CASE REPORTS

CONSERVATIVE TREATMENT IN AN INFANT WITH SUPERIOR VENA CAVA SYNDROME AFTER CARDIAC SURGERY

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Abstract: Superior vena cava (SVC) syndrome rarely responds to conservative treatment. We report the case of a 2-month-old boy with SVC syndrome and bilateral chylothorax after surgical repair of the hemitruncus. Medical management with low-dose heparin, dipyridamole, and aspirin resulted in improvements of head swelling and chylothorax. The chyle had disappeared 46 days postoperatively. Compensatory growth of collateral vessels was also found. Although surgical repair is sometimes advocated in patients with SVC syndrome, medical treatment is an important alternative if the risk of surgery is too high.

Superior vena cava (SVC) syndrome results from blood flow obstruction through the superior cava vein. In more than 80% of patients, this morbidity is due to compression by a malignant tumor [1]. Echocardiography is helpful in the early detection of, and intervention for, SVC obstruction [2]. When SVC pressure rises, the amount of lymphatic leakage from the thoracic duct will increase and result in chylothorax. The chyle involves large amounts of lymphocytes, proteins, and immunoglobulins spilling into the pleural cavity. Morbidity and mortality are high without early surgical intervention [3]. We report the case of a 2-month-old boy who developed SVC syndrome complicated with chylothorax after cardiovascular surgery for hemitruncus. The possible causes included decannulation during open heart surgery or repeated central venous catheter insertion. Under medical treatment with low-dose heparin, dipyridamole, and aspirin, the chyle disappeared from the pleural cavity 46 days after the operation.

Case Report

A 2-month-old boy was noted to have prolonged feeding time without cyanosis since birth. A heart murmur was accidentally found during physical check-up for an episode of respiratory tract infection. Cardiac catheterization and angiography performed at age 1.5 months revealed a hemitruncus with a right pulmonary artery arising from the ascending aorta, and a large patent ductus arteriosus (PDA) combined with pulmonary hypertension. As progressive dyspnea developed, an operation was arranged for PDA ligation and re-implantation of the right pulmonary artery to the main pulmonary artery (Fig. 1) at age 2 months (Day 1 was defined as the day of surgery). Five episodes of seizure were noted 3 days later. Brain computerized tomography (CT) studies showed subdural hematoma at the left frontal area without hydrocephalus. The seizures were controlled with phenobarbital.

Cough, poor feeding, and progressive dyspnea developed thereafter. Echocardiography showed decreased blood flow in the SVC on Day 9. Chest ultrasonography revealed bilateral...
pleural effusion with right upper lung consolidation. Pleural effusion study showed elevated triglyceride (346 mg/dL) and low cholesterol (37 mg/dL) concentrations compared to plasma triglyceride (66 mg/dL) and cholesterol (66 mg/dL) concentrations on Day 13. Chylothorax was suspected and nutritional support was started with Portagen formula (Mead Johnson, Nijmegan, the Netherlands), containing low fat (22%) and high protein (16.5%), with the fat composed of 86% medium-chain triglycerides, on Day 23. Because dyspnea with swelling of the head and upper trunk progressed, an endotracheal tube was inserted along with bilateral chest tubes. Ultrafast CT of the chest on Day 21 confirmed the SVC obstruction (Fig. 2). Balloon dilatation was delayed till Day 26 due to unstable vital signs. However, angiography showed complete SVC occlusion under the level of the innominate vein, and contrast medium drained via the collateral veins to the hemizygos vein (Fig. 3). Balloon angioplasty or stent implantation for opening the occluded vessels was scheduled, but was not feasible due to the high risk of SVC perforation in performing the procedure, and the collateral veins were considered adequate. Medical management without surgery was used with continuous infusion of low-dose heparin $2 \, \text{U} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$ along with $5 \, \text{mg} \cdot \text{kg}^{-1} \cdot \text{d}^{-1}$ dipyridamole and $3.3 \, \text{mg} \cdot \text{kg}^{-1} \cdot \text{d}^{-1}$ aspirin. The right chest tube was removed successfully on Day 41 (32 d after SVC syndrome onset), the left chest tube was removed on Day 55 (46 d after SVC onset), and heparin was discontinued on Day 46. Chest roentgenogram and chest ultrasonography 2 months later revealed resolution of the chylothorax; cardiac ultrasonography 6 weeks later showed many collateral vessels to the SVC. After 4 months, feeding was smoothly switched to usual infant formula without recurrence of chylothorax.

