Chylous Reflux Syndrome Involving the Pericardium and Lung*

Robert J. Toltzis, M.D.; Amnon Rosenthal, M.D.;** Kenneth Fellows, M.D.; Aldo R. Castaneda, M.D., F.C.C.P.; and Alexander S. Nadas, M.D.

A ten-year-old boy with symptom-free pulmonary interstitial edema and pericardial effusion was found to have a chylous reflux syndrome involving the pericardium and lung. Evidence suggests that unlike patients with isolated primary chylopericardium in whom pericardial window and/or ligation of the thoracic duct is curative, those with chylous reflux should be managed medically since the morbidity and mortality of surgical treatment is high in this syndrome.

Chylous reflux syndrome is an entity describing a primary disturbance in which there is a backflow of chyle from its normal pathway from the bowel lacteals to the cysterna chyli, through the thoracic duct and into the venous system. Chyle is discharged into various organs or may collect in serous cavities, with clinical features of the syndrome varying according to the major site of discharge or accumulation of chyle. Primary chylous pericardium, associated with pulmonary lymphedema is rare.²⁻⁵

The purpose of this report is to describe a patient with a chylous reflux syndrome involving the lung and pericardium.

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**From the Departments of Cardiology, Cardiovascular Surgery and Radiology, The Children's Hospital Medical Center and the Department of Pediatrics, Harvard Medical School, Boston.
**Presently at Department of Pediatrics, C.S. Mott Children's Hospital, Ann Arbor.
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Reprint requests: Dr. Nadas, Department of Cardiology, Children's Hospital Medical Center, Boston 02115

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Figure 1. Normal chest x-ray film obtained March 3, 1972 (A). Note enlarged cardiac silhouette and severe interstitial pulmonary edema with Kerley B lines and fluid in fissures Nov 9, 1976.

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CASE REPORT

The patient, born Aug 29, 1966 was referred from Mexico City with a diagnosis of probable constrictive pericarditis. A routine chest x-ray film was normal (Fig 1A) in 1972. Due to intermittent cough a chest x-ray film was made in 1974, revealing pulmonary interstitial edema.

Physical examination on Nov. 8, 1976, disclosed no paradoxical pulse, jugular venous distension or hepatosplenomegaly; the chest was clear and the heart examination yielded normal findings. Chest x-ray film showed mild cardiomegaly and severe interstitial pulmonary edema (Fig 1B). The electrocardiogram disclosed mildly diminished QRS voltages. Pulmonary function studies revealed minimal airways obstructive disease. Echocardiogram was normal except for a small posterior pericardial effusion. Serum lipoprotein electrophoresis was normal. Gastrointestinal loss of plasma albumin was 0.7 percent over a four-day-period (normal < 1 percent). Cardiac catheterization and cineangiography, at the high altitude of Mexico City, suggested constrictive pericarditis or early tamponade. Catheterization repeated at sea level one month later showed no abnormalities (Table 1).

On Nov 15, 1976 exploratory left thoracotomy was performed, revealing multiple dilated lymphatic channels over the parietal pleura and pericardium; the epicardium was not involved. Over 200 ml of chylous pericardial fluid was removed. A large pericardial window was created and lung biopsy obtained. Analysis of the pericardial fluid showed a triglyceride level of 2,825 mg/dl and cholesterol level of 250 mg/dl. Microscopic examination of the lung disclosed multiple dilated endothelial-lined valved channels filled with proteinaceous material in the subpleura and interstitium. The general architecture of the lung parenchyma was intact. The pericardium was thickened and contained a fibrous exudate with dilation of parietal and pleural endothelial lined channels.

Lymphangiography, performed Nov 23, 1976, using 5 ml of iodized oil (Lipiodol), revealed normal lymphatic drainage up to the cysterna chyli, with contrast material then ascending through multiple, diffuse, dilated and tortuous lymphatic...
channels to the thorax and emptying into the left subclavian vein. A normal unobstructed proximal thoracic duct was visible, but with multiple lymphatic fistulae constituting most of its length in the chest. Within 24 hours, contrast material was present in the lung, pleural fluid and pericardium (Fig 2).

Postoperatively, the patient developed a recurrent left chylothorax, transient scrotal edema, mild ascites and non-pitting edema of the left side of the chest and abdomen, as well as leakage of lymph into the urine. Despite a dietary regimen containing low fats and large amounts of medium-chain triglycerides, he had had recurrent left chylothorax.

**DISCUSSION**

In patients with isolated primary chylopericardium, creation of a pericardial window and/or ligation of the thoracic duct or large lymph channels in the lower thorax is usually curative, and no reaccumulation of chylous pericardial fluid occurs. Patients with chylopericardium associated with lymphangiectasia of the lungs or mediastinum do not fare as well, with recurrent chylopericardium and high morbidity and mortality described. The multisystem involvement, clinical course and adverse response to therapy suggest a different structural basis and pathogenesis for the chylopericardium in patients with associated lymphangiectasia of the lungs.

In our patient, the chylopericardium and interstitial pulmonary fluid were stimulated by diffuse fistulous and ectatic channels along the paravertebral thoracic lymph channel. In the absence of thoracic duct obstruction or history of trauma, the lymphangiectasia is probably congenital in origin. The contrasting size of the cardiac silhouette and hemodynamic findings during catheterization in Mexico City and in Boston may be attributable to the effects of altitude. Pulmonary arterial hypertension due to high altitude hypoxia could have resulted in elevation of right ventricular end diastolic pressure, and, subsequently, right atrial and systemic venous pressure. The pressure in the thoracic duct and its tributaries would be increased, resulting in increased reflux of lymph through the various fistulae and ectatic channels into the interstitial spaces of the lung and pericardium. This, in turn, would increase filling pressure in the right atrium and right ventricle, thereby setting up a self-perpetuating cycle while the patient resides at a high altitude.

In the present case, surgery appears to have complicated the situation by interrupting many of the lymphatic channels and fistulae. Thus we learned that surgical exploration, lung biopsy and pericardial window formation not only are unnecessary to establish the diagnosis but may be hazardous and cause undue morbidity. Pericardiocentesis will establish the diagnosis, while a lymphangiogram will help elucidate the pathophysiology. Initial management should be medical with dietary restriction of conventional long-chain fats and substitution of medium-chain triglycerides because the latter are principally absorbed via the portal vein and then absorption is not dependent on formation of chylomicrons and transport via the lymphatic system. Only after medical management has failed or if tamponade is present should creation of a pericardial window be considered.

**REFERENCES**


![Figure 2. Twelve hours after injection in lymphatic channel in foot, contrast material appears in pericardium and lung.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21013/ on 06/26/2017)