Occlusion of Azygos Vein Via Direct Percutaneous Puncture of Innominate Vein Following Cavopulmonary Anastomosis

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SUMMARY
A 2-year-10-month-old boy was diagnosed with a complex congenital heart disease: right atrial isomerism, left superior vena cava (LSVC), complete atrioventricular septal defect, secundum type atrial septal defect, transposition of the great arteries with pulmonary atresia, patent ductus arteriosus, absence of a right superior vena cava (RSVC), and dextrocardia. He had received a left Blalock-Taussig (BT) shunt at the age of 3 months and a left bidirectional Glenn shunt one year after BT shunt. Progressive cyanosis was noted after the second operation and cardiac catheterization showed a functional Glenn shunt with an engorged azygos vein, which was inadvertently skipped for ligation. Because of the absence of RSVC, transcatheter occlusion of the azygos vein was performed successfully via direct puncture of the innominate vein. (Jpn Heart J 2000; 41: 235-238)

Key words: Azygos vein, Interventional cardiac catheterization, Child

TRANSCATHETER coil embolization is a well-established procedure used for occlusion of many congenital vascular anomalies in children. The reported vascular anomalies include patent ductus arteriosus, systemic to pulmonary collateral arteries, coronary arterial fistula, pulmonary arteriovenous malformation, pulmonary sequestration, abnormal vein connection, and surgically created systemic to pulmonary artery shunt and fenestration in Fontan baffle.1-3) We describe a 2-year-10-month-old boy with an engorged azygos vein after a left bidirectional Glenn shunt for a complex congenital heart disease, which was successfully occluded with a coil via percutaneous puncture of the innominate vein.
CASE REPORT

A 2-year-10-month-old boy presented with cyanosis and a systolic murmur over the right middle sternal border at 3 months of age. Chest X-ray showed dextrocardia with decreased pulmonary vascularity. The liver shadow was transverse across the abdomen. Electrocardiogram showed dextrocardia and superior QRS axis. Howell-Jolly bodies were found in the peripheral blood smear. Two-dimensional echocardiography and color Doppler studies demonstrated right atrial isomerism, left superior vena cava (LSVC), complete atrioventricular septal defect, secundum type atrial septal defect, transposition of the great arteries with pulmonary atresia, patent ductus arteriosus and dextrocardia. Cardiac catheterization and angiography confirmed the echocardiographic findings. The additional angiographic findings were rudimentary pulmonary trunk with mild hypoplastic bilateral pulmonary arteries, absence of the right superior vena cava (RSVC) and left internal jugular vein. He adequately tolerated a palliative left Blalock-Taussig shunt. Progressive cyanosis was noted 1 year later. The second catheterization at the age of one year and three months showed a nearly totally occluded BT shunt. Therefore, a left bidirectional Glenn shunt was performed (LSVC anastomosis with left pulmonary artery). His condition was satisfactory after this procedure. However, progressive dyspnea and cyanosis were noted again one and a half years later. A third catheterization was done and the LSVC angiography showed a functional left bidirectional Glenn shunt and massive contrast material effluxed inferiorly into an unsuspected engorged azygos vein (Figure 1A, B). Reviewing the operative record of the bidirectional Glenn shunt, we found that the azygos vein was inadvertently skipped for ligation.

After explaining to the parent and obtaining their written informed consent, we decided to perform embolization of the azygos vein. Because of absence of the RSVC, if the catheter was inserted from the right jugular vein, it must follow a long course to the LSVC, then posterior to the azygos vein. Taking into consideration the long and zigzag vascular course and difficult manipulation of the catheter, we decided to embolize the azygos vein via direct puncture of the innominate vein.

Under general anesthesia and sterilization of the skin, a #18 needle was used to puncture at the cross point of the second intercostal space and right middle clavicle line. The needle directed to the patient’s left posterior axillary line and the angle was 45 degrees to the skin. After smooth puncture of the innominate vein and approach into the azygos vein, selective injection of a contrast material showed its large diameter (6.4 mm) and long course down to the abdomen (Figure 1C). The size of the coil was chosen according to well-documented principles. The azygos vein was entered with a C1 5.5-French catheter and a non-attached
system with Dacron stranded, stainless steel coil (MWCE-38-15-15, COOK Inc, Bloomington, Indiana, USA) was deployed. Angiography of the LSVC performed 10 minutes later confirmed successful occlusion of the azygos vein (Figure 1D). The oxygen saturation (SaO2) elevated from 74 % to 90 % in 5 minutes. On the next day, he was discharged with an SaO2 of 86-89 %.
DISCUSSION

It is well established that an azygos vein should be ligated in the case of a Glenn shunt. Inadvertent miss of this procedure will result in shunting of the blood flow into the azygos vein and decreased flow to the pulmonary arteries. The patient rapidly becomes cyanotic. In this case of absent RSVC and left internal jugular vein, to avoid the long and zigzag catheter course via the subclavian vein which may make embolization of the azygos vein difficult, we think that direct puncture of the innominate vein makes the manipulation much easier. However, an accurate puncture is important to prevent pneumothorax or even a hemopneumothorax because of the poor compression effect for hemostasis.

REFERENCES