Case Report

Pulmonary Circulation

Palliative balloon atrial septostomy in two pediatric patients with severe pulmonary arterial hypertension requiring extracorporeal membrane oxygenation support

Samantha L. Brackett, Nina Deutsch and Chinwe Unegbu

Division of Anesthesiology, Pain and Perioperative Medicine, Children's National Hospital, Washington, DC, USA

Abstract

Pulmonary arterial hypertension is a pernicious disease with a diverse etiology in the pediatric population. Despite the increased availability of drug therapies, pulmonary arterial hypertension continues to cause significant morbidity and mortality. In pediatric patients with severe pulmonary arterial hypertension who have failed medical therapy, a few studies have demonstrated the role of balloon atrial septostomy as a bridge to lung transplantation or a means of improving symptomatology. However, no data exists on the utilization of balloon atrial septostomy as a palliative intervention to wean from extracorporeal membrane oxygenation (ECMO) when all other therapies are exhausted. Here we describe a case series of two pediatric patients with severe pulmonary arterial hypertension, requiring ECMO support, who were successfully weaned from ECMO following balloon atrial septostomy.

Keywords

extracorporeal circulation, pediatric cardiovascular disease, pulmonary arterial hypertension

Date received: 10 June 2020; accepted: 7 August 2020

Pulmonary Circulation 2020; 10(3) 1–3
DOI: 10.1177/2045894020953714

Pulmonary arterial hypertension (PAH) is associated with significant morbidity and mortality in the pediatric population with a median survival of 10 months in children left untreated.[1,2] PAH is defined as a resting mean pulmonary artery pressure of >25 mmHg with a pulmonary capillary wedge pressure (PCWP) of <15 mmHg, and a pulmonary vascular resistance (PVR) of >3 Wood units × M^2 in those older than three months of age.[1,2] The etiology of PAH can be diverse and may include cardiac, pulmonary, and systemic diseases. PAH may also be idiopathic, though this remains a diagnosis of exclusion.[1]

Despite novel pharmacologic therapies, children diagnosed with PAH continue to have poor long-term outcomes.[1] Those with severe PAH who fail maximal pharmacologic therapy may be referred for lung transplantation; however, the few who meet transplantation requirements may die waiting, making salvage therapies more critical for these patients.[3]

Balloon atrial septostomy (BAS) is a palliative intervention recommended (Class IIb, Level C evidence) in adult patients with PAH awaiting lung transplantation who have failed maximal medical therapy.[3] No recommendations exist on the use of BAS in pediatric patients with severe PAH. Moreover, no literature exists on its use as a palliative therapy to transition off of extracorporeal membrane oxygenation (ECMO) support. In this case series, we describe two pediatric cases of severe PAH necessitating institution of ECMO in which subsequent palliative BAS resulted in successful weaning off of ECMO.

Corresponding author:
Samantha L. Brackett, Division of Anesthesiology, Pain and Perioperative Medicine, Children's National Hospital, 111 Michigan Avenue NW, Washington, DC 20010, USA.
Email: sammi.brackett@gmail.com

Case descriptions

Case 1

A 2-year-old male patient presented to the emergency room with decreased exercise tolerance, intermittent cyanosis, syncopal episodes, abdominal fullness, and periorbital edema. Abdominal ultrasound revealed flow reversal in the inferior vena cava. Therefore, a transthoracic echo (TTE) was requested and demonstrated severe dilation of the right atrium and right ventricle (RV), severe tricuspid regurgitation (TR), severely decreased RV function, systemic RV pressures, normal left ventricle (LV) function, and an intact atrial septum. The patient was admitted to the cardiac intensive care unit (CICU) and initiated on sildenafil, milrinone, and treprostinil therapy.

On hospital day 4, he underwent general anesthesia for a cardiac catheterization and peripherally inserted central catheter insertion. Anesthetic induction was uneventful with preemptive initiation of dopamine, and he remained on treprostinil and milrinone throughout the procedure. His diagnostic cardiac catheterization (on FiO_2 1.0) was significant for a femoral artery pressure of 73/47 mmHg (mean 58 mmHg) and main pulmonary artery (MPA) pressure of 140/75 mmHg (mean 95 mmHg). Right PCWP was 12 mmHg, and PVR was 45 Wood units $\times$ M^2. The pulmonary to systemic output ratio (Qp:Qs) was 1:1. His pulmonary vascular bed was nonreactive to inhaled nitric oxide (iNO).

