Case Report

Intravascular Stent Implantation for Refractory Chylothorax Secondary to Congenital Superior Vena Cava Stenosis in an Infant

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Abstract

Chylothorax is a rare but dangerous cause of respiratory failure in the pediatric population. It most commonly presents after cardiac surgery or, alternatively, due to retrograde pressure on the thoracic duct from narrowing or obstruction in the innominate vein due to thrombus or neoplasm. We observed an unusual presentation of chylothorax in an otherwise healthy 3-month-old infant with congenital superior vena cava stenosis leading to acute respiratory collapse. After initially not responding to medical therapy, the patient was successfully treated with an intravascular stent.

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Introduction

Respiratory failure secondary to chylothorax is a rare complication in patients with congenital heart disease [1]. In these patients, leakage of chyle into the thoracic space most commonly results from direct incidental damage to the lymphatic system during surgery or vascular obstruction in the innominate vein or superior vena cava (SVC) secondary to central venous line-associated thrombosis, postoperative vascular stenosis, or extravascular mass effect, all of which impede lymphatic decompression into the veins through the lymphatic duct. Very rarely, chylothorax is reported in infants with congenital stenosis of the SVC. In previous reports of chylothorax secondary to congenital SVC stenosis, patients have tended to present in the neonatal period and have almost always had additional complex congenital heart lesions that increased the index of suspicion for vascular anomalies as the underlying etiology [2, 3, 4]. Here, we present a unique case of an isolated congenital SVC stenosis leading to chylothorax and acute respiratory failure in an otherwise healthy 3-month-old. We describe the successful transcatheter stenting of the SVC and subsequent rapid resolution of the patient's effusion and symptoms.

Case Presentation

An ex-full-term and otherwise healthy 3-month-old boy was admitted to the hospital for acute respiratory failure in the context of a massive effusion surrounding the right lung visualized on chest x-ray. He was immediately intubated and the fluid drained with a tube thoracostomy. Fluid analysis showed high

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levels of triglycerides and cholesterol, consistent with a chylothorax. He was extubated shortly after thoracostomy tube placement and was stable on room air, although a significant amount of chylous fluid continued to drain from his thoracic cavity. A magnetic resonance lymphangiogram was obtained to evaluate for congenital lymphatic malformation or signs of genetic lymphedema-lymphangiectasia syndrome. However, imaging showed a normal thoracic duct with no obvious lymphatic malformation or intrathoracic mass. An upper extremity vascular Doppler ultrasound was negative for deep venous thrombosis in the right and left internal jugular, subclavian, and axillary veins. An echocardiogram had reportedly been notable for a patent foramen ovale and otherwise structurally normal heart at the referring hospital and was repeated upon admission. Transthoracic echo showed a narrowed right-sided SVC with low velocity but forward flow to the right atrium. There was retrograde flow in the unobstructed innominate vein and multiple large posterior venous structures that appeared to be decompressing the upper body central veins caudally to a dilated inferior vena cava. The echocardiogram was negative for additional intracardiac or vascular lesions. The patient showed

no signs of plethora or upper body edema on exam. After the patient was stabilized, medical management was attempted, including octreotide infusion and low fat formula feeding followed by central line placement in the right subclavian vein and initiation of total parenteral nutrition (TPN) when the clinical response was deemed inadequate. Chylous drainage from the chest tube continued to be significant despite these measures. Given the echocardiographic finding of SVC obstruction as the likely etiology of continued chylous effusion, the patient was referred to the cardiac catheterization lab for angiography and intervention 14 days after admission.

