Review Article



# Review of chylopericardium

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**Abstract:** Chylopericardium is a rare pathologic condition consisting of the accumulation of excess amounts of chylous fluid within the pericardial cavity. Most patients are asymptomatic at presentation; however, chest pressure, chest pain and lightheadedness have been reported, and the most common presenting symptom is shortness of breath. Patients are noted to have enlargement of the cardiac silhouette on routine chest radiograph, and evidence of a pericardial effusion on echocardiography. The diagnosis is only definitively confirmed with pericardiocentesis and fluid analysis. The fluid is typically turbid white or milky in appearance, with a triglyceride level in excess of 500 mg/dL. The mechanism by which chyle accumulates within the pericardium is believed to be secondary to abnormal or damaged lymphatics or due to elevated pressure within the thoracic duct that results in chyle reflux into the pericardium. Following drainage with a pericardiocentesis or pericardial drain, attempts at conservative therapy with nothing by mouth and parental nutrition can be made, but have a high rate of failure and subsequent reaccumulation of chyle. Surgical treatment provides the most definitive management and consists of ligation of the thoracic duct just above the level of the diaphragm and creation of a pericardial window. With this treatment, risk of recurrence is incredibly low (<5%).

**Keywords:** Chylopericardium; thoracic duct ligation; pericardial window

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## Introduction

Chylopericardium is a rare pathologic entity consisting of the accumulation of chylous fluid within the pericardial cavity (1-3). Isolated or primary chylopericardium was first described in detail by Dr. Groves in the *New England Journal of Medicine* in 1954, and includes patients with no antecedent trauma, cardiac or thoracic surgery, or mediastinal tumor (4). The pericardial cavity of healthy adults contains a small amount of serous fluid, similar in content to lymphatic fluid, that decreases friction between the myocardium and the surrounding fibrous pericardium (1). When fluid collects within the cavity, in excess of this normal amount, it causes gradual distention of the pericardial sac. If the fluid accumulates too rapidly, the pericardium cannot distend to accommodate and the fluid begins to exert pressure on the heart, which can impede filling of the heart during diastole.

## Symptoms

Chylopericardium can occur in all age groups, with reported adult cases ranging from age 18 to 70, and affects both sexes equally (5). Most of the cases in the literature describe patients that are asymptomatic at presentation (40–60%) (1,5), and the chylopericardium was diagnosed as an incidental finding, usually on a routine chest X-ray. Of the patients that are symptomatic, shortness of breath and dyspnea is the most common presenting symptom (39–64%) (1,2). Additional common symptoms include cough, chest pressure, chest pain, lightheadedness, and syncope. When fluid accumulates more rapidly, and enough pressure is generated, impairment of venous return to the heart results in cardiac tamponade physiology, which is the clinical

presentation in approximately 17–30% of patients (1,2).

## Diagnosis

The time between onset of symptoms and diagnosis displays a wide variation, with minimally or asymptomatic patients taking years to be finally diagnosed, and those with signs and symptoms of cardiac tamponade, being diagnosed immediately (2). Physical exam may demonstrate distant heart sounds, but in the absence of tamponade physiology, there are unlikely to be any other clinical findings. Those who present in tamponade will have elevated jugular venous pressures, Kussmaul's sign, and evidence of pulsus paradoxus (5). Patients are commonly noted to have enlargement of the cardiac silhouette on routine chest radiograph, which is often the first diagnostic study to be performed (1). This is characteristically followed by an echocardiogram, utilized to confirm the presence of a pericardial effusion and to assess for impairment of venous filling during diastole.

While these studies are useful to diagnose a pericardial effusion, they do no differentiate between a chylous effusion and other sources of benign and malignant effusions. The diagnosis is only confirmed with pericardiocentesis and fluid analysis (6). The fluid in a chylopericardium is typically turbid white or milky in appearance, has a high triglyceride level, a predominance of lymphocytes, and the presence of fat globules as confirmed by Sudan III staining (1). The fluid demonstrates a triglyceride level in excess of 500 mg/dL in the vast majority of cases (2). Cultures of the fluid are almost universally negative as chyle itself is bacteriostatic (7). Patients then are often assessed with a chest computed tomography (CT) scan. The CT scan functions to assess for possible secondary causes of impairment of lymphatic drainage, including assessment for mediastinal malignancies.

