

# Surgical Reconstruction for High-Output Chylothorax Associated with Thrombo-Occlusion of Superior Vena Cava and Left Innominate vein in a Neonate

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We report a case of high-output chylothorax associated with thrombo-occlusion of the superior vena cava (SVC) and left innominate vein (LIV) following an arterial switch operation in a neonate. The chylothorax was resolved by 3 weeks after surgical reconstruction of the SVC and LIV using fresh autologous pericardium. We confirmed the patency of the SVC and LIV with a 1-year follow-up computed tomographic scan at our outpatient clinic.

*Key words:* 1. Chylothorax  
2. Thrombosis  
3. Surgery

## Case report

A baby was born at 38 weeks of gestational age with a prenatal diagnosis of transposition of the great arteries (TGA) and intact ventricular septum (IVS). After birth, oxygen saturation remained low despite supplemental oxygen, ventilator care, and prostaglandin infusion, and an echocardiographic exam showed an aneurysmal interatrial septum without communication, in addition to TGA with IVS. Emergent balloon atrial septostomy was performed to maintain adequate oxygenation and stabilize the baby before corrective surgery. However, increased inotrope and vasopressor requirements led to urgent corrective surgery at the age of 2 days.

After an uneventful arterial switch operation (ASO), chylothorax developed on postoperative day 10 and persisted for 10 days with an increasing

amount of drainage (up to 100 mL/kg/day) despite conventional medical treatment, including the administration of low-fat medium-chain triglycerides by mouth, no food by mouth with total parenteral feeding, and intravenous administration of octreotide. Follow-up echocardiography and computed tomography (CT) revealed severe stenosis of the left innominate vein (LIV) and superior vena cava (SVC) (Fig. 1), which was strongly suspected to be the main cause of the high-output chylothorax. However, the patient did not have any symptoms of SVC occlusion, such as swelling or engorged veins in the face, arm, and upper trunk. The levels of protein C (61%; reference value in infants, 15%–50%) and protein S (111%; normal range in infants, >50%) were not deficient, but the level of anti-thrombin III (40%; normal range in infants, 60%–90%) was significantly decreased. In previous laboratory exams, the level of

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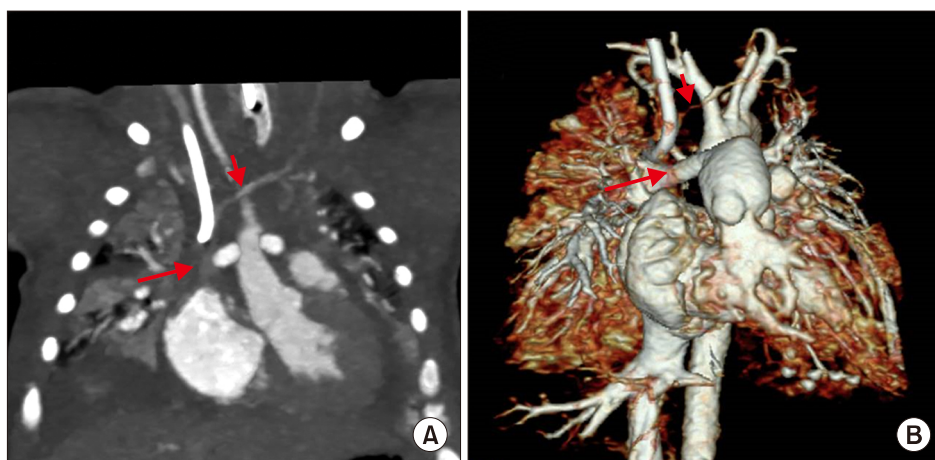


Fig. 1. (A, B) A preoperative computed tomographic scan shows the severely obstructed superior vena cava (long arrow) and left innominate vein (short arrow).

anti-thrombin III before and after the ASO was 39% and 51%, respectively. The patient had a central catheter for 19 days prior to the second operation. It was in the left jugular vein for 17 days, and was then revised to the right jugular vein immediately after the diagnosis of thrombo-occlusion of the SVC and LIV. On postoperative day 19, thrombectomy and reconstruction of the LIV and SVC were performed under normothermic cardiopulmonary bypass. An SVC cannula was inserted through the right atrial appendage. After incising the SVC and LIV, we found that the junction between the SVC and right atrium was almost occluded and that the lumen of the SVC and LIV was severely narrowed, with white thrombi and irregularly thickened intima. Thrombectomy was performed using a fully ballooned 3-Fr Fogarty catheter (Edwards Lifesciences Co., Irvine, CA, USA) and a 0.4-mL ballooned 4-Fr Fogarty catheter (Edwards Lifesciences Co.) through the left subclavian vein and left internal jugular vein. Finally, the SVC and LIV were reconstructed with fresh autologous pericardium (Fig. 2A, B).