In the catheterization lab, the left femoral vein was accessed, and a hydrophilic catheter was placed in the stenotic SVC for hemodynamics and angiography. Right atrial pressure was normal, with a 5 mmHg gradient across the SVC. Angiography showed a severe long-segment SVC stenosis measuring 18 mm in length and 2.6 mm at its narrowest diameter (Figure 1). The upper body central veins were decompressing through a very dilated hemiazygous system. The SVC was nearly occluded by the right subclavian vein central line that had been placed 3 days prior to catheterization. Balloon
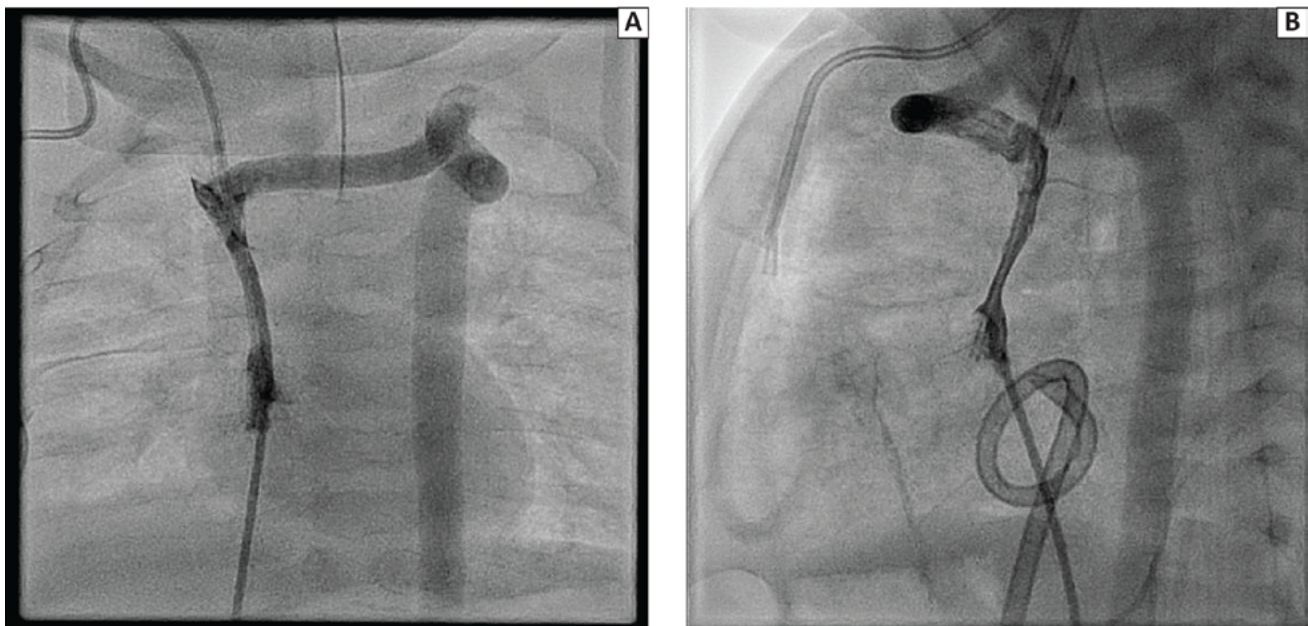

Figure 1. *Panel A*. Severe long-segment superior vena cava (SVC) stenosis and collateral decompression of upper body venous flow through a very dilated hemiazygos system. Note that the SVC stenosis was appreciated prior to insertion of the right subclavian central venous line. *Panel B*. Lateral view, with thoracostomy tube in view.

angioplasties were performed in the narrowed SVC using a 5 × 20 mm Sterling balloon (Boston Scientific, Marlborough, Massachusetts) over a .018 Platinum Plus wire (Boston Scientific, Marlborough, Massachusetts), but a tight waist persisted at 14 ATM of pressure (Figure 2). Dilations were then performed with a higher pressure 5 × 20 mm Dorado balloon (Bard Medical, Covington, Georgia) over the same .018 wire, which resulted in resolution of the tight waist at 24 ATM. However, on repeat angiography, although the SVC showed improved flow, there was significant vascular recoil and residual obstruction necessitating stent implantation. In anticipation of stent implantation, the central venous line from the right subclavian vein that crossed the SVC was pulled out of the SVC and into the left innominate vein using a snare catheter introduced from the hemiazygos vein. An 8F long sheath was placed in the SVC from the femoral vein over a .035 Amplatz Super Stiff wire (Boston Scientific, Marlborough, Massachusetts), and a Palmaz Genesis 1910 XD stent (Cordis, Fremont, California) mounted on a 6 × 20 mm Dorado balloon was deployed in the SVC and further dilated with a 7 × 20 mm Dorado balloon at 22 ATM. Afterward, there was a 1 mmHg gradient across the
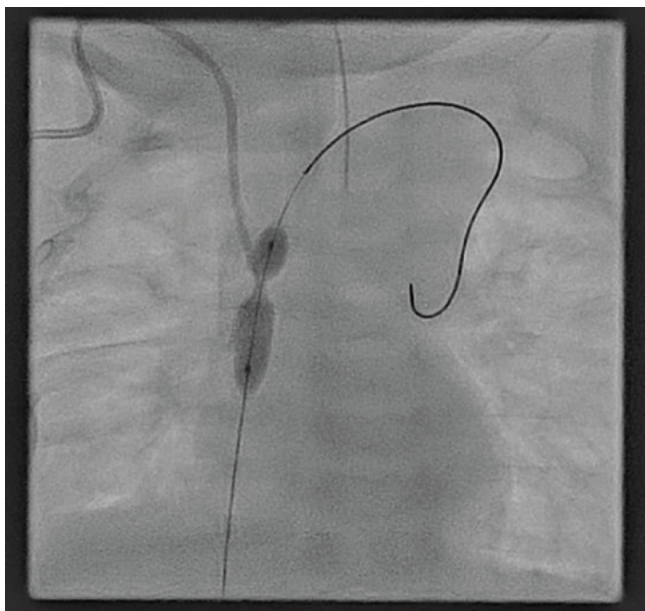

