients

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are at 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 [14,15] defined the technique and reported its feasibility. The initial series of 42 patients by Cope and Kaiser [16] 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 report by Nadolski and Itkin [6] reported that TDE for nontraumatic chylous effusions in 34 patients was primarily successful if there was thoracic duct occlusion and extravasation. Pamarthi et al [7] reported 85% technical success and 64% clinical success in 105 patients with all-cause chylous leaks. Additional series have yielded similar results. Collectively, TDE has higher clinical success treating traumatic compared with nontraumatic chyle leaks and with TDE compared with thoracic duct disruption [8,9,11,17]. Overall, acute complications associated with TDE are minor and generally self-limited and are estimated at 2% to 6% [5-7]. Long-term complications may be seen in up to 14% of patients and may include leg swelling, abdominal swelling, or chronic diarrhea [18].

Overview of Imaging Modalities

Different imaging studies serve different purposes in the evaluation and treatment of chylothorax.

Chest radiography

Chest radiographs are routine examinations to evaluate dyspnea, particularly in postoperative scenarios and in patients who require intensive care. Radiographs can reliably detect pleural effusions or alternative diagnoses and monitor the position of support lines and tubes [19]. Although there is a high sensitivity for pleural effusions, this technique cannot reliably characterize the type of effusion.

Ultrasound

Ultrasound (US) is sensitive for the detection of pleural fluid but cannot definitively discriminate between the types of pleural effusion [20]. US is now commonly used to help guide thoracentesis. Similarly, US can be used to facilitate intranodal lymphangiography, which is becoming a more accepted technique. Beyond facilitating these procedures, the role of US is limited with regard to the evaluation and management of chylothorax [21].

Conventional lymphangiography

Lymphangiography has historically been used to opacify lymphatic vessels, detect lymph nodes and metastatic lesions, and evaluate lymphatic flow. Technological improvements in alternative diagnostic modalities led to an abandonment of this technique for oncologic purposes because it was technically challenging and time intensive, provided less information, and was more invasive. Although proficiency and training in the performance of lymphangiograms decreased, the utility of lymphangiography to demonstrate lymphatic leak became an established indication [8,9,14,22,23].

Traditionally, lymphangiography is performed from a pedal approach with the patient in a supine position. In this technique, a dye such as methylene blue that stains the lymphatics is injected into the web spaces between the toes. After the lymphatic vessel fills with the dye, the tissue overlying the lymphatic vessel is incised vertically, the lymphatic vessel is carefully skeletonized, and a 30-gauge lymphangiography needle is used to access the vessel. After securing the lymphatic access, 6 to 8 mL of ethiodized oil is instilled at a rate of 4 to 10 mL/h. Serial spot radiographs from the foot to the chest are acquired approximately every 10 to 15 minutes to follow the progression of the ethiodized oil as it ascends [22,24-27].

More recently, an interest in nodal lymphangiography has developed [21,28]. In this alternative approach, an inguinal lymph node is targeted with a 25- to 26-gauge spinal needle under US guidance. The needle is positioned between the hilum and cortex of a lymph node. Hand injection of ethiodized oil at a rate of 1 mL per 5 to 7 minutes is then initiated for a total volume of 6 to 10 mL. Serial spot radiographs of the pelvis, abdomen, and thorax are then acquired to follow the progression of ethiodized oil [21,25,28-30]. Intranodal lymphangiography appears to decrease procedure time, is less technically challenging, and decreases the risk of wound infection when compared to pedal lymphangiography [21,28].

Lymphangiography is able to define the site of the leak, diagnose alternative lymphatic vessel diseases, and prevent unnecessary procedures. Several authors have documented the therapeutic benefit of lymphangiography to occlude the site of leakage in 37% to 70% of patients without additional procedures [24-30]. Moreover, as detailed earlier, lymphangiography is intrinsic to the performance of TDE and guides the transabdominal access to the cisterna chyli and thoracic duct [14-16].

