

Steeling Home: The Rarest Kind of Pulmonary Hypertension

*Y Moayed, M Aversa, JG Duero Posada, M Alhussein, HJ Ross,
M Simons, EM Horlick, J Granton, S Mak*

University Health Network, Toronto General Hospital and Mount Sinai Hospital,
University of Toronto, Ontario, Canada.

CASE PRESENTATION

A 40-year-old woman was transferred for double lung transplantation for pulmonary arterial hypertension (PAH) and worsening clinical status despite therapy including Epoprostenol, Macitentan and Tadalafil. The pathology of the explanted lungs showed intimal hyperplasia and plexiform lesions consistent with idiopathic pulmonary hypertension. Post lung transplantation, hospitalization was complicated by ongoing dyspnea and signs of severe right heart failure for several months. Failure to improve prompted review of her clinical and hemodynamic status. History and current physical examination was positive for a continuous murmur noted during pregnancy 8 years prior. She also had findings of severe tricuspid regurgitation with an elevated JVP with CV waves and a pulsatile liver.

INVESTIGATIONS

Transthoracic echocardiogram

Severe right ventricular enlargement with an right ventricular systolic pressure of 70 mmHg, severe tricuspid regurgitation.

Right Heart Catheterization:

- RA 20, RV 64/1/16, PA 56/14/37
PVR 1.73 WU, LVEDP 17
- PA saturation 74%, FA 89%,
- CO 9.8 L/min (Fick) CI: 6.5 L/min/m²

Repeat right heart catheterization revealed a large pelvic arterial venous fistula and a markedly dilated IVC, which was the cause of the high output heart failure and likely pulmonary vascular remodeling responsible for the pre-transplant development of PAH.

DIAGNOSIS



Figure 1.. CT angiogram showing massively dilated IVC and direct fistula between the left common iliac artery and vein.

TAKE HOME POINTS

1. PAH accounts for less than 5% of all lung transplants and associated with the highest 30-day mortality.
2. This patient's common iliac artery-venous fistula resulted in high-output cardiac failure. Systemic AV fistulas are less well recognized as a cause of PH mostly isolated to case reports.
3. This case highlights the importance of a systematic approach to the physical exam and investigations.

Figure 2. Panel A through E
Interventional procedure showing VSD closure device partial occluding of large left common iliac artery and vein (A-C). Subsequently treated with 3 overlapping covered stents (D-E).

