

Management of Acute Right Ventricular Dysfunction Following Patent Ductus Arteriosus Ligation & Coarctation of the Aorta Repair on Left Heart Bypass

Amy Sirizi SAA, Ellen Richter MD, Greg Jarvis CAA

Emory University School of Medicine, Master of Medical Science in Anesthesiology, Atlanta, GA, USA

Introduction

Patent Ductus Arteriosus (PDA) represents 5-10% of all congenital heart defects and can be associated with coarctation of the aorta (CoA). A PDA is failure of a shunt between the pulmonary artery to aorta, distal to the left subclavian artery, to close after birth¹. Failure to close leads to excessive pulmonary flow. The primary diagnosis is rare in adults as this finding is usually found in infancy and promptly repaired. When left untreated, a PDA can lead to pulmonary hypertension (pHTN), volume overload with left ventricle (LV) enlargement and dysfunction, and heart failure². The CoA is typically associated with other cardiovascular lesions and is defined as a focal constriction in the descending aorta. The most common clinical presentation in adults is hypertension (HTN), with angina and dyspnea seen in advance cases^{1,3}. Without correction of the CoA, the mean age of death for individuals is 34 years and 75% chance of mortality by age 43³. Surgical intervention for descending thoracic aorta surgery is performed in left lateral thoracotomy using left heart bypass (LHB). LHB is a protective strategy to reduce incidences of post-op paraplegia and paraparesis, where spinal cord injury can occur secondary to coarctation repair⁴.

Learning Objectives

1. Chronic effects of a PDA and aortic coarctation
2. Management of acute right ventricular dysfunction following ligation of a PDA
3. One-lung ventilation and inhaled pulmonary vasodilators in patients with a PDA

Patient Description

49 yo, male, 5'7", 94.3 kg, BMI 32.6, ASA 4. Presents with PDA and post ductal CoA, congestive heart failure (CHF) symptoms for the last 1-2 years (dyspnea on exertion and orthopnea), pHTN, obesity, and chronic renal insufficiency. Patient has atrial fibrillation with rapid ventricular rate, right bundle branch block, and left anterior fascicular block at baseline. Echo report indicates EF 45-50%, bicuspid aortic valve with aortic root aneurysm (5.1 cm), mild pulmonic, aortic, and mitral valve regurgitation, mildly increased LV wall thickness, bi-atrial dilation, and estimated right ventricular systolic pressure (RVSP) 89 mmHg. Patient presents for open descending thoracic aorta repair and PDA ligation.