The source of the chyle within the pericardial cavity was first demonstrated by the oral administration of Sudan III dye mixed with corn oil and milk and subsequent demonstration of the pink dye within the pericardial effusion, thereby documenting that the chyle within the pericardium was related to the thoracic duct (4). Lymphoscintigraphy and lymphangiography have additionally been utilized to document the source of the chyle in chylopericardium cases. Lymphoscintigraphy involves the subcutaneous injection of Technetium-99 sulfur colloid (Tc99-SC), usually in the medial interdigital web spaces of both feet, after which whole body images are obtained immediately, at 3 hours, and 5 hours after injection (3,5). The radiotracer is absorbed via the lymphatics in the lower extremities and transported to the cisterna chyli and ultimately into the thoracic duct; providing visualization of the duct and potentially identifying leaks or obstructions along its course.

A Tc99-SC injection performed in a patient with chylopericardium and a control patient demonstrated an abnormal accumulation of tracer within the mediastinum and hilar lymph nodes of the chylopericardium patient (3). Additionally, it demonstrates a delay in filling of the upper mediastinum and left subclavian vein compared to control patients, giving evidence to lymphatic obstruction in the upper mediastinum or central venous hypertension as being a primary cause of chylopericardium. Radiographic lymphangiography involves the injection of a radiopaque contrast agents into the lymphatic vessels, and fluoroscopic images to trace the contrast as it moves through the lymphatic system. Lymphangiography can also be utilized in combination with Computed Tomography to demonstrate evidence of fistulous connection between the thoracic duct and the pericardial sac, usually at the level of the bifurcation of the trachea (8). Lymphangiography with CT has become the gold standard for detecting the location and the source of the fistula or obstruction, as this provides both information about the course and flow of the thoracic duct, but also cross section anatomical information about the pericardium and upper mediastinum (3,8).

## Causes

The etiology of chylopericardium includes congenital lymphangiectasia, iatrogenic injury after cardiac or thoracic surgery, malignant tumors (thymic tumors and lymphoma especially), blunt or penetrating trauma, infection, radiation, pulmonary hypertension, subclavian vein thrombosis, and primary or idiopathic causes as well as other causes of elevated central venous pressure (2,6). The most common cause over the past few decades is idiopathic 56%, followed by post-operative 9%, and mediastinal neoplasms 6% (2). Lymph drains from the pericardium towards the anterior mediastinum, then the bronchopulmonary lymph nodes, and finally into the thoracic duct (1). The pathophysiologic mechanism by which chyle accumulates within the pericardium is believed to be secondary to either damaged lymphatics that result in an abnormal communication between the thoracic duct and the lymphatic vessels of the pericardium (9), or elevated pressure within the thoracic

duct or venous system that results in chyle reflux into the pericardial lymphatics (8). This elevated pressure can be secondary to obstruction distally within the thoracic duct itself, but also can be secondary to mechanical obstruction of the venous drainage of the duct, namely the left subclavian vein (1). Animal studies, however; have demonstrated that chylopericardium does not necessarily occur with distal obstruction of the thoracic duct, as there are multiple sources of collateral drainage, including the azygous, intercostal and lumbar venous systems (7,9), and therefore some mechanism by which normal collateral drainage does not occur is also necessary for the development of a chylous pericardial effusion (6). Also, generalized chronic elevation of central venous pressure (such as congenital heart disease) can result in chylopericardium.

Chyle can be produced in excess of 2,000 mL per day, and the thoracic duct is capable of generating a normal pressure of 30 cm of water, and this can increase to up to 65 cm of water when the thoracic duct is ligated or obstructed (7). Likely due to the high pressures that can be generated, fistulae of the thoracic duct will only close spontaneous in approximately 50% of cases, even when there is no identifiable distal obstruction. This is in stark contrast to most other types of hollow viscous fistulae that are much more likely to heal spontaneously (7).

