

CASE REPORT

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Stent Implantation for Effective Treatment of Refractory Chylothorax due to Superior Vena Cava Obstruction as a Complication of Congenital Cardiac Surgery

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Abstract: Chylothorax is a serious complication of congenital cardiac surgery and is significantly associated with increased morbidity and mortality. Central venous obstruction, which is often related to the insertion of central venous catheters for postoperative management, is known to be an important risk factor for treatment failure and mortality associated with this condition. We present the case of a 6-month-old girl with refractory chylothorax after surgical repair of tetralogy of Fallot. The chylous drainage continued for more than 2 months despite maximal conservative therapy (water restriction, total parenteral nutrition, and infusion of somatostatin and steroid) and surgical ligation of the thoracic duct. Subsequently, we observed stenosis of the superior vena cava (SVC) caused by large thrombi possibly associated with the prolonged use of central venous catheter placed in the internal jugular vein. Because transcatheter balloon dilation failed to relieve the stenosis, we performed stent implantation for the SVC and innominate vein. After the procedure, chylous drainage dramatically reduced, and the patient was discharged from the hospital. In conclusion, central venous obstruction due to thrombosis should be routinely examined when chylothorax is diagnosed and is resistant to conservative therapy after congenital heart surgery. Stent implantation can effectively relieve the venous obstruction and thus be a life-saving treatment option for this difficult condition.

Keywords: chylothorax, thrombosis, central venous catheter, stenting

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Introduction

Chylothorax is an infrequent but a serious complication of pediatric cardiovascular surgery and could often jeopardize postoperative recovery. The incidence of this condition has been increasing over the past decades, possibly due, in part, to the increased complexity of cardiac surgery.¹ Central venous obstruction, which is often related to the insertion of central venous catheters for postoperative management,^{2,3} is known to be an important risk factor for treatment failure and mortality associated with this condition.⁴⁻⁷ In this report, we present the case of an infant with prolonged chylothorax associated with thrombosis or obstruction of the superior vena cava (SVC). The chylothorax was resistant even to thoracic duct ligation performed after maximal conservative therapy. However, stent implantation for the SVC dramatically resolved the chylous drainage.

Case report

The patient was a 6-month-old girl with a small right ventricle (70% of normal) who underwent surgical correction of tetralogy of Fallot. She had postoperative heart failure represented by low cardiac output and elevated central venous pressure of 13 mmHg. She presented with massive pleural drainage from postoperative day 1. Examination of the drained fluid revealed cell counts of 4,250 cells/ μ L with a predominant lymphocyte fraction of 99%. The triglyceride levels were as high as 144 mg/dL, which confirmed that she had chylothorax. Because the chylous drainage did not reduce with the generally recommended conservative therapy, including water restriction (60 mL \cdot kg⁻¹ \cdot day⁻¹) and total parenteral nutrition,

we initiated administration of a somatostatin analogue at a dose of 10 μ g \cdot kg⁻¹ \cdot h⁻¹ on postoperative day 7. Nonetheless, chylous drainage persisted, and thoracic duct ligation was performed on postoperative day 35. Although chylous drainage temporarily decreased after the surgery, it did not stop completely and increased gradually while supportive therapies, including water restriction, total parenteral nutrition, and somatostatin analogue infusion, were continued. Steroid administration was also ineffective. An echocardiogram on postoperative day 70 revealed stenosis of the SVC due to large thrombi (Fig. 1). A central venous catheter had been placed in the right internal jugular vein from postoperative day 0 to 10 and from day 28 to 58. An important observation was that there was an unexpected decrease in the platelet count from 444 \times 10³/ μ L on postoperative day 44 to 152 \times 10³/ μ L on postoperative day 54. Balloon dilation for the SVC stenosis performed on postoperative day 80 (Fig. 2) only temporarily and partially reduced the chylous drainage. The chylous drainage began to increase 1 week after the procedure. Thrombosis developed despite the administration of tissue plasminogen activator followed by continuous infusion of heparin. Therefore, we performed stent implantation in the SVC (Express LD 8 mm \times 27 mm) and innominate vein (Express LD 7 mm \times 17 mm) on postoperative day 102 (Fig. 3). Stent implantation dramatically reduced the chylous drainage from 40 mL \cdot kg⁻¹ \cdot d⁻¹ to 3.5 mL \cdot kg⁻¹ \cdot d⁻¹ (averaged during the 5 days before and after the procedure). The chest tube was successfully removed, and the patient was finally discharged from the hospital with diuretics and dietary control with milk enriched with medium-chain triglycerides.

