Successful surgical treatment of hypoplastic left heart syndrome associated with a divided left atrium that was diagnosed intraoperatively

Yuji Naito, MD, a Yorikazu Harada, MD, a Shunji Uchita, MD, a Koki Takizawa, MD, a Gengi Satomi, MD, b Satoshi Yasukochi, MD, b and Hikoro Matsui, MD, b Nagano, Japan

Hypoplastic left heart syndrome (HLHS) often involves abnormalities in atrial septal morphology, including a restrictive atrial communication and anomalous attachment of a malaligned septum primum. Cor triatriatum is a rare congenital heart abnormality in which the left atrium (LA) is divided into 2 chambers by a diaphragm. Association of cor triatriatum with HLHS is rare, and cor triatriatum is difficult to diagnose preoperatively. We herein describe an unusual case of HLHS with cor triatriatum.

Clinical Summary
The patient was a neonate (7-day-old boy, 2.46 kg) in whom HLHS was diagnosed prenatally. Delivery was uncomplicated at 37 weeks’ gestation. Intravenous infusion of prostaglandin was initiated to maintain the systemic circulation through the patent ductus arteriosus. Chest radiography showed an enlarged cardiac shadow with a cardiothoracic ratio of 58% and lung congestion with signs of severe pulmonary venous congestion. Echocardiography revealed anatomy consistent with HLHS: restrictive patent foramen ovale (PFO), patent ductus arteriosus, persistent left superior vena cava (SVC), severe mitral valve stenosis, and severe aortic valve stenosis. The progressive pulmonary congestion was presumed to be due to the restrictive PFO, and first-stage palliation was indicated.

The patient underwent Norwood repair at 8 days of age. An abnormal pulmonary venous connection was anticipated from the unidentified anatomic structure found at the superior surface of the LA. After initiation of cardiopulmonary bypass, hypothermic cardioplegic arrest was induced. The right atrium (RA) was cut open to confirm that the fossa ovalis was intact, with no interatrial communication when the right pulmonary artery by using continuous sutures because the atrial septum, which was enlarged during the first operation, was the communication between the RA and the CPVC. The patient gradually showed symptoms of pulmonary venous obstruction, and echocardiography showed accelerated left-to-right shunt flow across the surgically enlarged interatrial communication.

Second-stage palliative surgery consisting of a bidirectional Glenn anastomosis and creation of unrestricted communication between the CPVC and RA was successfully performed when the patient was 5 months old and weighed 3.8 kg (Figure 2B). After establishment of cardiopulmonary arrest, the RA was reopened. Through the surgically created PFO, the atretic mitral valve and left atrial appendage were confirmed, but the aperture on the diaphragm was not identified. Both the atrial septum and the diaphragm were widely excised to create sufficient communication from the CPVC to the RA. After resection of the diaphragm, the unobstructed orifices of all pulmonary veins were found in the CPVC. The right SVC alone was anastomosed to the right pulmonary artery by using continuous sutures because angiographic examination after first-stage palliation had confirmed that the left SVC was occluded.

The postoperative course was uneventful, MRI depicted an unrestricted pulmonary venous pathway, and echocardiography showed no acceleration of the pulmonary venous drainage.

Discussion
Association of cardiac anomalies with HLHS is uncommon. Such anomalies reportedly include intact atrial septum, total anomalous pulmonary venous connection (TAPVC), levopatral cardinal vein, coronary sinus atresia, atritic pulmonary veins, complete atrioventricular septal defect, and transposition of the great arteries.1 Eidem and Cetta2 first described the rare association between cor triatriatum, which is not a well-recognized morphologic defect, and HLHS.2 Cor triatriatum is a rare congenital cardiac anomaly in which the pulmonary veins enter a posteroseparable proximal left atrial chamber that is separated from the anteroinferior...
distal left atrial chamber bearing the mitral valve and left atrial appendage by a diaphragm in which there are 1 or more restrictive ostia.

In the present case the clinical diagnosis presented some morphologic puzzles. The connection between the CPVC and RA could have been diagnosed as either TAPVC or cor triatriatum. In cases of cor triatriatum, each pulmonary vein can be considered as not joining the LA but rather as entering a separate chamber, called the CPVC or proximal left atrial chamber, which is analogous to the common pulmonary venous sinus found in patients with TAPVC. In this respect cor triatriatum is similar to TAPVC, but the pulmonary veins in cor triatriatum are incorporated into the structure of the LA, whereas in TAPVC the pulmonary veins connect to sites separate from the LA.3 Because the diaphragm between the LA and CPVC was confirmed and resected in our patient’s second palliative operation and all pulmonary veins entered a single chamber thought to be part of the LA, we concluded that the atrial morphology was that of cor triatriatum with a morphologic classification of IB1 according to the Lucas–Schmidt system or subtype A2 according to Lam’s classification system.