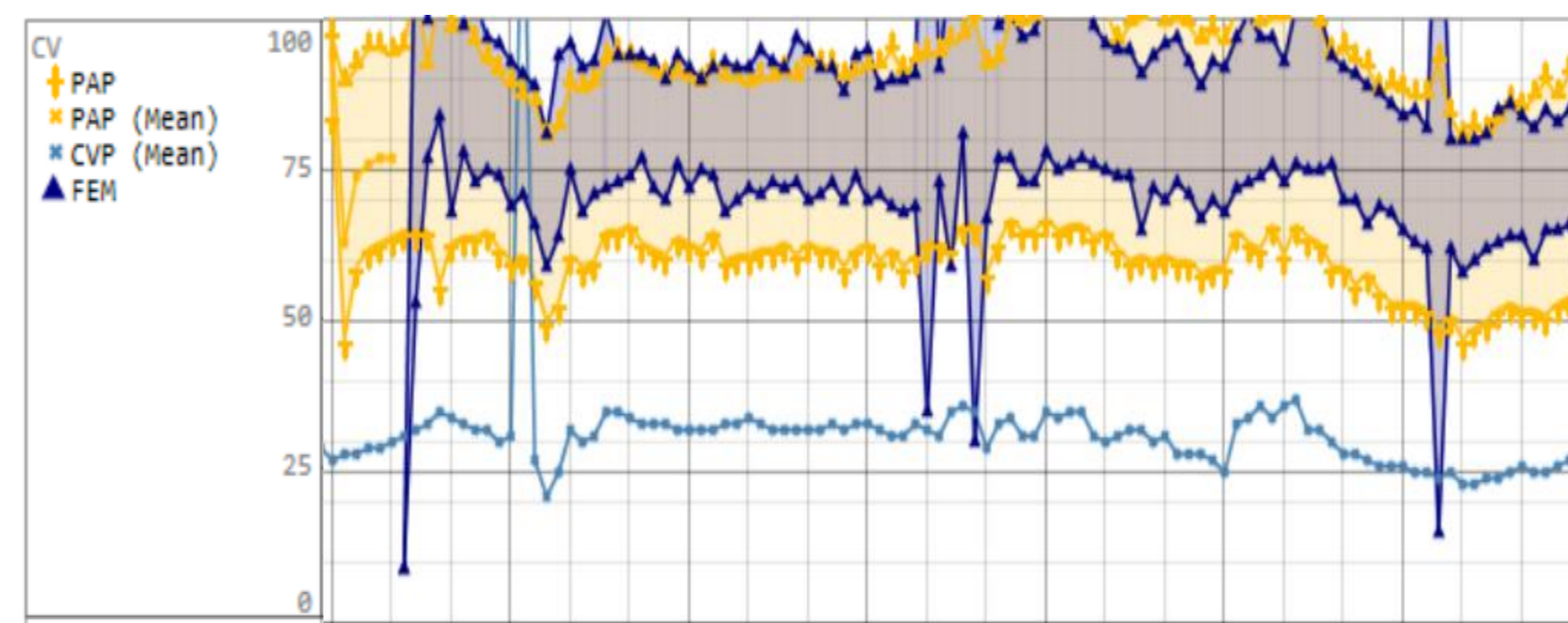


Figure A: Patient PAP, mean PAP, mean CVP, and Femoral arterial pressure waveforms

Significance

Right sided heart failure can be caused by volume loading due to congenital heart disease lesions and pressure loading resulting from pHTN, to name a few causes⁵. Challenges faced in this case included management of right ventricle (RV) dysfunction following ligation of the PDA. The RV is not built to handle large or rapid increase in pulmonary artery pressure (PAP) or large volumes. When coming off pump and attempting to perfuse the heart, there is potential for RV overload and distention. When insufficient systolic functional adaptation occurs, cardiac output is limited, resulting in hypotension and cardiogenic shock⁶. The increased resistance that the RV meets when pHTN (mean PAP \geq 25mmHg) is present can progress to pulmonary vascular disease, increased RV afterload, and RV failure⁷.

Intervention

- Risk of spinal cord injury due to decreased cord perfusion following descending aorta repair is ~20%. Utilizing cerebrospinal fluid (CSF) drainage, the intraspinal pressure was decreased and blood flow to the spinal cord was increased⁸. Patient received spinal drain in order to remove 10 mL of CSF every hour to maintain intracranial pressure \leq 10 mmHg.
- Double-lumen endotracheal tube was used to provide one-lung ventilation (OLV) during the lateral thoracotomy. Patient did not tolerate OLV and SpO₂ dropped to 80%. Continuous Positive Airway Pressure (CPAP) was applied to the nonventilated lung to improve oxygenation.
- Patient had elevated central venous pressure (CVP) and PAP indicative of severe pHTN. Ventilation was exchanged from the anesthesia machine to intensive care unit (ICU) ventilator with aerosolized Flolan (epoprostenol sodium). Inhaled nitric oxide (iNO) was added later to achieve decreased pulmonary vascular resistance.
- Patient developed hemorrhagic shock and profound coagulopathy during surgical repair. The surgeon requested transfusion using CPB, two cell savers, Belmont rapid infuser, and fluid warmer. Perioperatively, a total of 2,250 mL albumin, 40 units cryoprecipitate, 8 units fresh frozen plasma, 7 units packed red blood cells, and 6 units platelets were given.
- Patient developed ventricular tachycardia upon separation from cardiopulmonary bypass (CPB) necessitating internal defibrillation, initiation of a dobutamine infusion, calcium chloride bolus, amiodarone bolus, and volume challenge.
- Left femoral intra-aortic balloon pump (IABP) was inserted to augment coronary perfusion pressure and reduce resistance to systolic outflow.
- Airway exchange from a double-lumen tube (DLT) to endotracheal tube (ETT) in setting of anasarca utilizing a GlideScope and endotracheal tube exchanger was performed.

