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Single center experience with the Potts shunt in severe pulmonary arterial hypertension☆

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A B S T R A C T

Background: There remains limited options for end stage pulmonary arterial hypertension that is refractory to medical therapy. The reprisal of the Potts shunt (pulmonary artery to descending aorta anastomosis) has been used to decrease right ventricle (RV) afterload by creating a right to left shunt in effort to promote lung transplant free survival.

Methods: Retrospective review of three patients ages 16, 19, and 27 years old who underwent the creation of a "reverse" Potts shunt at our institution in 2016.

Results: The patients were WHO functional class III or IV and on three classes of pulmonary hypertension specific drug therapy including IV prostacyclin. All patients had RV dysfunction and suprasystemic RV pressures. Two patients had a catheterization for dilation and stenting of a tiny patent ductus arteriosus (PDA) and one had emergent surgical placement of a pulmonary artery to descending aorta conduit. The catheterization procedures were well tolerated without complications. The surgical procedure was complicated with heavy bleeding and respiratory failure. All patients recovered from their procedure and experienced improved functional class with decrease in RV pressures to systemic levels but no improvement in RV function. One catheterization patient required restenting due to stent fracture at 7 months. Our surgical patient died from massive hemoptysis 13 months after the procedure.

Conclusions: Reverse Potts shunt physiology may be an option for end-stage PAH patients with suprasystemic RV pressures. Optimal timing of this procedure remains unclear, but if feasible, PDA stenting, even in adult patients may be accomplished with low morbidity.

1. Introduction

The progressive and fatal nature of pulmonary arterial hypertension (PAH) has been the impetus for the creation of better PAH therapies. Fortunately, these newer therapies are likely responsible for the improved outcomes in PAH over the past few decades. As noted by the REVEAL registry, one and five year adult survival is now 85% and 57% compared to 68% and 36% reported by the NIH in the 1980's [1,2]. Similarly, pediatric survival has improved with five year survival of 74% in the REVEAL registry [3] compared with a historical average pediatric survival less than one year post diagnosis [2].

Current pediatric and adult treatment guidelines for PAH describe a tiered approach to medical therapy with escalation to combination drug therapy for inadequate clinic response and finally lung transplant for medically refractory disease [4,5]. The use of an atrial septostomy is the only interventional procedure described in the adult guidelines for medically refractory PAH. In contrast, the Potts shunt has just recently been noted as an additional therapy in pediatrics to consider before transplantation in the European guidelines [5].

Blanc et al. first described the use of a "reverse" Potts shunt (descending aorta to left pulmonary artery anastomosis) in two children to create a right to left shunt to decrease afterload of the right ventricle (RV) while preserving saturations to the upper part of the body in patients with suprasystemic PAH [6]. Subsequently, several centers have reported series of patients with medically refractory PAH receiving Potts shunt with improvement in RV function and overall functional status [7,8]. We report our single center experience with creating reverse Potts shunt physiology in three adolescent/young adult patients with medically refractory PAH.

☆ Declarations of interest: none.

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Table 1
Pre-procedure characteristics.

Patient #	Age at PAH diagnosis	Age at Potts shunt	Diagnosis	WHO FC	Oral Med	IV PC dose	NT-PROBNP	O2	6MWT
1	10 years	16 years	IPAH	III	ERA, PDE5I	316 ng/kg/min‡	74 (BNP)	1-2 L	330 m
2	8 years	19 years	HPAH	IV	ERA, PDE5I*	210 ng/kg/min‡	9000	None	–
3	11 years	27 years	IPAH	III	ERA, PDE5I	102 ng/kg/min§	3340	4-5 L	441 m

ERA = Endothelin receptor antagonist. PDE5I - phosphodiesterase-5 inhibitor. IPAH = idiopathic pulmonary arterial hypertension. HPAH = Hereditary PAH. WHO FC = world health organization functional class. PC = prostacyclin. O2 amount in liters. *Resumed 2 weeks prior. ‡ treprostinil, § epoprostenol.

2. Methods

This is a retrospective review of patients who underwent creation of reverse Potts shunt physiology at the Children's Hospital of Wisconsin in 2016. Electronic medical record was reviewed to extract demographic and clinical data obtained during routine follow up at the Pulmonary Hypertension clinic at the Herma Heart Institute. When available, pre-procedural cardiac catheterization and echocardiogram reports were reviewed. Patient functional class was classified using the World Health Organization (WHO) functional class scale.

