
# Pulmonary Hypertension: Common Condition, A Relatively Rare Entity

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Introduction: PoPH (portopulmonary hypertension) is being increasingly recognized as an important cause of PAH despite lacking precise diagnostic and treatment modalities. The mortality rate is significantly higher compared to IPAH. Here is a case of PoPH presenting with cardiogenic shock.

Report:
A 42 year old female with no known comorbids or high risk behaviours presented with complaints of acute onset dyspnoea for 2 days, starting off as grade 2 NYHA rapidly progressing to Grade 4, and oliguria. The patient also had complaints of lower limb edema for the past 1 week. Examination revealed pan digital clubbing with pallor and cyanosis. BP was 70/40, SpO2 was 89%. RS and CVS examination revealed no abnormality. Abdominal examination showed moderate ascites with shifting dullness. Patient was started on ionotropes. Blood work up **showed raised total count with thrombocytopenia, deranged RFT and LFT with increased billirubin and reduced albumin**. A 2d echo was done and showed a **dilated RA and RV with no RV hypertrophy**, **dilated MPA and severe TR with increased TRPG**. A **CTPA was done and showed no evidence of Pulmonary embolism**, acute or chronic. However, visualized liver sections were showing corase echoes. An ultrasound was ordered which showed coarse liver echoes with hepatofugal flow, minimal spleen and moderate ascites. She however continued to be ionotrope dependent. ANA profile and APLA were done and were negative. Viral markers were negative. Sr.Ceruloplasmin was also within limits. Anti LKM antibodies, AMA ASMA were negative. In order to rule out cardiac shunts, Bubble contrast echo was done. It showed an RA-LA cycle time of 8 cycles implying no extra or intra cardiac shunts. Putting the picture together, we came to a possible diagnosis of PORTO PULMONARY hypertension in the setting of DCLD, probably cryptogenic. A V/Q scan was needed to exclude CTEPH but could not be done. The patient was given supportive care and ionotrope therapy, but succumbed to hypoxemia and had to be intubated and put on mechanical ventillator and suffered a cardiac arrest.

Conclusion:
The identification of Portal hypertension as a sole cause of Pulmonary hypertension requires a high index of clinical suspicion as these patients are considered poor candidates for liver transplantation. Numerous studies have been conducted to explore treatment options and to mitigate risk before transplant, some of them being the PATENT 1 trial and the PORTIGO trial which showed good promise with Macitentan.