Nuclear lymphoscintigraphy
Nuclear lymphoscintigraphy images the pathways of lymphatic flow and lymph nodes and is most commonly used to identify draining lymph nodes proximal to neoplasms. A few reports of lymphoscintigraphy with Tc-99m–labeled radiotracers or orally administered iodine I-123-labeled 15-(4-iodophenyl)-3(R,S)-methylpentadecanoic acid are present and demonstrate the potential to visualize the anatomic configuration of the thoracic duct, reveal abnormal lymphatic drainage patterns, and potentially detect leaks [31-33]. However, aside from a few small series, little is present in the literature to support its routine use in the diagnosis or treatment of chylothorax.

Magnetic resonance imaging chest and abdomen
Visualization of the cisterna chyli, thoracic duct, and tributary lymphatic vessels with magnetic resonance imaging (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 magnetic resonance cholangiopancreatography. Further refinements of sequences, particularly heavily T2-weighted sequences with and without and fat suppression, combined with 3-D techniques, maximum-intensity projections, and higher magnetic fields, increased the reliability and quality of MR lymphangiography [35-39]. Morphological features of the cisterna chyli and thoracic duct can be noted with identification of these structures in over 90% of preoperative patients, potentially providing valuable information and decreasing their risk of lymphatic leak [40-43].

The vast majority of studies are performed with unenhanced techniques, although delayed-phase cisterna chyli enhancement has been noted [44]. Respiratory gating and further technical refinements have the potential to better elucidate minor lymphatic vessels and lymphatic vessels in antidependent areas, which may not be seen through conventional lymphangiography. Recent studies are beginning to document the feasibility of using gadolinium-based contrast material injection within groin lymph nodes 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, but these are preliminary research experiences and are not widely available [45,46].

Computed tomography chest and abdomen
Older studies noted that noncontrast computed tomography (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 [47]. At least some portion of the thoracic duct was visualized in 55% of patients in a different series [48].

Although MRI more reliably visualized more segments of the thoracic duct than CT, the addition of CT increased the number of visualized segments [36]. More recent 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 [49]. Older reports using a combination of lymphangiography and CT did not find any additional value of CT in diagnosing the lymphatic injury, although in a more recent series, a 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 [30]. Moreover, in this series of 24 patients, the lack of thoracic duct leakage was managed with nonoperative therapy with higher success rates [30]. No evidence is present to suggest a role for intravenous contrast material.

When the underlying etiology of chylothorax is unknown or nontraumatic, the speed, sensitivity, and specificity of CT imaging can narrow the broader differential diagnosis.

Discussion of the Imaging Modalities by Variant

Variant 1: Chylothorax treatment planning: traumatic etiology.

Traumatic chylothoraces are a result of direct injury to thoracic lymphatics. Iatrogenic traumatic chylothorax complicates up to 4% of esophageal resections [1,2,4-7]. 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. Sampling the pleural effusion confirms the diagnosis of chylothorax. Imaging a patient with a known traumatic chylothorax serves only to confirm the diagnosis and assist in therapeutic planning.

Chest radiography
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 [19].

Ultrasound
US can be helpful in the guidance of thoracentesis and intranodal injection during lymphangiography [21]. Otherwise, US has little, if any, diagnostic role in the setting of a known traumatic chylothorax.

Conventional lymphangiography
Conventional lymphangiography is the gold standard for visualization of lymph nodes, lymphatic vessels, cisterna chyli, the thoracic duct, and sites of injury [14,22,23]. Lymphangiography alone has been shown to be therapeutic in a small percentage of patients, irrespective of attempts at TDE or disruption [24-27]. 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].

Nuclear lymphoscintigraphy
Although nuclear lymphoscintigraphy may be able to confirm a lymphatic leak and identify the site, little evidence is present to support its routine use [31-33]. Moreover, this adds little to the clinical care of a patient as the traumatic etiology is usually known and any information gained would be redundant if conventional lymphangiography was performed as part of TDE.

Magnetic resonance imaging chest and abdomen
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 where identifying the cisterna chyli and/or thoracic duct is difficult or conventional lymphangiography is unsuccessful [40-43].

