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BEGINNER

CASE REPORT: CLINICAL CASE

# Chylothorax Due to Superior Vena Cava Obstruction in a Patient With Complex Congenital Heart Disease

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## ABSTRACT

Obstruction of the superior vena cava represents an under-recognized cause of chylothorax in the adult population. Our case report describes the successful conservative management of chylothorax due to bilateral superior vena cava obstruction in an adult patient with complex congenital heart disease. (**Level of Difficulty: Beginner.**)
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## HISTORY OF PRESENTATION

We described a 37-year-old woman with complex congenital heart disease (CHD), including dextrocardia, right atrial isomerism, double outlet right ventricle with transposition of the great arteries, and severe pulmonary stenosis, presenting with pleural effusion. The echocardiographic examination showed severely impaired systemic right ventricular contractility with severe atrio-ventricular valve regurgitation. The computed tomography (CT) scan revealed bilateral pleural effusion, prevalent on the left with appearance of sac-like effusion and scissural involvement. Extensive, multisegmental disventilatory phenomena were also noted. The patient underwent thoracentesis and analysis of the pleural fluid. The latter showed a milky, transudative fluid (low protein and lactate dehydrogenase level less than two-thirds of the upper limit of the normal serum value), with high triglyceride levels (>110 mg/dl) and low cholesterol levels (<200 mg/dl), suggesting chylothorax.

### LEARNING OBJECTIVES

- To review the diagnostic tools for chylothorax.
- To describe a conservative management strategy of nontraumatic chylothorax in a high-risk patient with complex CHD.

## MEDICAL HISTORY

The patient underwent Blalock-Taussig shunt at the age of 1 year and bilateral Glenn anastomosis of the right and left superior vena cava (SVC) with the pulmonary artery, associated with Blalock-Taussig shunt closure, at the age of 22 years. In 2018, the patient was referred to our division for clinical evaluation. The

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echocardiographic examination showed severely impaired systolic function of the systemic right ventricle, therefore, the patient underwent subcutaneous implantable cardioverted defibrillator implantation for primary prevention.

## DIFFERENTIAL DIAGNOSIS

A differential diagnosis with other possible causes of pleural effusion (i.e., empyema and heart failure) was performed using pleural fluid analysis.

## INVESTIGATIONS

A few days after hospital admission, the patient underwent angiographic CT scan that revealed the presence of metallic clips at the inferior anastomosis of left SVC with the main pulmonary artery, and a dilated azygos vein, both presenting several thrombi. Thrombotic occlusions of the right SVC, right subclavian vein, and ipsilateral jugular vein with multiple collateral vessels were also detected (**Figure 1**).

## MANAGEMENT

Once the diagnosis of chylothorax was made, a chest drain was inserted to ensure complete lung expansion; a medium-chain triglyceride (MCT) diet and therapy with octreotide were started as well. Although bilateral obstruction of the SVCs was present, our patient was considered to be at high risk for endovascular intervention, therefore anticoagulation therapy with intravenous heparin was commenced.

## DISCUSSION

Chylothorax is the accumulation of lymphatic fluid in the pleural cavity (1). Common causes include direct injury of the thoracic duct after surgery or the infiltration of the lymphatic systemic secondary to malignant disease, whereas SVC obstruction represents a rare cause (2–4). Chylothorax is a frequent complication of CHD surgery, occurring in about 1% to 9% (5), increasing morbidity and mortality. Univentricular palliation procedures contribute to raise the central venous pressure, increasing the production of lymph by the liver; some patients may have a failure of the lymphatic circulation in draining this excess of lymph back into the vein circulation. These patients may have chylothorax early after Fontan operation or from protein-losing enteropathy and plastic bronchitis at a later stage. It has been suggested that the rate of chylothorax is higher after operations for single-ventricle palliation, especially