Discussion and Conclusion

Surgical repair of CoA in adulthood has an increased mortality risk due to the presence of collateral peri-aortic vasculature, pHTN, and end-organ damage caused by chronic HTN³. The shared chronic hypertensive effects of a PDA and CoA presented challenges during the case. The increased resistance to forward flow that the RV meets when pHTN is present can be reduced with the use of inhaled drug therapy. Inhaled administration of Flolan and iNO improve ventilation/perfusion matching by dilating vessels that supply ventilated regions, thereby improving gas exchange. The inhaled route allows for a greater volume of drug targeting the lungs and avoids systemic effects like hypotension. iNO is used to treat acute right-heart failure and acute vasoreactivity in pulmonary arterial hypertension⁹. The addition of iNO to Flolan, a prostacyclin, effectively vasodilated the pulmonary vasculature in combination with an IABP, which provided mechanical circulatory support after CoA repair and PDA ligation¹⁰. These steps allowed for safe separation from CPB, closure of the chest, and transport to the ICU.

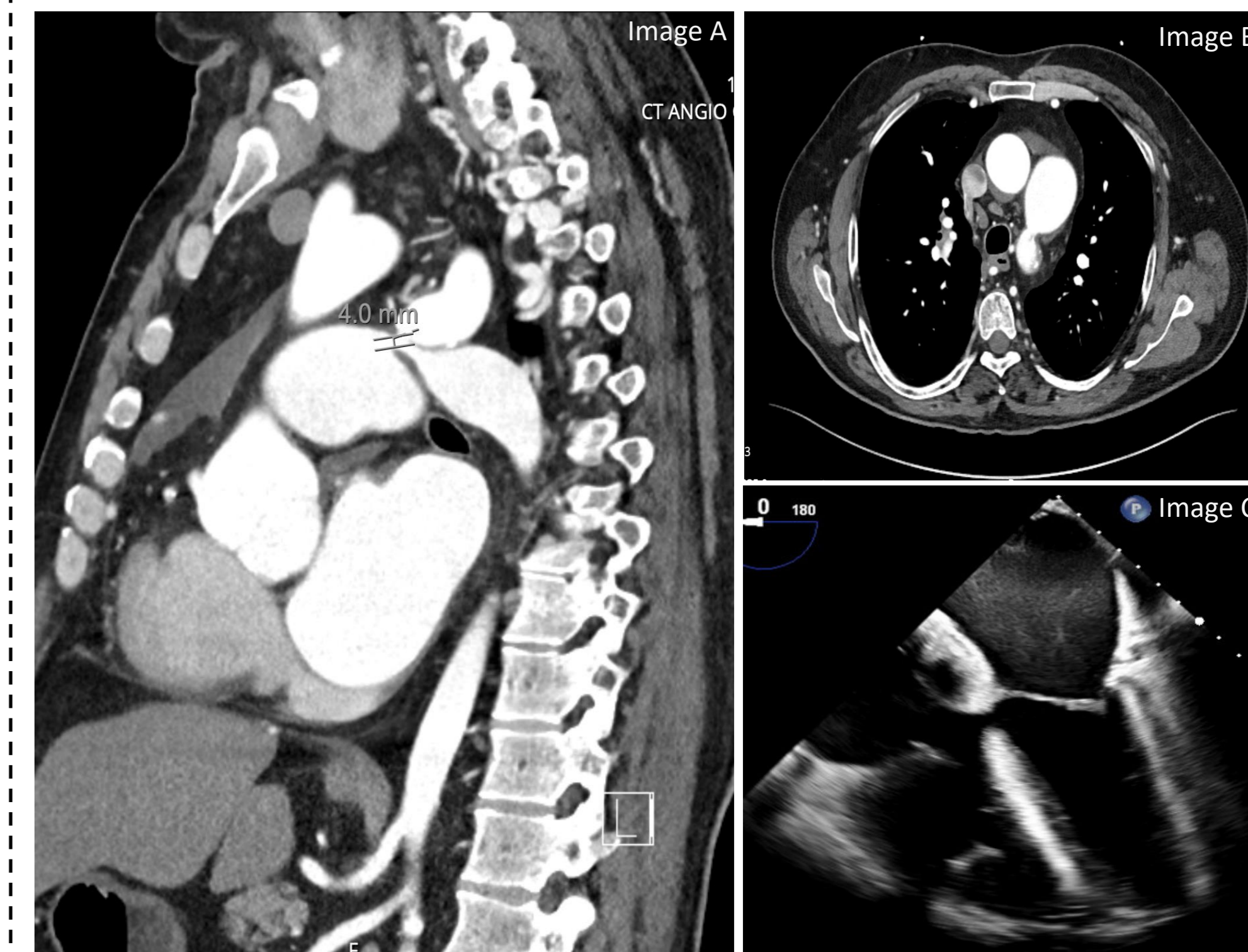


Image A: Sagittal CT scan shows the PDA measurement as the aorta pinches down to the CoA and connects to the left pulmonary artery (PA)
Image B: Axial CT image depicts PDA connecting to an enlarged left PA
Image C: Transesophageal echo image depicts biatrial enlargement, spontaneous echo contrast in the left atrium in the setting of atrial fibrillation and low cardiac output, and RV dilation.

References:

1. Mutluer, F, Celiker, A. (2018). General Concepts in Adult Congenital Heart Disease. *Balkan Med J*; 35: 18-29