**Discussion**

Obstructed or high pressure SVC interrupts the normal venous return of blood from the head, upper extremities, and thorax to the right atrium. Thus, venous blood must return to the heart through the azigos veins and collateral vessels, which enter the inferior vena cava (IVC). When the pressure of the venous system exceeds the pressure in the thoracic duct (10–25 cmH$_2$O), rupture of the duct, lymphatic leakage resulting in chylothorax, and collateral vessel development can be expected. In 1936, Blalock et al showed that occlusion of the SVC could produce chylothorax [3]. SVC obstruction may be caused by intrathoracic neoplasm, aortic aneurysm, or iatrogenic procedures such as the development of subsequent venous thrombosis after the use of in-dwelling central venous catheters.
venous catheters, after decannulation of extracorporeal membrane oxygenation, or by cardiac bypass surgery [2]. Certain cardiovascular procedures are more prone to this complication, such as Fontan or Senning procedures and bicaval pulmonary anastomosis.

Complete SVC occlusion with development of SVC syndrome is an unusual complication after decannulation following open heart surgery [4]. A review of the literature found that SVC syndrome with chylothorax in children was only reported in 14 cases of transposition of the great artery (TGA) after Mustard’s [5] and Senning’s operations [6], and that it is an unusual complication of surgery of the hemitranunus. Among 14 reported cases, only two had bilateral chylothorax [5]. Chest CT and angiography to locate the site of SVC obstruction and define important collateral pathways are the best method for early detection of SVC obstruction [2]. In our patient, the SVC obstruction was below the level of the azygos arch as seen on ultrafast CT, and was defined as Grade IV, which indicates a high risk and poor prognosis [7].

While SVC syndrome with chylothorax developed 1 week after open heart surgery in our patient, it may have been caused by repeated central venous catheter insertion or cannulation for cardiopulmonary bypass. Stent implantation was not performed because of the high risk of SVC rupture when it is totally obstructed, and also because collateral vessels were developing without any hydrocephalus sequelae. Thus, medical management was tried initially. Although surgery has been recommended if the amount of chyle leakage is more than 15 mL/kg in several reports [8], this could deteriorate the development of thrombolized SVC, and increase the risk of collateral vessel destruction or rupture because of the many prominent posterior vertebral veins, as seen on chest CT. Beghetti et al suggested increasing the conservative treatment duration to 30 days for chylothorax, because they observed that 80% of patients had a good response to conservative treatment, and the chyle leakage rate was suspected to be higher in patients with surgery than in those who received conservative treatment [8]. Although treatment of chylothorax induced by SVC syndrome requires more time and a higher volume of chyle leakage increases the risk of failure for conservative treatment [8], our patient was successfully treated medically with heparin for 46 days, and was then switched to dipyridamole and aspirin. Successful conservative treatment for SVC syndrome in an infant with bilateral chylothorax after cardiac surgery has been reported, and achieved complete resolution of the chylothorax after 15 days [9]. The rapid development of collateral circulations in children, which is more prominent than in adults, plays an important role in successful conservative treatment of SVC syndrome. In our patient, the azygos, hemiazygos, and IVC system collateral pathways developed within 2 weeks after the clinical symptoms of head and neck swelling. A literature review found that thrombolytic therapy has been used successfully only in rare cases [10]. Heparin is thought to relieve SVC syndrome through the stabilization of the capillary endothelium via reducing the leakage of protein into the extravascular space, a thrombolytic effect, and the alteration of the cytoskeletal architecture in the perivascular space to augment resorption of protein into the lymphatic return [11].

Early performance of echocardiography or ultrafast CT evaluation in patients with suspected SVC syndrome after bypass surgery is recommended, and early intervention with thrombolytic therapy and stent implantation are worth trying before a second operation to prevent or reduce the severity of SVC syndrome or chylothorax.

References