

An Unusual Cause of Severe Pulmonary Hypertension in a Young Woman

Dear Editor,

A 25-year-old Cambodian lady diagnosed with pulmonary hypertension in her native country came for second opinion on the further management of her condition. She had become increasingly breathless in the last 2 years, with further deterioration from New York Heart Association (NYHA) II to III in the last few months despite starting sildenafil therapy. She had history of congenital heart disease with cyanosis at birth and had heart operation done for “hole in the heart” at the age of 9 years in Paris, France. However, her operation notes had been lost. After operation, she was pink and well for many years.

Physical examination was unremarkable except for the presence of loud second heart sound at the pulmonary area. Her blood pressure was 97/55 mmHg and heart rate was 73 bpm. The baseline SpO₂ was 96%, which dropped to 74% after walking 100 meters. The SpO₂ slowly picked up to 88% after a few minutes of rest.

Her electrocardiography (ECG) showed right ventricular hypertrophy with tall R wave in the right precordial leads and right axis deviation. She was in sinus rhythm.

Her transthoracic echocardiogram showed severe right ventricular hypertrophy with moderately dilated right ventricle and mildly reduced right ventricle systolic function. There was severe pulmonary hypertension with pulmonary artery systolic pressure of 106 mmHg. Interestingly, there was significant flow turbulence seen at the superior aspect of the interatrial septum near entrance of the pulmonary vein (Fig. 1) with continuous wave (CW) Doppler velocity of 2.6 m/s suggestive of pulmonary vein stenosis. This was adjacent to the interatrial septum patch from previous surgical repair. Agitated bubbles contrast saline studies were performed with no right to left shunt demonstrated.

Computed tomography (CT) thorax was performed mainly to visualise all the pulmonary veins. This showed severe stenosis of all the pulmonary venous ostia with engorgement and dilatation of the proximal portion of the pulmonary veins. In addition, there was also partial anomalous pulmonary venous drainage (PAPVD) of the small diminutive (3 mm) right superior pulmonary vein into superior vena cava. The ostium of this vein was severely stenotic and slit-like. The

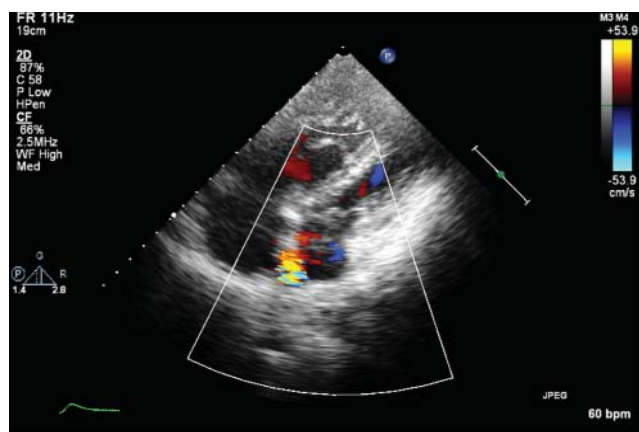


Fig. 1. Significant flow turbulence seen on color Doppler at the orifice of the right pulmonary vein.

pulmonary arteries were dilated in keeping with pulmonary arterial hypertension.

Pulmonary vein stenosis can occur congenitally or as a complication of radiofrequency catheter ablation for atrial fibrillation, or after repair of total anomalous pulmonary venous connection. The management for this condition continues to remain challenging and it can be lethal if not treated properly. This lady had severe pulmonary hypertension as a result of the pulmonary vein stenosis. The current available treatment options for the pulmonary vein stenosis include transcatheter balloon angioplasty with or without stenting or direct surgical repair. The surgical techniques include anastomotic revision (for patients with acquired PV stenosis), intraoperative stent placement, patch venoplasty (using atrial tissue, pericardium, or polytetrafluoroethylene), scar excision with primary repair and sutureless pericardial marsupialization. Sutureless pericardial marsupialization was associated with satisfactory midterm results and appeared superior to other conventional techniques.¹⁻³

The patient was still considering the various options for the management of her condition.

In patients with significant pulmonary hypertension, where there is no obvious common cause of pulmonary hypertension being identified from echocardiogram, we may need to pay special attention to rare causes such as pulmonary vein stenosis while performing echocardiogram studies. This is important especially in patients who have history of radiofrequency catheter ablation done for atrial flutter or fibrillation, or after repair of total anomalous pulmonary venous connection. Radiofrequency catheter ablation done for patients with atrial flutter or fibrillation has become commoner nowadays and there is increasing number of paediatric patients with the repair done for total anomalous pulmonary venous connection and who have grown into their adulthood. It is important for us to pay special attention to this echocardiographic finding that suggests pulmonary vein stenosis while performing echocardiogram in this group of patients, so that we do not miss this important clue to the diagnosis of pulmonary vein stenosis.

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