## ABBREVIATIONS AND ACRONYMS

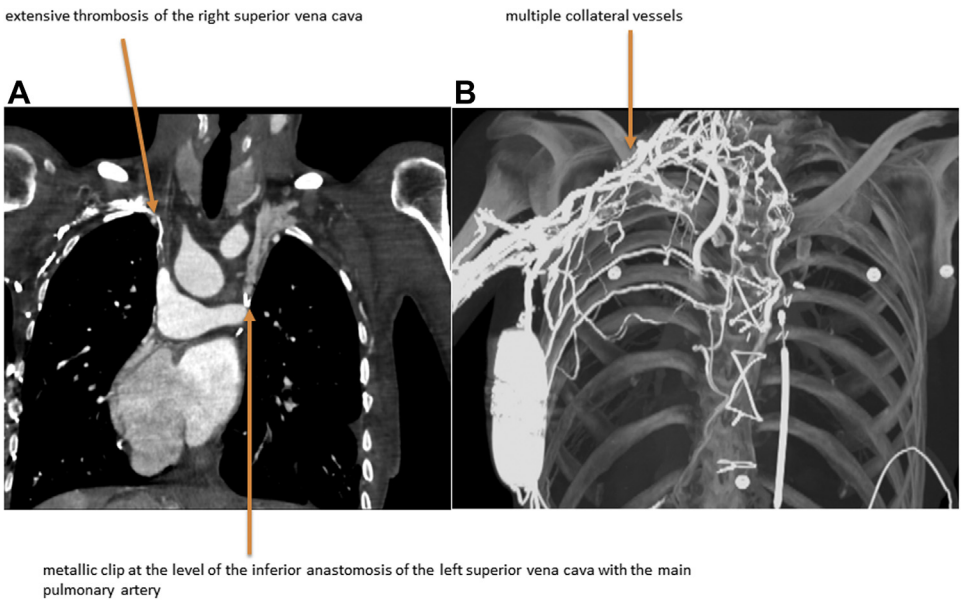
**CHD** = congenital heart disease

**CT** = computed tomography

**MCT** = medium-chain triglyceride

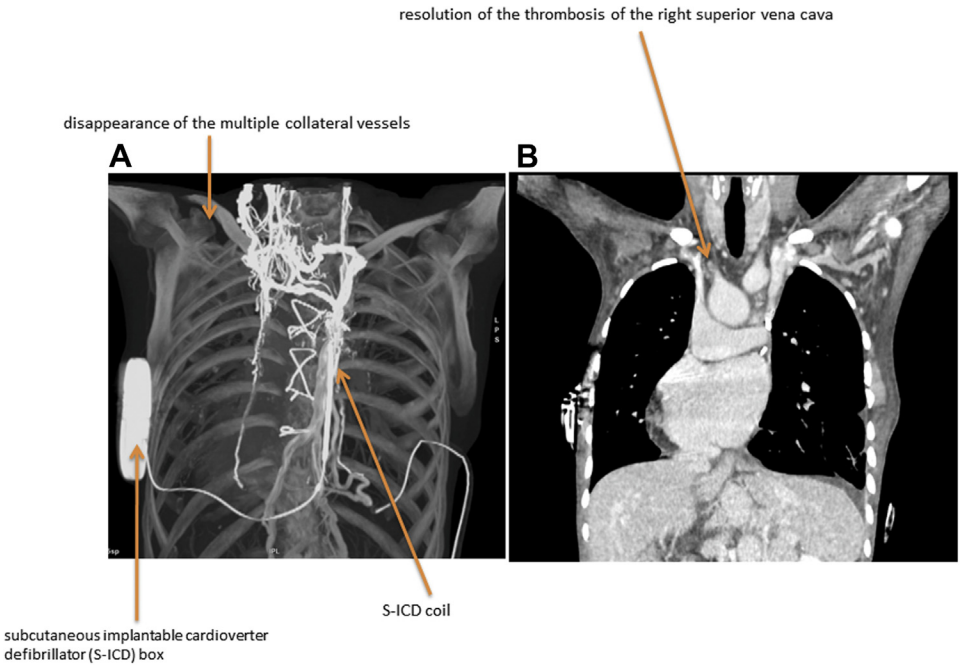
**SVC** = superior vena cava

**FIGURE 1 Thrombosis of the Right Superior Vena Cava With Collateral Vessels**




Coronal maximum intensity projection **(A)** and coronal plane **(B)** images showing extensive thrombosis of the right superior vena cava **(A)** with multiple collateral vessels **(B)**; a metallic clip is also visible at the level of the inferior anastomosis of the left superior vena cava with the main pulmonary artery **(A)**.

**FIGURE 2 Disappearance of the Collateral Vessels and Resolution of Thrombosis**




Disappearance of the collateral vessels in the scapular region **(A)**; subcutaneous implantable cardioverter defibrillator box and its coil **(A)**; resolution of thrombosis in the right superior vena cava **(B)**.

in patients with systemic right ventricle. Although postoperative chylothorax may be caused by direct injuries to the thoracic duct, it is now believed that increased central venous pressure and central vein thrombosis represent the most common causes after CHD surgery (6). Treatment of the underlying causes is key to resolution of chylothorax. Thoracentesis represents an initial intervention for both diagnostic and therapeutic purpose. SVC thrombosis is primarily treated with endovascular intervention (7), however, anticoagulation alone in case of central vein thrombosis has been reported with successful resolution of chylothorax (8). MCT diet and octreotide may be prescribed to reduce the chyle flow in the thoracic duct. The use of MCT diet leads to diminished lymph flow and intralymphatic pressure; thus, MCT are directly absorbed into the portal system, not requiring chylomicron packaging or transport via the lymphatic system.This reduces the flow into the thoracic duct allowing the opportunity to heal (9). Somatostatin and its synthetic analogue octreotide promote the closure of thoracic duct leaks, decreasing the lymph flow. This is, at least in part, secondary to decreased absorption of triglycerides induced by the drug. The most frequently administered dose in the adult population is 50 mg every 8 h. The optimum duration of treatment is unknown, but most often it has been administered for 1 or 2 weeks (10).

## FOLLOW-UP

The angiographic CT scan performed after 3 weeks of conservative therapy with intravenous heparin, MCT diet, and octeotride showed resolution of the thrombosis in the right SVC and disappearance of the collateral vessels in the scapular region (**Figure 2**). The chest drain was removed with no new onset of pleural effusion and the patient was discharged soon after with stable clinical conditions.

## CONCLUSIONS

This case demonstrated that obstruction of the Glenn anastomosis may be an uncommon cause of chylothorax in patients with CHD. Treatment is

challenging, however, conservative management may be successful even in high-risk patients.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**KEY WORDS** chylothorax, congenital heart disease, superior vena cava obstruction