Severe pulmonary hypertension is associated with a poor prognosis [1]. Chronic suprasystemic pulmonary hypertension and right ventricular pressure overload usually result in right ventricular failure [2]. A decrease of cardiac output is not only related to impaired right ventricular function but also could be a consequence of impaired left ventricular filling due to right-to-left ventricular interaction [3] or reduced left ventricular function, ie, due to a borderline left ventricular morphologic condition [4]. As a novel approach for palliating children with suprasystemic idiopathic pulmonary arterial hypertension (IPAH), creation of a Potts shunt has been reported, which could be an alternative treatment to lung transplantation in selected cases [4, 5]. This surgical procedure implies the construction of an anastomosis between the left pulmonary artery and the descending aorta that allows right-to-left shunting and thereby leads to decompression of the failing right ventricle (RV) without provoking desaturation in the upper part of the body.

We describe our experience with a modified Potts shunt in a 20-year-old young adult with suprasystemic postcapillary pulmonary hypertension caused by Shone’s complex, including parachute mitral valve, aortic valve stenosis, small muscle ventricular septal defect, persistent ductus arteriosus, and a LV of borderline size. The patient had successful balloon valvuloplasty of the stenotic aortic valve at the age of 1 month and an interventionial closure of the arterial duct at the age of 2 years. At the age of 20 years he was admitted to our center with decompensated biventricular failure corresponding to the age of 20 years. He was already listed for combined heart-lung transplantation. Horizontal positioning became impossible and resulted in acute congestive heart failure associated with pulmonary edema and significant cyanosis despite continuous supplemental oxygen therapy and pulmonary arterial hypertension (PAH)-specific medication that included a combination of bosentan and sildenafil.

Cardiac imaging by echocardiography/cardiac magnetic resonance imaging showed a small LV with impaired function (indexed end-diastolic left ventricular volume, 77 mL/m² body surface area; mitral valve annulus, 31 mm; ejection fraction, 34%), an enlarged left atrium, and dilated pulmonary veins. The apex-forming RV and the pulmonary arteries were markedly dilated, the right ventricular myocardium was hypertrophied, and right ventricular function was significantly impaired (ejection fraction, 16%) (Figs 1, 2). Right and left heart catheterization performed in the sitting position revealed low cardiac output (cardiac index, 2.2 L/min/m²; Qp/Qs [ratio of pulmonary blood flow to systemic blood flow], 1.0) associated with severe left ventricular diastolic dysfunction (left ventricular end-diastolic pressure, 28 mm Hg) and “suprasystemic” postcapillary pulmonary hypertension (pulmonary arterial pressure, 125/75 mm Hg; systemic arterial pressure 105/55 mm Hg). Nitric oxide administration...
led to an increase of the pulmonary capillary wedge pressure from 32 mm Hg to 41 mm Hg. Pressure gradients across the aortic (7 mm Hg) and mitral valve (2 mm Hg) were measured to exclude valvular heart disease as a confounding factor contributing to the rise in pulmonary artery pressure.

Based on our experience with hybrid interventions in newborns with borderline LV and recent reports of the use of a Potts shunt in IPAH [4–7], we decided our patient should undergo an interventional/surgical hybrid approach consisting of the creation of an interatrial communication combined with a modified Potts shunt. Using the Brockenbrough technique and gradual balloon dilation, an 8- to 10-mm atrial septal defect was created with a significant left-to-right shunt associated with a drop of the left atrial pressure from 30 to 22 mm Hg (Fig 3A). The size of the atrial septal defect chosen was large enough to allow partial decompression of the LV but was small enough to avoid unfavorable volume load of the RV. A surgical graft tube (13-mm polytetrafluoroethylene) was then implanted between the left pulmonary artery and the descending aorta through a lateral thoracotomy (Fig 3B). The size of the graft tube was chosen to be at least the same as the diameter of the descending aorta.