Figure 2. Balloon dilation shows a significant residual waist using a 5-mm Sterling balloon at 14 ATM of pressure. The stenosis ultimately required 22-24 ATM of pressure to resolve at full inflation.

stent, and the SVC was widely patent, measuring 6 mm. At the conclusion of the procedure, there was excellent flow through the well-positioned stent, and the veins no longer decompressed through the hemiazygos system; rather, all venous flow entered the heart briskly through the SVC (Figure 3). Of note, the patient required balloon venoplasty of the right iliofemoral vein due to narrowing, likely from a previous line placement. After venoplasty, a PICC line was placed in the right femoral vein so that the tunneled subclavian line could be removed.

The patient was transitioned off octreotide over the following 4 days. Medium-chain triglyceride formula was replaced with breast milk, and no recurrence of chylous pleural effusion was observed. The chest tube was removed on post-catheterization day 6.

To preserve and rehabilitate the obstructed right femoral vein, repeat catheterization was performed 11 days after initial catheterization, at which time the right femoral vein PICC was removed and angioplasty was performed to maintain the patency of the right iliofemoral vein. Repeat angiography of the SVC stent showed continued unobstructed flow across the SVC stent with no short-term recurrent narrowing (Figure 4). The patient was discharged 2 days later on low-dose aspirin for stent endothelialization.

At the most recent follow-up 5 months after discharge, echocardiogram showed excellent flow through a stably positioned SVC stent with a mean Doppler gradient of 3.8 mmHg. The dilated hemiazygous vein was no longer visualized. No recurrent pleural effusions were noted on follow-up chest radiography. The patient will require periodic repeat catheterizations with angioplasty of the SVC stent for somatic growth until the vessel reaches adult size.