## Treatment

Conservative management is attempted initially in most patients diagnosed with idiopathic chylopericardium. This includes initial evacuation of chyle from the pericardial space, consisting of intermittent pericardiocentesis or continuous drainage with a pericardial catheter. This is followed by NPO dietary status with the exception of medium chain triglycerides, and electrolyte and nutritional repletion with total parental nutrition (TPN) (2). NPO status reduces the amount of fatty acids absorbed from the gastrointestinal tract and ultimately transported into the thoracic duct as chyle (10), however, medium chain fatty acids are absorbed directly into the blood stream via the portal vein, and can therefore safely be administered without increasing chyle production.

Surgical intervention is recommended as the initial treatment in all patients with a hemodynamically significant chylous effusion on presentation. Conservative therapy fails in approximately 50–60% of patients, and therefore surgical intervention is indicated (1,2). Failure of conservative therapy is considered ongoing drainage in excess of 500 mL of fluid per day, over a 7-day period of time, or concern over complications related to nutritional deficiency (2). Additionally, any symptomatic recurrence of a chylous pericardial effusion should prompt consideration of surgical intervention, as these are less likely to be successfully managed with conservative interventions.

The most common surgical management includes a right video-assisted thoracoscopic surgery (VATS) or thoracotomy and ligation of the thoracic duct, partial pericardiectomy, and creation of a pericardiopleural fistula (a pericardial window). Thoracic duct ligation for chylothorax was first demonstrated to be safe and effective by Lampson (11), carries a very low risk of mortality (10), and has no known long term nutritional or immunologic sequela (7). If the chylopericardium is associated with a chylous pleural effusion, then the approach should be ipsilateral to the side of the effusion.

Given the anatomic variations of the thoracic duct, the duct should be ligated as low as possible within the chest, just above the level of the diaphragm (1). The lymphatic drainage is more commonly a single duct from the cisterna chyli in the abdomen up to the level of the eighth thoracic vertebra. Above this level there is an increased possibility of duplication or multiplication of the mediastinal portion of the duct, which occurs in up to 40% of patients according to cadaveric studies (12).

The surgical procedure includes division of the right inferior pulmonary ligament to allow exposure of the posterior mediastinum, at the level of the diaphragm (10). Mass ligation of the duct, and all of the soft tissue between the esophagus anteriorly, the azygous vein laterally, and the aorta medially is achieved by mobilization of the esophagus (8). Mass ligation is recommended as opposed to attempting identification of the duct and direct clipping, as the attempt to identify the duct is more likely to lead to iatrogenic injury and resultant right sided chylous effusion (6).

Attempts at treatment by creation of a pericardial widow, without duct ligation, are associated with high rates of recurrence, as described in the first report of primary chylopericardium by Groves (4). Whereas recurrence after the combination of duct ligation and pericardial window creation is incredibly rare, less than 5% (2,13). Pericardial window creation, and adequate drainage of the pericardial space is paramount to prevent complications such as constrictive pericarditis (5), and therefore a pericardial window anterior to the right phrenic nerve is most appropriate (10). Due to the possibility of malignant

etiologies, the fluid and pericardial tissue should be collected for pathologic analysis, however the histologic evaluation of the pericardium commonly demonstrates only acute pericarditis, with no other histologic abnormalities. The use of any sclerosing agents to minimize risk of recurrence, such as tetracycline or bleomycin, within the pericardial sac is contraindicated due to a high prevalence of post intervention constructive pericarditis (1). Finally, treatment of any underlying cause of lymphatic obstruction is recommended for all causes of secondary chylopericardium (i.e., treatment of a malignancy).

## Conclusions

Chylopericardium represents an unusual and rare clinical entity. The accumulation of chyle can be secondary to recent cardiac or thoracic surgery, trauma, mediastinal malignancy, or central venous hypertension, but the vast majority of cases are idiopathic with no identifiable cause of mechanical obstruction of the thoracic duct. The definitive diagnosis is made with pericardiocentesis and drainage of milky white fluid with a significantly elevated triglyceride count. Further clinical assessment should be made with CT and lymphangiography to evaluate for potential secondary causes. Surgical treatment provides the most definitive management and consists of mass ligation of the thoracic duct just above the level of the diaphragm and creation of a pericardial window. Following this treatment, the risk of recurrent chylopericardium is exceedingly low, less than 5%.

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