Unfractionated heparin was used to maintain adequate levels of anticoagulation (4–12 U/kg/hr; target activated partial thromboplastin time, 60–80 seconds) for 14 days until enteral feeding was initiated. The amount of pleural drainage gradually decreased, and all tubes could be removed 3 weeks after the second operation. Two months after the second operation, the levels of protein C, protein S, and anti-thrombin III were 46%, 111%, and 69%, respectively. At the outpatient clinic, warfarin was administered for 3 months (target international normalized ratio,

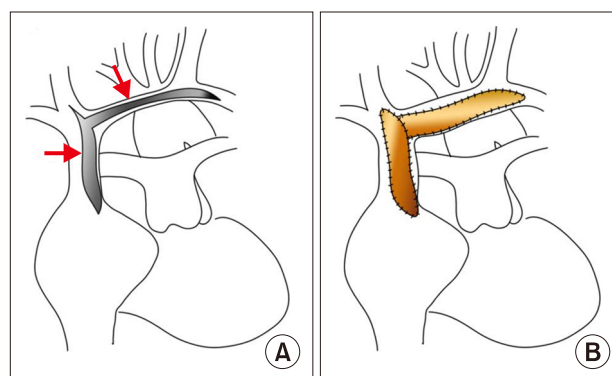


Fig. 2. Diagram of the operative procedures. (A) Arrows indicate the range of the incision for the thrombectomy and reconstruction. (B) The superior vena cava and left innominate vein were reconstructed using fresh autologous pericardium.

1.5–2.0) and a 1-year follow-up CT scan showed broad patency of the LIV and SVC (Fig. 3A, B).

## Discussion

Chylothorax is a significant complication after surgery for congenital heart disease. The development of chylothorax can lead to respiratory difficulty, malnutrition, and deficiencies in the immunologic and hematologic system, and can be associated with other morbidities and mortality [1]. There are many causes of chylothorax, but high-output chylothorax can be caused by thrombo-occlusion of the upper venous system. Thrombosis can occur due to certain acquired clinical conditions or hereditary risk factors involving blood composition, vessel wall integrity, and blood flow [2,3]. Furthermore, since functional

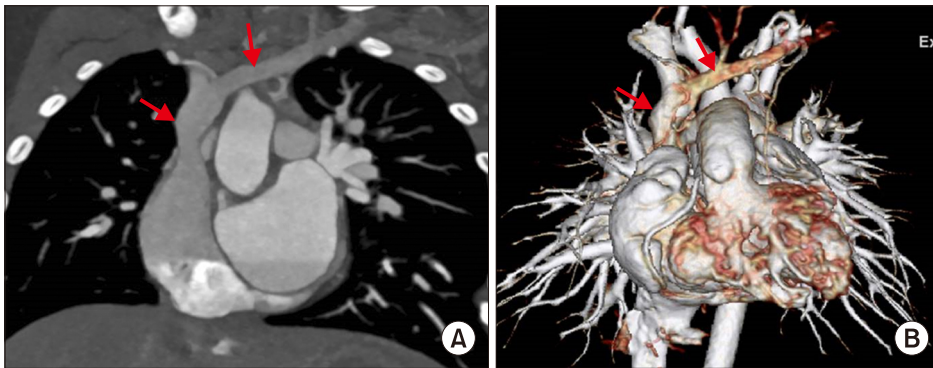


Fig. 3. (A, B) A postoperative computed tomographic scan reveals luminal patency of the superior vena cava and left innominate vein.

anti-thrombin levels and anticoagulant protein levels are normally low in neonates and infants due to developmental hemostasis [3], the hypercoagulability test must be performed for the diagnosis of suspected thrombosis in neonates or infants. In our case, the patient was a newborn and had risk factors for a thrombus, including a surgical history for congenital heart disease and the presence of a central venous line. We also observed low anti-thrombin III levels, which could have caused the thrombus.

Although thrombosis of the venous system can be treated with conservative treatment, including anticoagulant therapy, catheter-based interventions and surgical reconstruction with homologous grafts have also been reported to be effective treatments for upper venous thrombosis with complications [4-6]. In our case, chylothorax associated with thrombo-occlusion of the upper venous system was effectively treated with a surgical intervention, and the luminal opening was well maintained on 1-year follow-up imaging.

In summary, high-output chylothorax unresponsive to conventional medical therapy may be suspicious for upper venous thrombosis, and an accurate diagnosis should be made through imaging and laboratory tests. Active surgical treatment could be helpful in such cases, and we found that fresh autologous pericardium might have acceptable durability as a venous reconstruction material.

### Conflict of interest

No potential conflict of interest relevant to this article was reported.

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