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Hepatopulmonary syndrome (HPS)

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Case Presentation summary: 55 year old gentleman, Married with three children, currently unemployed, normally fit and well, Known Type 2 Diabetes Mellitus (on metformin) Pancytopenia with Macrocytosis(two Bone marrow biopsies. Reduced Cellularity and bone marrow trephine was inadequate. HIV negative)Patient presented to Ealing Hospital on 9/9/16 with a dry cough and increased shortness of breath, on a background of chronic progressive dyspnoea. CT Chest excluded pulmonary embolism or significant pneumonic process. CT study of the abdomen showed moderate cirrhosis of the liver, with portal hypertension and splenic and gastric varices. OGD confirmed oesophageal varices. The aetiology remains unclear; liver screen showed normal autoantibodies, Heptititis screen negative along with negative gene for haemachromatosis in the past as per Gastroenterology clinic letters.Ventilation perfusion studies of the lung showed evidence of a right to left shunt, although could not determine cardiac or pulmonary origin.Subsequently he had an initial TOE which showed Normal RV size and function with no evidence of cardiac shunt on Doppler studies.The TOE suggested the possibility of a right to left shunt but the picture quality was suboptimal Patient had another review by cardiology Consultant who performed a contrast TTE on 12/12/2016 which confirms the presence of a Right to Left shunt with agitated gelofusine microbubbles appearing in the left atrium 5 cardiac cycles after appearing in the right atrium. Cardiac MRI does not show evidence of a cardiac shunt and with a Qp:Qs ration of <1.0 supports the possibility of a right to left shunt.In summary the above findings are compatible with Pulmonary Right to Left shunt.Patient will be presented in the cardiac MDT and was given cofmirmed diagnosis of HPS Patient was reviewed by the Gastroenterology team while in patient and discharged home with Home oxygen and referred to Liver transplant unit king's college London.

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