Pulmonary artery in patients with severe suprasystemic PAH.

or probe-patent ductus arteriosus (PDA) to establish a stable approach using a transcatheter technique by stenting residual with direct side-to-side anastomosis.2–4 We describe a similar story IPAH into an Eisenmenger physiology using a Potts shunt reported improved survival after converting severe and refrac-
tively better than patients with IPAH. Prior surgical series have Eisenmenger syndrome where the life expectancy is signifi-
cantly better than patients with IPAH. During the past 5 years, we have performed cardiac catheterization in 28 IPAH patients and found 4 with a small or probe-patent PDA. One of these patients had infrasystemic PA pressures and did not undergo PDA stenting. The other 3 patients, whose data are summarized in the Table, had the PDA stented.

Patient 1 was diagnosed with idiopathic PAH at the age of 6 months. Transthoracic echocardiography at the time of the diagnosis showed an atrial septal defect with left-to-right shunt and a small PDA. He remained stable on oral medica-
tions until 9.7 years of age when he presented with recurrent syncope. Severe right ventricular dilatation and bowing of the interventricular septum to the left were noted on transthoracic echocardiogram. Cardiac catheterization confirmed suprasys-
temic PAH and the presence of a small PDA shunting right-to-
left with significant restriction (Figure 1).

Patient 2 was diagnosed to have suprasystemic PAH at the age of 2.6 years. He was started on oral medication with no significant improvement. Echocardiography showed partial atrioventricular septal defect with a small ostium primum.

atrial septal defect and a small PDA. Cardiac catheterization at the age of 3.3 years confirmed suprasystemic PAH. A small PDA was seen only on the pulmonary angiogram with exclusive right-to-left shunt.

Patient 3 was diagnosed with familial PAH at the age of 5 months. PA pressure was noted to be suprasystemic by echo-
cardiogram with poor response to triple therapy at 16 months of age. Cardiac catheterization at 2.3 years showed suprasys-
temic pressures with right ventricular failure. Angiography diagnosed the presence of an incidental small PDA (Movie I in the online-only Data Supplement).

**PDA Stenting**

A written, valid, and witnessed consent was obtained from the parents before the procedure. All procedures were done under sedation. The PDA was crossed with a wire from the PA to the aorta except in patient 3 and was stented with a bare metal stent large enough to equalize the systolic PA and aortic pressures in a stepwise and sequential manner. Postdilatation of the stent was done in patient 1 (10 mm Conquest balloon, Bard, USA). Patient 3 required deployment of a second stent, as angiogram after the first stent showed a tight angulation between the stent and the roof of the main PA. The second stent was placed to avoid possible PA rupture by the end of the first stent. Pulse oximetry saturation in the lower limbs dropped after stenting in all patients. Patient 3 had vagally mediated vascular collapse just after crossing the PDA with guide wire, requiring brief cardiac resuscitation. All other procedures were done without major complications. There was no evidence of stent collapse, tear, extravasation, or dissection of the PA or the aorta in any patient. Post–procedure transthoracic echocardiography showed a widely patent stent with unrestricted flow across the stented PDA in all patients. The right ventricular function improved immediately after the procedure. All patients were discharged on the next day.

**Diagnosis**

Idiopathic pulmonary arterial hypertension (IPAH) continues to be a progressive and fatal disease.¹ Patients with congeni-
tal heart disease and PAH constitute a separate subset with Eisenmenger syndrome where the life expectancy is signifi-
cantly better than patients with IPAH. Prior surgical series have reported improved survival after converting severe and refrac-
tory IPAH into an Eisenmenger physiology using a Potts shunt with direct side-to-side anastomosis.² ³ ⁴ We describe a similar approach using a transcatheter technique by stenting residual or probe-patent ductus arteriosus (PDA) to establish a stable communication between descending thoracic aorta and left pulmonary artery in patients with severe suprasystemic PAH.

**Patents**

During the past 5 years, we have performed cardiac cathe-
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Follow-up

After a mean follow-up of 14±9 months (range, 8–24 months), all patients showed improved functional capacity and patent PDA stent on echocardiography with a mean value of peak velocity across PDA of 1.4±0.17 m/s (Figure 2; Table). On echocardiography, right ventricular function improved in all patients (Table). Patient 3, however, succumbed to a lethal RSV infection after 24 months.

Discussion

The poor overall response to medical management for patients with chronic IPAH has compelled the need to find alternate ways to palliate such patients. Lung or heart–lung transplantation, balloon atrial septostomy or surgical Potts shunt are some limited alternatives for patients who worsen while receiving medical therapy for IPAH or who experience repeated syncope or right heart failure.2–4 The rationale of creating a right-to-left shunt at the arterial level is to switch IPAH physiology to more favorable Eisenmenger physiology. By doing so, cyanosis and the risk of paradoxical embolism are limited to the lower half of the body, and the growing brain is thus spared from the chronic ill effects of hypoxia.

We present 3 medically managed cases with suprasystemic PAH in whom we successfully stented a small existing PDA. We chose stent diameter based on our experience of surgical Potts shunt where the mean opening diameter was 9.25 mm (target diameter). However, in contrast to surgical Potts shunt where the shape of the opening is elliptical or slit-like, transcatheter Potts opening is more circular, providing a larger cross-sectional area. Therefore, rules for selecting stent diameter may not apply in a similar manner. The criteria we used for selecting initial stent diameter were an aggregate consideration between age, surgical target diameter, and narrowest PDA diameter. Most importantly, we chose to gradually enlarge the diameter of the connection and thus flow by sequential balloon inflation with the aim of equalizing the PA and aortic systolic pressures. By doing so, we were also able to maintain pulmonary blood flow and limit lower limb desaturation. A 6-mm stent was used in the smallest patient and 9-mm stent for the oldest patient. Postdilatation and over-expansion were necessary in 1 patient. The objectives were achieved with a single stent in 2 patients.

There was a dramatic improvement in the functional status immediately after PDA stenting, and the improvement was sustained through last follow-up. The significant improvement without midterm complications from this interventional approach should allow larger series with longer follow-ups. In comparison with the most recent surgical series, which had 8 patients >5 years from 2 high volume institutions,4 we report our interventional approach in 3 patients from a single center >5 years. The promising results compelled us to change the approach and protocol for management of patients with suprasystemic PAH. We now perform a detailed cardiac catheterization in all patients with suprasystemic PAH to look for a PDA.

Conclusions

Palliative transcatheter Potts shunt established by probing and stenting restrictive PDA constitutes an innovative approach for severely ill patients with suprasystemic PAH.
Further long-term multicenter studies will be required to evaluate its impact on survival.

Disclosures

None.

References


Key Words: congenital heart defect • patent ductus arteriosus • pulmonary hypertension • stent
Patent Ductus Arteriosus Stenting (Transcatheter Potts Shunt) for Palliation of Suprasystemic Pulmonary Arterial Hypertension: A Case Series
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Movie 1. Cineangiogram from patient 3 showing the presence of a clinically silent and incidentally diagnosed small PDA. The PDA was crossed with a wire from the aorta. A pre-mounted Genesis stent (6*18mm, Cordis, Fr) was positioned in the PDA but there was a tight angulation between the stent and the roof of the main PA. Therefore, a second Genesis stent (7*18mm, Cordis, Fr) was inserted. Note the smooth curve and the wide opening of the PDA.