2. Van de Sandt, F, Boekholdt, S, Bouma, B. (2011). Patent ductus arteriosus in adults – indications and possibilities for closure. *Neth Heart J*; 19:297-300
3. Alkashkari, W, Albugami, S, Hijazi, Z. (2019). Management of Coarctation of the Aorta in Adult Patients: State of the Art. *Korean Circulation Journal*; 49(4): 298-313
4. Lin, K, Mahmood, F. (2003). A Complication of Left Heart Bypass: A Transesophageal Echocardiographic Finding. *Anesthesiology*; Vol. 98, 1283-1285
5. Konstam, M, Kiernan, M, Bernstein, D. (2018). Evaluation and Management of Right-Sided Heart Failure. *Circulation*; 127:e578-e622
6. Harjola, V, Mebazaa, A, Celutkiene, J. (2016). Contemporary management of acute right ventricular failure: a statement from the Heart Failure Association and the Working Group on Pulmonary Circulation and Right Ventricular Function of the European Society of Cardiology. *European Journal of Heart Failure*; 18, 226-241
7. Vachriery, J, Adir, Yochaj, Barbera, J. (2013). Pulmonary Hypertension Due to Left Heart Disease. *Journal of the American College of Cardiology*; 62(25):D100-D108
8. Epstein, N. (2018). Cerebrospinal fluid drains reduce risk of spinal cord injury for thoracic/thoracoabdominal aneurysm surgery: A review. *Surgical Neurology International*; 9:48
9. Hill, N, Preson, I, Roberts, K. (2015). Inhaled Therapies for Pulmonary Hypertension. *Respiratory Care*; 60(6): 794-805
10. Kong, A, Sin, S. (2020). Short-term mechanical circulatory support (intra-aortic balloon pump, Impella, extracorporeal membrane oxygenation, TandemHeart): A review. *Ann Transl Med*; 8(13):829



EMORY
UNIVERSITY
SCHOOL OF
MEDICINE

Master of Medical Science
Program in Anesthesiology