Hypoplastic Left Ventricle and Scimitar Syndrome

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Hypoplastic left ventricle with scimitar syndrome is a rare combination of anatomic lesions. Five patients with this anatomy have been followed up at our institution. Four of these patients are alive: 2 after heart transplant, 1 underwent a Norwood operation, and 1 has survived staged palliation, culminating in a Fontan operation.


Scimitar syndrome (SS) consists of partial anomalous pulmonary venous return to the inferior vena cava or inferior cavoatrial junction and other anomalies, including abnormal lung lobation, lung hypoplasia, dextrocardia, systemic arterial supply to the right lung, and rarely, atrial septal defects. SS has an incidence of 1 to 2 per 100,000 births [1]. Previous reports have classified patients, according to severity and timing of presentation, into infantile and child/adult forms. The infantile form has a mortality rate of 33% to 60% [2–4]. The combination with hypoplastic left ventricle (HLV) is rare, and the incidence of this combination is unknown, with only 3 patients reported previously [5–7]. We detail our experience with 5 patients with HLV-SS at The Children’s Hospital of Philadelphia.

Case Reports

Patients were identified by query of our institution’s cardiac surgery and cardiology databases. Each medical record was reviewed and is presented in chronologic order. The study was exempted from review by the Institutional Review Board of The Children’s Hospital of Philadelphia.

Patient 1

This was a 3.0-kg boy with double-outlet right ventricle, HLV, bilateral superior vena cavae, and SS. The anomalous pulmonary vein drained into the inferior vena cava, and he had severe right pulmonary artery (RPA) hypoplasia (diameter, 1.7 mm; z score, –6.5) and a large aortopulmonary collateral (APC) supplying the right lung. He underwent a Norwood operation with a 5-mm Sano shunt (scimitar vein and APC were not addressed). An attempt to perform bilateral superior cavopulmonary connections (SCPC) at age 11 months was abandoned due to the severity of RPA hypoplasia. He underwent patch angioplasty of the left pulmonary artery (LPA) and upsizing of the Sano shunt. He was discharged home with a tracheostomy and chronic ventilatory support. The scimitar vein was patent and unobstructed at discharge. He died at home after a respiratory event at age 3 years.

Patient 2

A 5-year-old girl had a history of hypoplastic left heart syndrome (HLHS) and SS. She initially underwent a Norwood operation with a 5-mm Sano shunt and ligation of the APC, without addressing the scimitar vein. At age 6 months, she had a right hemi-Fontan and left bidirectional Glenn with stenting of her proximal RPA. An atrial septectomy was performed at 2 years. She presented to our institution at 5 years for a second opinion regarding Fontan operation in light of a relatively high pulmonary vascular resistance (PVR) of 3.7 Woods units (WU). Angiography demonstrated unobstructed anomalous venous drainage to the right atrium with mild RPA (diameter, 8.6 mm; z score, –0.4), and lung hypoplasia. She underwent an extracardiac nonfenestrated Fontan. She was last seen at age 8 and was doing well, with saturations in high 90s without any further intervention.

Patient 3

This was a 3.5-kg girl with HLHS and SS with severe RPA hypoplasia (diameter, 3.3 mm; z score, –2.2). Catheterization demonstrated obstructed anomalous pulmonary venous return from the entire right lung to the cavoatrial junction and a large APC supplying the right lung. She underwent a hybrid procedure with bilateral PA bands and a ductal stent. Worsening heart failure developed and at catheterization required an interatrial stent and ductal stent angioplasty. At age 5 months, she underwent a biatrial heart transplant. The atrial septum was left intact with the scimitar vein draining into the right atrium. Initial hemodynamic instability required extracorporeal membrane oxygenation support before she was successfully weaned. She was discharged home with a tracheostomy for chronic respiratory failure but has since been decannulated. Catheterization at age 5 years demonstrated a mean RPA and LPA pressures of 17 and 22 mm Hg, respectively, with a PVR of 1.6 WU. She continues to do well 6 years after transplant.

Patient 4

This was a 2.4-kg girl with HLHS, dextrocardia, bilateral superior vena cavae, and SS. The scimitar vein drained the entire right lung to the right atrium, with severe RPA and lung hypoplasia (diameter, 2.5 mm; z score, –3.4).
She had a right-sided pulmonary sequestration with an APC arising from the celiac trunk. She underwent a hybrid procedure with bilateral PA bands and an interatrial stent, but the ductal stent was not attempted for concern of reverse coarctation. She was maintained on intravenous prostaglandin and at age 5 months underwent a bicaval transplant with reimplantation of the scimitar vein into the left atrium. The sequestration was not addressed at this operation. Her postoperative course was notable for pulmonary hypertension and poor ventricular function. Catheterization demonstrated no antegrade flow to the right lung, with occlusion of the reimplanted scimitar vein. She was discharged home with a tracheostomy for chronic respiratory failure but continues to do well. Her most recent catheterization, at age 22 months, demonstrated a mean LPA pressure of 25 mm Hg and a PVR of 2.7 WU.

**Patient 5**

A 2.6-kg boy with HLHS, right lung hypoplasia, and left superior vena cava to coronary sinus underwent a Norwood operation with a 3.5-mm right modified Blalock-Taussig shunt. A postoperative catheterization revealed undiagnosed SS. The anomalous vein from the right middle and lower lobes was obstructed at the cavoatrial junction. He had a moderate-sized APC to the right lung and mild RPA hypoplasia (diameter, 4.0 mm; z score, –0.5). At 5 months, he underwent balloon angioplasty and stenting of the scimitar vein and coiling of the APC. He was unsuitable for stage 2 palliation due to elevated PVR and decreased pulmonary blood flow and was listed for transplant. At 8 months, progressive hypoxemia prompted a catheterization and stenting of the proximal Blalock-Taussig shunt and balloon angioplasty of an in-stent stenosis of the scimitar vein. At his most recent catheterization, at age 9 months, his mean LPA and RPA pressures were 14 and 15 mm Hg, with a PVR of 3.5 WU.

