



FIT Clinical Decision Making

PULMONARY THROMBI AS A CAUSE OF DYSPNEA IN ADULT CYANOTIC HEART DISEASE

Poster Contributions
Poster Hall B1
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Session Title: FIT Clinical Decision Making: Structural Heart Disease and Pulmonary Hypertension
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Background: Silent pulmonary thrombi can occur in up to 15-20% of Fontan-corrected adult congenital patients, but the prevalence or effects of pulmonary thrombi in uncorrected cyanotic heart disease is not well defined.

Case: A 37 year old male with uncorrected cyanotic heart disease, characterized by double inlet left ventricle, large secundum atrial septal defect, left pulmonary artery (PA) atresia, status-post classic Blalock-Taussig shunt to the right PA, multiple aorto-pulmonary collaterals with pulmonary hypertension (PH), diastolic heart failure, and severe restrictive lung disease secondary to dextro-scoliosis, presented with progressive dyspnea on exertion for one week. He also noted a six pound weight gain and right-sided pleuritic chest pain. He reported dietary and diuretic compliance. On exam, he appeared somnolent and dyspneic. Initial laboratory analysis showed acute on chronic respiratory acidosis, normal renal function, hypoxemia, and polycythemia.

Decision Making: After admission, the patient became progressively more hypoxemic, requiring non-invasive ventilation with increasing oxygen to maintain saturations at his baseline of 60-70%. Central venous pressure (CVP) measurements through a right internal jugular vein line were 1-3 mmHg. Due to PH from the aorto-pulmonary collaterals, with concern for poor pulmonary perfusion at low filling pressures, we targeted a higher CVP of 5-7 mmHg with fluid resuscitation. Also considering the patient's PH and likely impaired ability to augment tidal volumes secondary to restriction, resulting in hypercapnia, we evaluated for pulmonary embolism with CT angiography. The initial read suggested asymptomatic mural thrombi, but given our high index of suspicion that these were pathogenic, we performed a ventilation-perfusion scan, showing multiple V/Q mismatches. The patient was started on anticoagulation, with improvement.

Conclusion: This case demonstrates the importance of understanding cardio-pulmonary physiology in the care of complex adult congenital patients. Although silent pulmonary mural thrombi can occur with PH, they may contribute to symptoms in the setting of impaired lung function.