The perioperative period remained uneventful. A “harlequin”-like oxygen saturation pattern with desaturation of the lower limb was caused by the pulmonary artery-to-aorta right-to-left shunt (arterial oxygen saturation of the ascending aorta was 97% and that of the descending aorta was 78%–84%) combined with a decrease of the pulmonary arterial pressure from 125/75 to 115/59 mm Hg in the presence of unchanged systemic arterial pressure. The postoperative Qp/Qs was 0.65, whereas the indexed pulmonary circulation was 3.0 L/min/m² and the systemic circulation was 4.6 L/min/m². Left ventricular end-diastolic pressure decreased markedly from 28 to 15 mm Hg, associated with a right ventricular end-diastolic pressure of 12 mm Hg and an increase of right ventricular ejection fraction from 16% to 27%. Left ventricular function improved (ejection fraction, 55%) and the shape of the LV became more spherical because of decompression of the RV. Recovery of ventricular function was further expressed by a decrease in levels of brain natriuretic peptide from 340 pg/mL at presentation to 135 pg/mL at discharge from the hospital. World Health Organization functional class decreased from class IV to class III at hospital discharge and 3 months later to func-

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**Abbreviations and Acronyms**

- DAO = descending aorta
- IPAH = idiopathic pulmonary arterial hypertension
- LPA = left pulmonary artery
- LV = left ventricle
- PAH = pulmonary arterial hypertension
- RV = right ventricle

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**Fig 1.** (A) Echocardiograms showing the right ventricular–left ventricular imbalance with a small left ventricle (LV) and (B) the dilatated and hypertrophied apex-forming right ventricle (RV).

**Fig 2.** Cardiac magnetic resonance image showing the small left ventricle (LV) with a dilatated left atrium and enlarged pulmonary veins (asterisks). The right ventricle (RV) is dilatated and severely hypertrophied.
tional class II, whereas the distance for a 6-minute walk improved from 75 meters to 375 meters.

Comment

Based on the poor prognosis of children with pulmonary hypertension [1, 8], different rescue treatments have been proposed in patients with severe PAH and right ventricular failure. Atrial septostomy is currently recommended for bridging these patients to lung transplantation [8]. Recently, surgical anastomosis between the left pulmonary artery and the descending aorta has been described in patients with end-stage IPAH with suprasystemic pulmonary artery pressures who are refractory to medical treatment [4, 5]. This pulmonary-to-systemic connection changes cardiac pathophysiologic features to those of Eisenmenger’s physiology.

To our knowledge, we report the first patient with end-stage biventricular failure with out-of-proportion postcapillary PAH caused by Shone’s complex with associated borderline left ventricular morphologic features, in whom an interventional/surgical hybrid approach consisting of balloon atrial septostomy combined with a Potts shunt was successfully performed. Suprasystemic pulmonary arterial pressure decreased to the systemic level combined with an increase of the systemic blood flow. Left ventricular preload decreased because of the atrial left-to-right shunt and the reduced transpulmonary blood flow through the Potts shunt. The change of the cardiac pathophysiologic characteristics into those of Eisenmenger’s physiology preserved highly oxygenated coronary and cerebral blood flow and avoided extreme oxygen desaturation of the lower body parts by the atrial left-to-right shunt. However a prerequisite for the hybrid operation presented, which includes both atrial and pulmonary arterial level shunts as opposed to a pulmonary arterial shunt alone as reported by Blanc and colleagues [4], is that postoperative left ventricular end-diastolic pressure has to be higher than right ventricular pressure such that the atrial level shunt is left to right, avoiding complete systemic desaturation (eg, upper and lower body), which may not be tolerated. As such, it may not be advised in other conditions associated with severe pulmonary hypertension in which this criterion is not met.

References


Solitary Fibrous Tumor Causing Cardiac Tamponade

Akinori Tamenishi, MD, Yasumoto Matsumura, MD, and Hiroshi Okamoto, MD

Department of Cardiovascular Surgery, Yokkaichi Municipal Hospital, Yokkaichi, Japan

Solitary fibrous tumor of the pleura is a rare primary tumor arising from mesenchymal cells in the areolar tissue subjacent to the mesothelial-lined pleura. Most solitary fibrous tumor of the pleura arises from the visceral or the parietal pleura, and asymmetrically occupies the hemithoracic cavity. We report a rare case of solitary fibrous tumor of the pleura causing cardiac tamponade.

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Address correspondence to Dr Tamenishi, Department of Cardiovascular Surgery, Yokkaichi Municipal Hospital, 2-37 Shibata 2-chome, Yokkaichi 510-8567, Japan; e-mail: tamenishi89@yokkaichihp01.jp.