Discussion

Chylothorax is a known cause of respiratory compromise and appears at an increased frequency in patients with congenital heart disease, in particular following intrathoracic surgical intervention. Chylothorax is most commonly due to incidental damage to the thoracic duct during a surgical procedure or, alternatively, to venous stenosis/obstruction causing
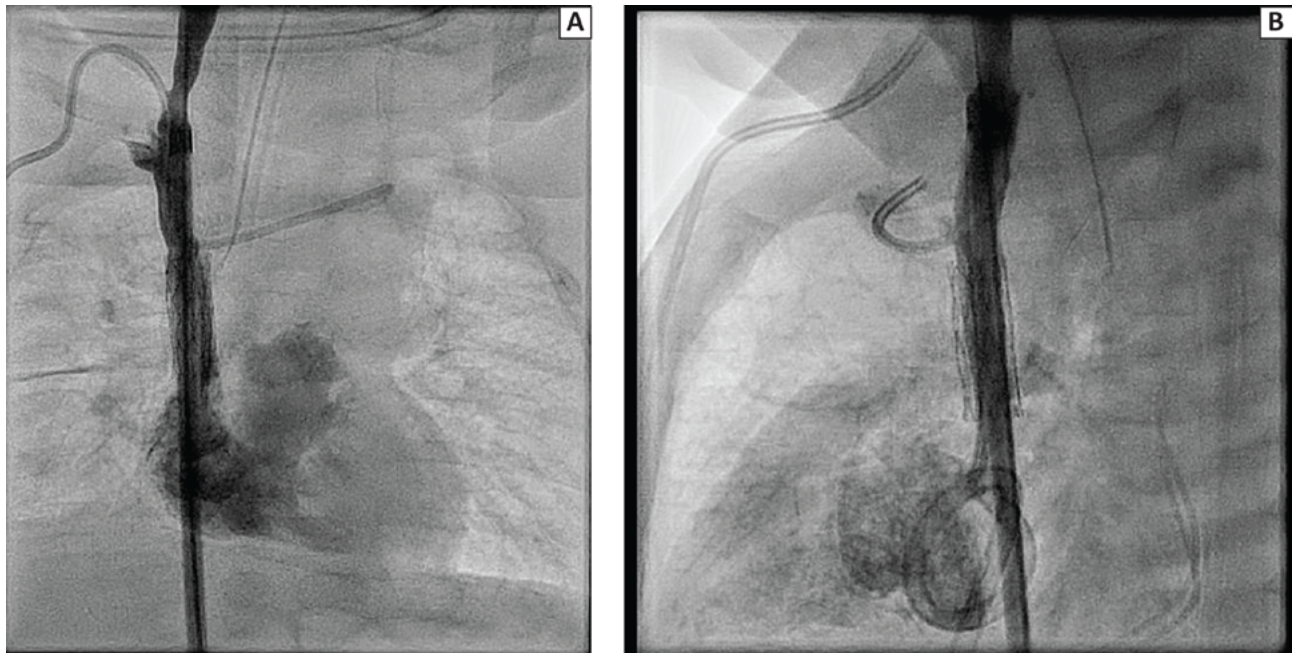
Figure 3. *Panel A*. After implantation of a PG1910XD stent in the superior vena cava and high-pressure dilation to 7 mm, there was increased forward flow across the stent into the right atrium and no retrograde flow in the innominate vein or decompression through collateral veins. Note that the right subclavian central venous line was repositioned in the left innominate vein using a snare catheter from the hemiazygos vein (snaring not pictured). *Panel B*. Lateral view after stent dilation.
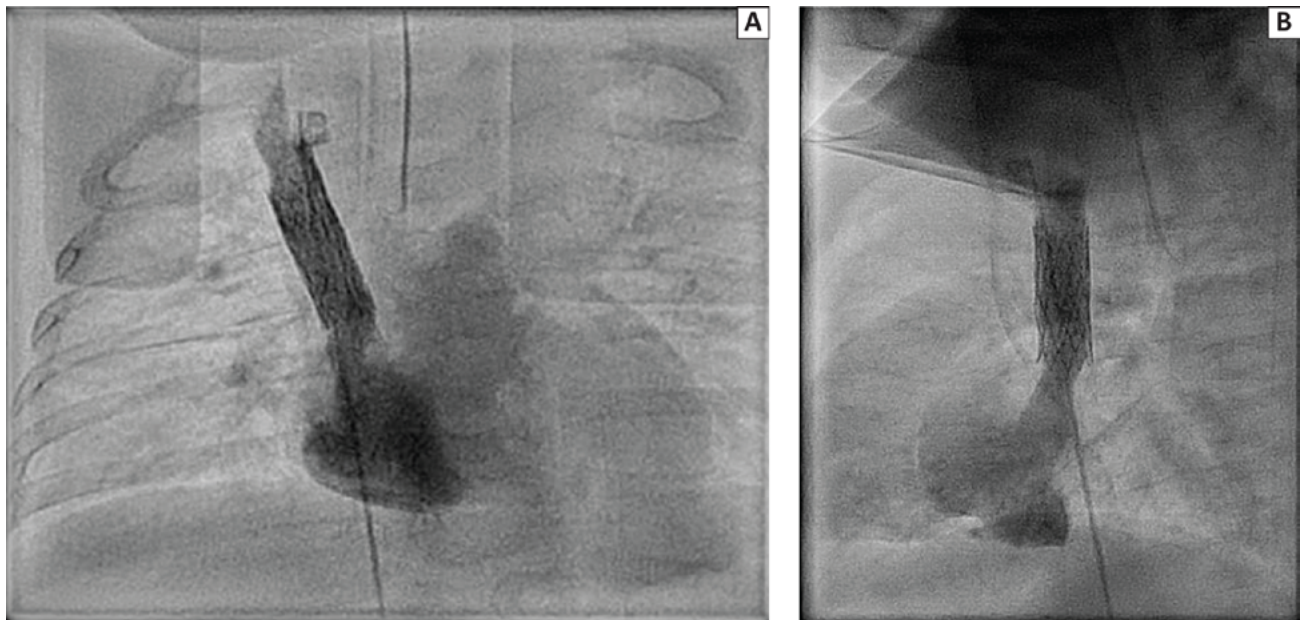

Figure 4. *Panel A*. Two weeks after the initial procedure, there was continued unobstructed flow across the stent and no short-term recurrent stenosis. *Panel B*. Lateral view of 2-week follow-up.

increased pressure in the thoracic duct [5]. The most common site for this type of obstruction is the innominate vein [6], but as demonstrated in this case, the SVC must also be considered as a potential site of vascular compromise. The most common causes of SVC stenosis or obstruction leading to chylothorax in children

are acquired rather than congenital. The three most common etiologies are thrombosis surrounding a port or central line, external compression from tumors or other mass effect, and surgical complications including direct injury of the thoracic duct or postoperative stenosis of the SVC or innominate vein [4, 7, 8, 9, 10].