3. Results

3.1. Patient Characteristics

Patient characteristics are presented in Table 1. All patients were on three therapies for PAH which included IV prostacyclin, oral phosphodiesterase-5 inhibitor and an endothelin receptor antagonist. Patient #1 was a 16-year-old female with idiopathic PAH diagnosed at age 10 years old who had a decrease her WHO functional class from II to III over the year prior to elective patent ductus arteriosus (PDA) stent placement. She and her family refused referral for lung transplant.

Patient #2 was a 19-year-old male with hereditary PAH due to a BMPR2 mutation diagnosed at age 8 years old who had been lost to follow up and was non-compliant with medical therapy. Upon re-presentation, he was hospitalized with RV failure and WHO functional class IV. Despite attempts to reinstitute and increase pulmonary vasodilator therapy, he had worsening RV ischemia as evidenced by ST segment elevation and increase in cardiac troponins. As he was not felt to be a good candidate for catheter intervention to create an atrial level shunt given that the atrial septum bowed far into the left atrial space, he underwent emergent placement of a surgical Potts shunt. Due to the urgency of his clinical situation and history of non-compliance he was not considered a lung transplant candidate at this time.

Patient #3 was a 27-year-old female with idiopathic PAH diagnosed at age 11 years old who had been declined as a lung transplant candidate secondary to non-cardiac comorbidities. She was stable with WHO functional class III symptoms at the time of elective PDA stent placement.

3.2. Pre-Procedural Data

Pre-procedure echocardiograms revealed suprasystemic RV pressure (estimated by TR gradient + central venous pressure) in all three patients. Patient #1 had mildly decreased, #2 moderately decreased and #3 severely decreased RV function (Table 2). Prior to their procedures, NT Pro BNP was significantly elevated in patients #2 and 3.

Patients #1 and 3 both had a tiny PDA noted on chest computerized tomography (CT) not well visualized on echocardiogram. Cardiac catheterization prior to PDA stent placement in patients #1 and 3 confirmed suprasystemic RV pressures (Table 3) and a PDA. Patient #2 had a catheterization 4 years prior showing subsystemic RV pressures but repeat catheterization immediately preceding surgical Potts shunt was not performed due to hemodynamic instability.

3.3. Procedures

Patients #1 and 3 underwent stenting of their PDA using a single pre-mounted baremetal EV3 Visipro stent initially implanted at 7 mm in diameter. A retrograde ductal approach was utilized, advancing a long sheath into the aorta via the pulmonary artery and using contrast injections from the sheath and aortic catheter to guide placement. The length was determined by angiography and prior CT imaging. The stents were further inflated with the goal of reducing the main PA pressure down to the aortic pressure, although we chose to stop at 9 and 10 mm diameter for patients #1 and 3, respectively, given the tiny initial PDA size (1–2 mm) and thus did not immediately reach the goal pressures with the initial implantation.

Patient #2 underwent placement of a surgical Potts shunt performed via a left thoracotomy. Because of significant collateral vessel burden and suprasystemic pulmonary artery pressures with a thin walled main pulmonary artery (MPA), cardiopulmonary bypass was utilized to decompress the MPA to allow safe clamping and placement of a 10 mm diameter Dacron conduit between the descending aorta (DAO) and MPA. The procedure was complicated by bleeding from the MPA anastomosis which required extensive felt reinforcement and blood product transfusion, but was ultimately successful.

Warfarin to a goal INR of 2–2.5 plus aspirin was used for anticoagulation with the PDA stents and aspirin alone was used for the surgical Potts shunt.

3.4. Outcomes

Both PDA stent patients (patients #1 and 3) tolerated the procedure well without complications. Immediately after PDA stenting while in the catheterization lab, there was a difference between the pre and post shunt saturations of 9–13% by pulse oximetry, however pulmonary artery pressures still measured suprasystemic. Both PDA stent patients were discharged home after 2 days on decreased doses of IV prostacyclin.

The surgical Potts shunt in patient #2 was complicated by significant hemorrhage from the surgical site and a pulmonary contusion with respiratory failure requiring ventilator support for 19 days. Though early on he needed inotropes, evidence of previous myocardial ischemia immediately resolved post-procedure. He had wound dehiscence from the thoracotomy requiring a VAC dressing. He was able to wean IV prostacyclin while hospitalized and the patient eventually recovered to discharge 51 days post shunt. His thoracotomy site was surgically closed a month after discharge.

Early echocardiograms around the time of discharge did show a decrease in estimated RV pressures to systemic levels with right to left flow across the shunt in all three patients.

3.5. Latest Follow-Up

All patients have been followed closely and subjectively report feeling better overall and improvement in WHO functional class (Table 4) despite no significant improvement in RV function by echocardiogram (Table 2). All patients continue to have a pulse oximetry saturation difference between pre and post-shunt as well as a slight