During transfer onto the CICU stretcher, the patient experienced a pulmonary hypertensive crisis with severe hypoxia and hypotension that resolved with sedation, paralysis, and the initiation of iNO. In the CICU approximately 1.5 h later, the patient developed profound bradycardia and hypotension requiring multiple epinephrine boluses and eventual venoarterial (VA) ECMO cannulation. The next day, he underwent an uneventful BAS and was decannulated from VA ECMO 24 h later. He was extubated on postoperative day (POD) 3 following his BAS and then discharged a week later to home on sildenafil, ambrisentan, and treprostinil. Of note, BAS was not considered during this patient's initial cardiac catheterization since he tolerated the diagnostic procedure reasonably well and the hemodynamic collapse was encountered in the postoperative period.

The patient returned nine months after BAS for an elective and uneventful Potts shunt from the left pulmonary artery to aorta. He is currently medically managed at home with improved exercise tolerance and good symptom management.

Case 2

A 14-year-old male patient, with a history of PAH secondary to a later diagnosis of pulmonary venoocclusive disease who was initially managed on an outpatient regimen of ambrisentan, milrinone, and treprostinil, presented with increased dyspnea. TTE demonstrated suprasystemic RV systolic pressures, moderately decreased RV systolic function, tricuspid annular plane systolic excursion (TAPSE) 0.75 cm (>3 standard deviations below mean for age), severe RV dilation, moderate TR, and normal LV function.

Shortly after admission to the CICU, the patient experienced a pulmonary hypertensive crisis with subsequent bradycardic arrest. He was intubated, received 13 min of cardiopulmonary resuscitation, and was cannulated onto VA ECMO. He underwent BAS on ECMO day 3 after unsuccessful attempts at weaning off ECMO. He was decannulated from ECMO 24 h later.

However, despite continued unrestricted flow through his atrial septum, he became hypotensive and non-responsive to escalating doses of vasoactive agents on POD 6 and was recannulated onto VA ECMO. In order to further offload RV pressure and provide more permanent stability, on POD 10 he underwent a Potts shunt via thoracotomy and was decannulated from ECMO in the operating room immediately after the procedure. He was extubated four days after his Potts shunt and was discharged home three weeks later on treprostinil and milrinone. Of note, in patients with pulmonary venoocclusive disease, treprostinil should be administered cautiously given the potential risk of pulmonary edema. Since the diagnosis of venoocclusive disease was made one month after initiation of treprostinil therapy, via open lung biopsy, it is possible that treprostinil therapy worsened this patient's clinical status on initial presentation. However, the decision was made not to wean the patient off of this medication.

This patient is currently undergoing workup for lung transplantation candidacy, which should be performed early in these patients.

Discussion

There is limited data on the success of BAS in pediatric patients with severe PAH, and no literature exists describing its use as a palliative option to wean from ECMO support when all other therapy has been exhausted.[4,5] In both cases presented, ECMO was discontinued within 24 h of BAS.

Current literature describes BAS as a "bridging" therapy for lung transplantation or a palliative therapy to decrease symptomatology and increase short-term survival due to favorable hemodynamic changes.[3–7] BAS does not appear to have ever been performed as a salvage therapy to allow weaning and decannulation from ECMO.

As demonstrated in this novel case series, BAS may prove beneficial as a palliative measure to wean from ECMO support, particularly when all other options are exhausted. In both cases presented, BAS was a minimally invasive therapy that enabled decannulation from ECMO and provided an eventual bridge to a Potts shunt. However, as evidenced in case 2, there may be limitations in the size and stability of the atrial septal defect (ASD) created by a BAS, especially in older patients. At this time, no data exists to guide sizing of the ASD created. As a result, in older and larger patients, a

Potts shunt may be needed sooner. Current literature demonstrates that the Potts shunt improves hemodynamics, functional status, and transplant-free survival in children with severe PAH.[8] Further studies are needed in this patient population to determine outcome measures.

Acknowledgments

The authors would like to thank the Divisions of Anesthesiology, Cardiology, and Cardiac Surgery at Children's National Hospital.

Conflict of interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Author Contributions and Consent

All listed authors have contributed to and approve the manuscript. All listed authors consent to publication.

Ethical approval

Not applicable. IRB at Children's National does not require parental consent for this case report/case series.

Guarantor

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