Computed tomography chest and abdomen
CT imaging is able to visualize portions of lymphatic system but provides less anatomic detail than MRI [36,47,48]. If the etiology is known, CT of the chest and abdomen, with or without intravenous contrast material, has little value in that it does not help guide therapy directed at chylothorax in most cases.

Variant 2: Chylothorax treatment planning: nontraumatic or unknown etiology.

Nontraumatic chylothorax accounts for approximately 46% of chylothoraces and can be subcategorized as resulting from malignancy, as 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 1 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 American College of Radiology (ACR), and the imaging evaluation includes chest radiography and chest CT [50,51]. The consistent finding of chylothorax on initial imaging is the presence of a pleural effusion, which is a common medical problem with more than 50 recognized causes [52]. Pleural fluid studies are necessary for definitive diagnosis and to narrow the cause etiology of chylothorax.

Chest radiography
Chest radiographs are routine examinations to evaluate dyspnea and have been designated as "usually appropriate" in the workup of ARI. Radiographs can reliably detect pleural effusions or alternative diagnoses and monitor the position of support lines and tubes [19,50,51]. This technique cannot reliably characterize the type of effusion.

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 [20]. US can be helpful in the guidance of thoracentesis and intranodal injection during lymphangiography [21].

Conventional lymphangiography
Conventional lymphangiography is the gold standard for visualization of lymph nodes, lymphatic vessels, cisterna chyli, and the thoracic duct and for detection of lymphatic leakage [14,22,23]. 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. Additionally, TDE is less clinically effective in a nontraumatic chylothorax unless thoracic duct occlusion or extravasation is present [6].

Nuclear lymphoscintigraphy
Nuclear lymphoscintigraphy has only a few reports that suggest it may be able to localize the site of chylous leak, particularly if used with 3-D single-photon emission CT/CT techniques [31-33]. Scintigraphic imaging alone provides limited localizing information and would not reliably narrow the differential diagnosis.

Magnetic resonance imaging chest and abdomen
MRI accurately visualizes lymphatic structures without intravenous contrast material, depicting abnormal lymphatic malformations. With the addition of contrast material, MRI can characterize mediastinal masses, pleural-based lesions, and chest wall pathology. However, thoracic MRI has limited utility for parenchymal lung pathology [35-39].

Computed tomography chest and abdomen
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 [36,50,51]. Moreover, it is a rapid examination that is easily tolerated by a supine patient. Intravenous contrast material accurately defines vascular and mediastinal structures and provides information on enhancement characteristics, which is a consideration when the etiology of chylothorax is unknown.

Summary of Recommendations

- In traumatic chylothorax, chest radiographs are useful to confirm the presence of pleural fluid, lateralize the process, and monitor the position of support lines and tubes. If further evaluation is warranted, lymphangiography can precisely define the leak and offer therapeutic benefit, particularly if paired with TDE. MRI and CT imaging are reserved for cases when lymphangiography is not diagnostic.
- Nontraumatic chylothorax can arise from a variety of disorders and may be a diagnostic dilemma. Chest radiographs are useful to confirm the presence of pleural fluid and lateralize the process. MRI and CT imaging are useful to narrow the differential diagnosis. Lymphangiography is helpful if a minimally invasive approach to treatment is desired.

Summary of Evidence

Of the 52 references cited in the *ACR Appropriateness Criteria® Chylothorax Treatment Planning* document, 22 are categorized as therapeutic references, including 6 good-quality studies and 8 quality studies that may have design limitations. Additionally, 30 references are categorized as diagnostic references, including 3 good-quality studies and 5 quality studies that may have design limitations. There are 30 references that may not be useful as primary evidence.

The 52 references cited in the *ACR Appropriateness Criteria® Chylothorax Treatment Planning* document were published from 1989 through 2015.

Although there are references that report on studies with design limitations, 9 good-quality studies provide good evidence.

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, both because of 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 to 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**
O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	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 (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies".		

Supporting Documents

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

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