Title

Group I PAH Requiring Bilateral Orthotopic Lung Transplant (BOLT) and Deceased Donor Renal Transplant (DDRT)

Journal

Proceedings of UCLA Health, 0(0)

Authors

Siddiqui, Mohammed
Balasubramanian, Satish
Velazquez, Ignacio

Publication Date

2023-05-30

UCLA Health

Department of Medicine
2023 Solomon Scholars Research Program

Abstract Form

Hospital Affiliation:	Harbor-UCLA Medical Center
Presenter Name (Last, First):	Siddiqui, Mohammed MD
Co-Authors:	Satish Balasubramanian MD, Ignacio Velazquez MD FACC
Project Title:	Group I PAH requiring Bilateral Orthotopic Lung Transplant (BOLT) and Deceased Donor Renal Transplant (DDRT)

Research Category (please check one):

☐	**Original Research**	☒	**Clinical Vignette**	☐	**Quality Improvement**	☐	**Medical Education Innovation**

Abstract

Introduction: Idiopathic pulmonary artery hypertension (PAH) can present with several complications ranging from vascular to cardiac complications. More common complications include pulmonary trunk dilation and Cor pulmonale. We discuss the presentation of a patient with idiopathic PAH who originally presented with anginal symptoms and shortness of breath with exertion who eventually underwent lung transplant, tricuspid valve replacement and annuloplasty given the extent of PAH.

Case Report: 58-year-old female with a medical history of severe PAH, idiopathic thrombocytopenic purpura, chronic kidney disease and type II diabetes presents with two weeks of chest pain and a longer history of shortness of breath with exertion. She describes the pain as substernal, exertional chest pressure with radiation to her back. She states this pain improves with rest. In the ED, vitals were notable for a blood pressure of 97/46 and oxygen saturation of 91% on room air. Physical exam was notable for a right ventricular heave and IV/VI systolic murmur auscultated loudest at the left lower sternal border. Labs were notable for indeterminate troponin levels of 0.11 and 0.14. B-type natriuretic peptide was 1,148. An electrocardiogram revealed sinus rhythm with an incomplete right bundle branch block and right ventricular hypertrophy without evidence of ischemia. A transthoracic echocardiogram (TTE) from 6 months prior revealed a normal left ventricular ejection fraction (LVEF) of 65-70%, normal wall motion, severe tricuspid regurgitation (TR), severe right atrial and right ventricular (RV) dilation as well as an increased estimated right ventricular systolic pressure (RVSP) suggesting pulmonary hypertension. Right heart catheterization revealed severe group I PAH with pulmonary artery mean pressure of 61mmHg and left heart catheterization revealed external compression of the ostial left main coronary artery by the pulmonary trunk with a minimum lumen area (MLA) of 13.5mm^2. During this admission, the patient's anginal symptoms resolved and she was discharged with outpatient follow-up. Patient was re-admitted to the hospital several times over the next 2 months due to volume overload thought to be secondary to worsening PAH and decreasing diuretic effectiveness. During these admissions, the patient required a bumetanide drip, metolazone and vasopressor support for diuresis. She eventually underwent continuous renal replacement therapy (CRRT) on several occasions for fluid removal given her inadequate diuresis with medical therapy. The patient was eventually referred to an outside hospital for lung transplant given her severe group I PAH. At the outside hospital, the patient was optimized with Epoprostenol and eventually underwent bilateral orthotopic lung transplant (BOLT), tricuspid valve replacement and annuloplasty. A TTE post-transplant revealed a LVEF of 55-60%, an RV/LV ratio of 0.92, trace TR and an estimated RVSP of 27mmHg. Given her worsening CKD over the course of multiple hospitalizations, she eventually underwent deceased donor renal transplant after BOLT.

Discussion: This case describes several interesting complications of idiopathic PAH. Complications of pulmonary hypertension include vascular, cardiac, and pulmonary complications. Among vascular complications, there can be pulmonary dilatation, pulmonary artery dissection and in situ thrombosis. Cardiac complications can include right heart failure, pericardial effusion, and tamponade. Pulmonary complications include cavitation and infection. Few case reports describe external compression of the left main coronary artery by a dilated pulmonary trunk. In our case, the MLA of 13.5mm^2 would suggest the compression is not hemodynamically significant, and instead a benign finding. Typically, an MLA cutoff closer to 5-7mm^2 is more indicative of hemodynamic significance. In other case reports, there have been instances of external compression requiring stent placement. In addition to this benign but rare finding, our case report reveals a successful BOLT which helped normalize this patient's pulmonary artery pressures as evident in the post-transplant echocardiogram. The patient is no longer experiencing anginal symptoms or shortness of breath with exertion.