The prospective detection rate of pulmonary venous abnormalities by means of MRI is reported to be superior to that by means of echocardiography.4 If pulmonary venous abnormality is suspected and the neonate’s hemodynamic condition is stable, MRI can provide useful information.

Figure 1. Photograph obtained during the first-stage palliative operation. The arrow indicates the orifice of the communication between the common pulmonary vein chamber (CPVC) and the right atrium (RA). RV, Right ventricle.

Figure 2. Schemas of atrial and ventricular morphology in the first-stage (A) and second-stage (B) palliative operations. A, The intact fossa ovalis (solid circle) was cut open to create an unrestricted interatrial communication. The stenotic communication between the CPVC and RA (dashed circle) was enlarged to relieve the pulmonary venous congestion. B, Both the atrial septum and the diaphragm (solid circle) were widely excised to create sufficient communication from the common pulmonary vein chamber to the right atrium. CPVC, Common pulmonary venous chamber; IVC, inferior vena cava; LA, left atrium; LSVC, left superior vena cava; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava.
In pulmonary arterial sling (PAS), the left pulmonary artery (LPA) arises from the right pulmonary artery (RPA) and goes leftward between the trachea and the esophagus. This produces a sling around the distal trachea and the proximal bronchi. The LPA thus compresses the superior part of the right bronchus and the distal part of the trachea. Most patients with PAS have clinical symptoms related to tracheal or tracheobronchial compression. When tracheal hypoplasia coexists (ring-sling complex), acute episodes of dyspnea and cyanosis are common and may cause major respiratory distress and death. Although the effect of PAS on lung ventilation is well known, the effect of the surgical correction of PAS on left lung perfusion has never been evaluated.

Clinical Summary
A 5-year-old boy with a history of recurrent pulmonary infections was referred to the pediatric pneumology department.

Preoperative examination. On examination, the patient was an alert and vigorous child weighing 18 kg. Results of clinical examination at rest were normal. A chest radiograph revealed that the left lung was smaller than the right lung, with a minimal shift of the heart and mediastinal structures to the left. Bronchoscopy revealed a mild compression of the distal trachea by as much as a third of its lumen. A 2-dimensional echocardiogram showed a PAS with a small LPA (7-8 mm) and a large RPA (13-14 mm). A multislice computed tomographic scan confirmed the presence of the PAS and showed the smaller LPA stretched around the trachea (Figure 1).

Ventilation scintigraphy with xenon 133 revealed a mild ventilation asymmetry (55% for the right lung and 45% for the left lung), whereas perfusion scintigraphy with albumin, aggregated technetium macro aggregated albumin labeled with Technetium-99m (Tc 99m MAA) revealed severe hypoperfusion of the left lung, with a right to left perfusion ratio of 90% to 10% (Figure 2, A).

Surgical correction of the vascular malformation was elected to preserve both the perfusion and the function of the left lung.

Operative technique. The operation was performed through a median sternotomy under normothermic cardiopulmonary bypass. The ligamentum arteriosum was divided. The LPA was detached from the RPA, dissected free where it coursed behind the trachea, and translocated to the left side of the trachea. It was then shortened to be implanted to the left side of the main pulmonary artery where the ligamentum arteriosus was inserted. The operative sizing of the pulmonary arteries revealed a mildly hypoplastic LPA relative to the RPA. The anastomosis was performed with continuous 6.0 absorbable monofilament sutures.

Postoperative status. The child was extubated in the operating room. He remained less than 24 hours in the intensive care unit and was discharged from the hospital on the sixth day.

Follow-up. At 5 months, the child was perfectly well, with strictly normal results of clinical examination. According to chest radiography, the left lung remained a little bit smaller than the right one. The perfusion scan showed an increase in the left lung perfusion, with a right to left ratio of 71% to 29%. At 1 year, the echocardiographic estimate of the LPA diameter remained similar to the preoperative value, but the perfusion scan showed an improvement of the right to left perfusion ratio at 65% to 35% (Figure 2, B).

Discussion
In PAS, left lung perfusion can be dramatically decreased. First, the LPA is usually smaller than the RPA. Second, the course of

References