**Comment**

Our experience with HLV-SS represents the largest in the literature, highlighting the low prevalence of this anatomic combination. Three patients with HLV-SS have been reported: 1 required heart transplantation after failed palliation [6], and 2 successfully underwent SCPC [5, 7]. No patients surviving to Fontan completion have been reported.

Our experience with HLV-SS raises several interesting points. First, the degree of right lung and pulmonary artery hypoplasia may help guide initial operative therapy. For patient 1, severe lung and RPA hypoplasia made SCPC prohibitive. The patients with severe RPA hypoplasia (z score < –2) and right lung hypoplasia (Fig 1B) have died or staged palliation failed. The only patient to complete staged palliation had an RPA diameter z score of more than –2. The other patient with an RPA z score exceeding –2 (Fig 1B) had obstructed anomalous venous drainage, and his elevated pulmonary vascular resistance made SCPC unfeasible. Despite low numbers, our experience suggests that patients with an RPA z score of less than –2 may benefit from a hybrid procedure or early listing for transplant rather than attempted staged palliation.

Second, the hybrid approach may allow for stabilization without exposure to cardiopulmonary bypass and homograft. Patient 1 presented before the hybrid procedure was performed at our center, but given his prohibitively hypoplastic RPA, the hybrid approach would have allowed us to stabilize the patient and reassess staged palliation vs early transplantation.

Third, appropriate operative treatment of the scimitar vein remains challenging. In 4 of 5 patients, the scimitar vein was not addressed, leaving a small but persistent volume load to the right heart. The scimitar vein in patient 4 was anastomosed directly to the left atrium, which subsequently thrombosed, resulting in a nonperfused lung. The patient ultimately required tracheostomy and chronic mechanical ventilation. In cases of SS with severe lung and RPA hypoplasia, because most of the pulmonary blood flows to the left lung, it may be prudent to leave the scimitar vein alone. The residual left-to-right shunt is typically of little hemodynamic or clinical significance.

In summary, HLV-SS represents a challenging combination of lesions. Infants with severe RPA hypoplasia (z score < –2) may be best served by a hybrid approach,
with less initial perioperative risk than a Norwood operation, providing time to assess whether transplantation or staged palliation is preferable.

References

Off-Pump Total Arterial Revascularization for Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery

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Anomalous origin of the left coronary artery from the pulmonary artery is a rare but potentially lethal congenital anomaly, especially if presented later in adulthood. We report a previously healthy young man who had a sudden cardiac arrest at a Zumba class, was resuscitated, and was successfully treated with off-pump total arterial revascularization and ligation of the left main stem of anomalous origin.


Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), or Bland-White-Garland syndrome, is a developmental anomaly caused by an abnormal connection of the left coronary bud to the pulmonary artery rather than normal communication to the aorta. Myocardial ischemia occurs soon after birth when antegrade left coronary blood flow from the low-pressure pulmonary artery is decreased as a result of a physiologic decrease in pulmonary vascular resistance. Coronary perfusion is dependent on the right coronary artery by way of collaterals, with retrograde flow through the left coronary shunting into the pulmonary artery, resulting in a “coronary steal” phenomenon [1]. Most patients with this condition present soon after birth. Few survive to adulthood and present with left ventricular dysfunction, mitral regurgitation, or sudden death. Surgical correction is the treatment of choice, although medical management is reserved for high-risk inoperable patients [2, 3]. We describe a case of ALCAPA treated with off-pump coronary artery bypass grafting (OPCABG) total arterial revascularization, with discussion of various surgical techniques used.

A 30-year-old man, generally well, suddenly collapsed at a Zumba fitness class. Basic life support was commenced immediately by a bystander. Upon arrival of the ambulance crew, the patient was noted to be in ventricular fibrillation arrest. He was successfully resuscitated with direct current cardioversion and amiodarone. An electrocardiogram showed ST depression with Q wave inversion in lateral leads. He was transferred to our primary angioplasty center for intervention. Intubation of the left main stem (LMS) was unsuccessful, and injection into the right coronary artery filled up the left coronary system and showed the origin of the LMS from the main pulmonary artery, as is typical of ALCAPA (Fig 1A). The patient was stabilized with an intraaortic balloon pump and inotropic agents. He also received therapeutic hypothermia in the intensive care unit for 3 days. Eventually he recovered from the initial insult and was transferred to a ward. An echocardiogram showed a dilated left ventricular cavity and mild overall systolic impairment, anterior and posterolateral wall hypokinesia, and multiple color flow jets associated with the septum, consistent with aberrant coronary arterial flow (Fig 1B). There was mild to moderate mitral regurgitation. He had normal carotids and vertebral arteries. Cardiac computed tomographic angiography (Fig 1C) confirmed the diagnosis. Cardiac magnetic resonance imaging showed subendocardial fibrosis and remodeling, as is usually seen in chronic ischemia, in the territory of the left coronary system (Fig 1D). In consideration of his age, his fitness, and these findings, the patient was worked up for surgical repair with OPCABG and total arterial revascularization.

Surgical Technique

The patient underwent a median sternotomy. The coronary arteries were hugely dilated and tortuous. The LMS originated from the posterior aspect of the main pulmonary trunk; no other anomalies were found. Bilateral pedicled internal mammary arteries were obtained. A half dose of heparin was given. The right internal mammary was anastamosed to the left anterior descending artery and the left to the first obtuse marginal (Fig 2). The origin...