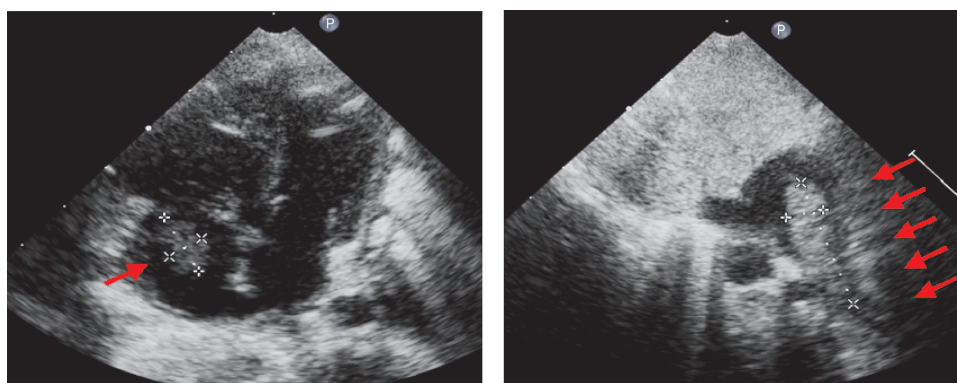


Figure 1. Two-dimensional transthoracic echocardiography showing the large-sized thrombus (arrows). Four-chamber view in the left and superior vena cava seen in the right atrial view in the right.

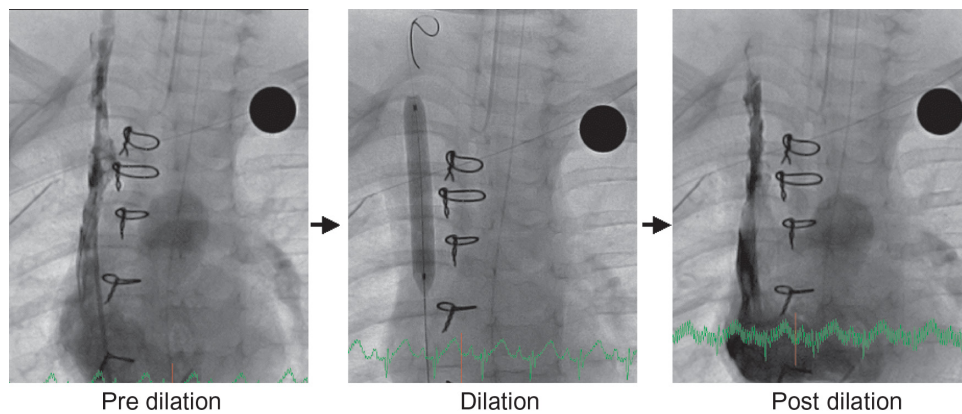


Figure 2. Transcatheter balloon dilation of the superior vena cava (SVC). Note that the lumen of the SVC remained irregular after the procedure, indicating incomplete relief of the stenosis.

Discussion

Chylothorax is a serious complication of cardiac surgery and is associated with increased mortality. Because the thoracic duct enters the venous system at angulus venosus, central venous obstruction in the upper body and/or high central venous pressure inevitably causes resistance to the flow of the lymph into the venous system, which leads to congestion of the lymph thus resulting in chylous drainage into the pleural space.^{8,9} This pathophysiology is directly linked to the well-recognized clinical finding that central venous obstruction and/or high central venous pressure is often associated with prolonged high volume of chylous drainage and is a significant risk factor for treatment failure.^{4-7,10} Central venous catheterization required for administration of medication or parenteral nutrition after cardiac

surgery is known to be an important cause of central venous thrombosis.^{2,3,11} Once chylothorax develops, prolonged chylous drainage, in turn, necessitates the use of a central venous catheter for a prolonged period for parenteral nutrition, which potentially leads to a viscous cycle of thrombosis formation and chylous drainage. Antithrombin lost in chyle^{2,12} and/or vascular inflammation induced by hyperalimentation solution² may also contribute to increased susceptibility for the development of thrombosis in patients with chylothorax.