Congenital—rather than acquired—SVC stenosis as presented in our case is exceedingly rare and thus far has only been reported in the presence of other significant cardiac anomalies. For our patient, the absence of upper body edema or plethora, along with highly developed collateral flow, indicates that this lesion had likely been present since birth. Although the causes of chylothorax are varied, the workup and management are similar across all etiologies. The diagnosis of chylothorax is made based on pleural fluid analysis containing a triglyceride concentration greater than 110 mg/dl, whereas it is excluded by a concentration less than 50 mg/dl. Intermediate values between 50 and 110 mg/dl can be further diagnosed by lipoprotein electrophoresis of the fluid to detect chylomicrons. Alternatively, a ratio of cholesterol-to-triglycerides in pleural fluid of less than 1 is also diagnostic [11]. Chylothorax without a clear underlying cause (e.g., trauma, obstruction, or post-surgical damage) accounts for 5–10% of cases and is typically diagnosed on lymphangiography or lymphoscintigraphy [12].

After initial invasive drainage of the effusion, if indicated for therapeutic or diagnostic purposes, the goal of therapy is to minimize production of chyle medically while treating the offending clot or obstruction, if possible. Medical treatment typically involves a fat-restricted diet supplemented with medium-chain triglycerides, followed by octreotide (0.3–10 mcg/kg/hour titrated to response, with a median dose of 2.8 mcg/kg/hour) and then gut rest and TPN if other measures are unsuccessful. Approximately 80% of pediatric patients respond to medical therapy, although octreotide therapy and TPN carry their own risks [1, 13]. Recommendations vary regarding indications and timing for surgical management with thoracic duct ligation or pleurodesis, but cases are generally considered refractory when effusion persists for more than 2 weeks despite conservative management [14]. Overall, these surgical interventions have a high success rate, and the main risk factor for death or

chylothorax recurrence after surgery is thrombosis of the upper body venous vessels [15], thus confirming the importance of relieving such an occlusion whenever possible.

There is only one previous report of isolated congenital SVC stenosis leading to chylous effusion and respiratory failure; in that case, the patient presented in the immediate neonatal period with severe hydrops and upper body edema consistent with SVC syndrome [16]. Here, we report the first case without an obvious acquired etiology presenting outside the immediate perinatal period. Notably, the patient did not present with obvious clinical findings of SVC syndrome and had developed significant venous collateralization, suggesting that the chronicity of his obstruction likely began during fetal life. Ultimately, his upper body central venous pressure was only mildly to moderately elevated due to collateral decompression, which likely allowed for a relatively slow accumulation of chyle in the pleural space until respiratory decompensation.

Furthermore, our patient's hospital course confirms the importance of understanding the various potential underlying causes of chylothorax and early intervention via relief of venous obstruction when possible. Our patient experienced a delay in treatment (i.e., abnormal echocardiogram on day 2, cardiology consulted on day 10, and the procedure itself performed on day 14) due to the standard of care of 2 weeks of medical therapy before declaring a chylothorax refractory and proceeding with an invasive means of treatment. An argument can be made that if a chylothorax is due to a known venous obstruction, intervention could be performed immediately to relieve this obstruction in lieu of medical management. As such, it is essential to perform the necessary imaging studies to evaluate for venous anomalies, thrombosis, or intrathoracic mass affecting both the innominate vein and/or the SVC.

Our patient's presentation and rapid recovery following acute decompression of the upper body central veins with effective intravascular stent implantation demonstrates that significant chylous effusion can occur from severe congenital vascular obstruction alone, in the absence of surgical complications or other factors. When chylothorax is caused by vascular obstruction, this phenom-

enon is most commonly observed at the site of the innominate vein. However, our case provides an important reminder to consider the SVC as another potential site of stenosis or obstruction, even in the absence of other intracardiac lesions. Anatomic narrowing causing obstruction to lymphatic drainage should be considered in all patients with chylothorax, even in the absence of other signs of venous congestion such as upper body edema. Early catheter-based interventions should be considered because they are effective in relieving vascular obstructions and resolving chylous effusions.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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