In our patient, chylothorax developed soon after surgery, which suggested that operative injury to the thoracic duct was the primary cause of chylothorax.¹ Elevated central venous pressure possibly associated with an underdeveloped right ventricle could have further accelerated and prolonged the chylous drainage.

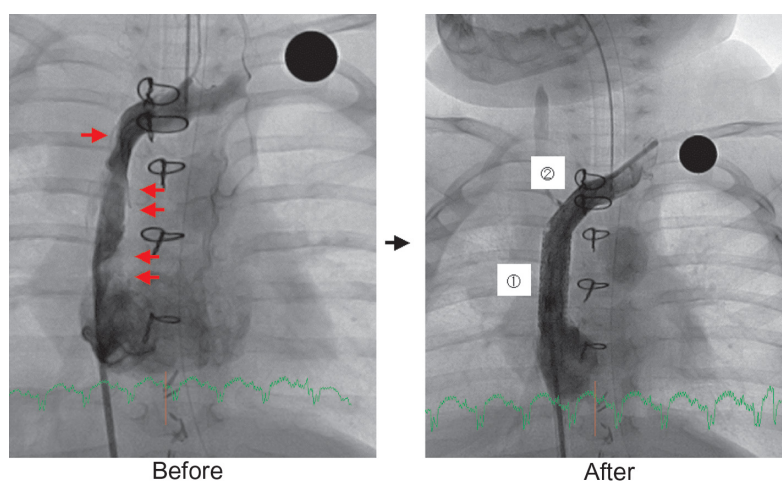


Figure 3. Stent implantation in the superior vena cava. Angiographic defects due to thrombi-induced stenosis (indicated by arrows in the left panel) were completely resolved after the stent implantation (right).



Formation of venous thrombosis was also believed to have contributed significantly to the refractory chyloous drainage observed even after thoracic duct ligation, because chylothorax resolved after the venous obstruction was relieved by stent implantation. Nath et al reported an important finding that although the overall outcome of surgical thoracic duct ligation for chylothorax refractory to conservative medical therapy is excellent, patients with upper body venous thrombosis were at a high risk of treatment failure and mortality.⁷ Considering the clinical course and the huge size of the thrombus formed in our patient, it might be assumed that the process of thrombus formation might have started much earlier than we recognized it. Thus, the findings in our patient and those in previous reports indicate the importance of high index of suspicion for central venous thrombosis and obstruction, especially in patients with refractory chylothorax. A decrease in platelet count, as observed in our patient, and possibly an increase in the level of D-dimers may provide useful information for this purpose.

Conventionally, surgical intervention is indicated when conservative therapy fails to control the lymphatic leak; however, specific and precise criteria for abandoning conservative therapy remain to be standardized.^{1,6,11} A consensus for treatment of chylothorax with central venous stenosis/occlusion remains to be established. To our knowledge, this is the first report on stent implantation for stenosis of the SVC due to thrombosis for effective treatment of otherwise uncontrollable chylothorax after congenital cardiac surgery. Our findings are in line with those reported in an infant born with congenital SVC obstruction in whom successful placement of an intravascular stent led to the resolution of the chylothorax with rapid clinical improvement.¹³ Balloon angioplasty can be an alternative to stent implantation, but in our patient, the treatment efficacy of stenting was greater than that of balloon dilation. Administration of anticoagulants or thrombolytics may be effective in some patients with chylothorax associated with thrombosis,^{3,11} but these drugs were ineffective in our patient.

Conclusion

Central venous thrombosis and resultant venous stenosis/occlusion can cause a life-threatening refractory chylothorax after congenital heart surgery. Stent

implantation can effectively relieve the venous obstruction and thus be a life-saving treatment option for this difficult condition. Early diagnosis and intervention may lead to early recovery and eliminate unnecessary treatments. Thus, physicians should maintain a high index of suspicion for venous obstruction when chylothorax is diagnosed and is resistant to conservative therapy.

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Disclosures

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