63-year old female with dyspnea

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A 63-year-old female presented to pulmonary clinic with the complaint of dyspnea. Shortness of breath had been slowly worsening over the previous 4 years, to the point that it took her one hour to make her bed. She complained of an occasional nocturnal cough which was non-productive. One week prior to this presentation, she had an episode of syncope while walking outside.

Her PMH was significant for atrial fibrillation, CAD (70% LAD lesion without intervention), obstructive sleep apnea with good compliance with CPAP therapy. Her current medication list included esomeprazole, felodipine, isosorbide dinitrate, lisinopril, metoprolol XL, warfarin, and PRN hydrocodone. She had a 60 pack-year smoking history, and currently smoked ¼ pack per day. She denied alcohol or illicit drugs. She was disabled secondary to her dyspnea, and formerly held office jobs. She denied any bird or dust exposures.

Her physical exam revealed the following: pulse 53, BP 149/78, 97% saturation on room air, respirations 22, weight 192 pounds. She was mildly obese in no acute distress. Cardiovascular exam was irregularly irregular, with no murmurs, no JVD, and a split S2. Lungs were clear bilaterally, and she had mild resting tachypnea. Abdomen was soft, nontender, and without hepatojugular reflex. She had no clubbing or peripheral edema.

Labs and studies revealed a normal CBC, complete metabolic profile, thyroid studies, brain natriuretic peptide, and ANA. Her ESR was 11. Chest X-ray was unremarkable, and sniff test demonstrated normal movement of the diaphragms. Pulmonary function testing revealed an FVC of 1.43 L (40% pred), FEV_1 1.02 L (37% pred), FEV_1/FVC 71%, TLC 3.11 L (54% pred), DLCO 16.37 (73% pred). She was sent for a treadmill stress test, but the cardiologist thought her too dyspneic to safely do the test, so a catheterization was performed. This demonstrated an EF of 65%, with 3-vessel coronary disease. Her right heart catheterization demonstrated a pulmonary arterial pressure of 55/19 (mean 35), PCWP 19, and CO 13.4 L/min. Saturation study is demonstrated in figure 1.

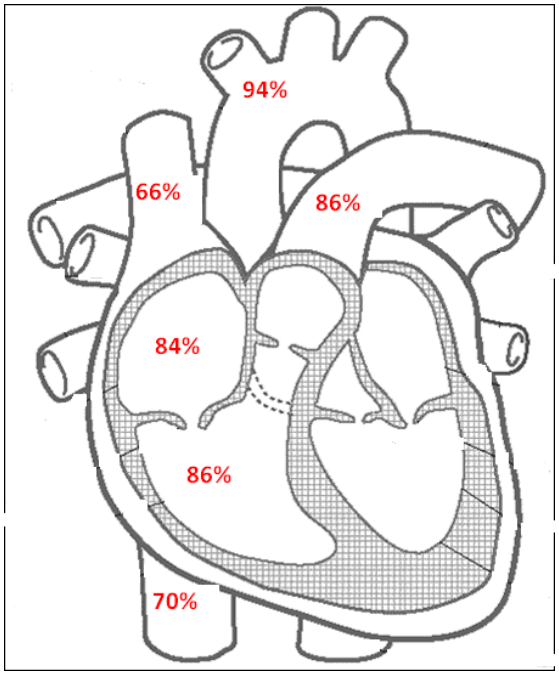

Figure 1- saturation study results

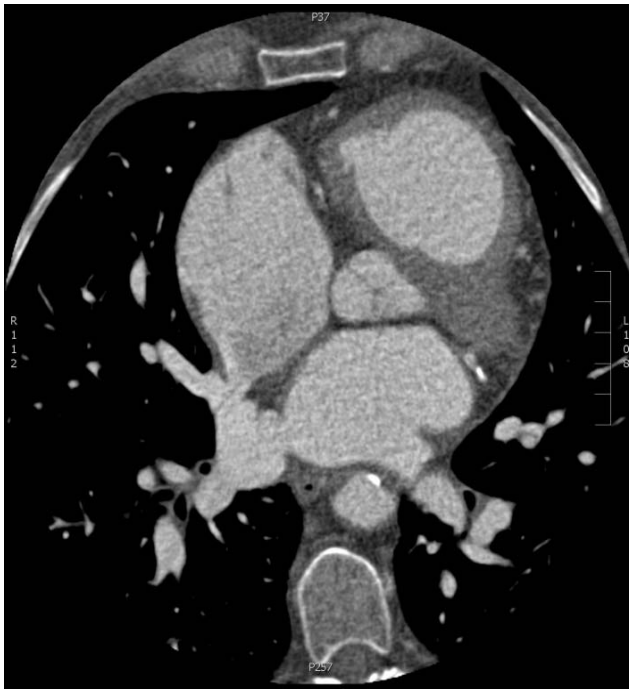

Figure 2- Mediastinal view of chest CT

Cardiac echo demonstrated normal LV function, moderate to severe right atrial and ventricular dilatation, and an estimated RVSP of >65 mm Hg. No ASD or VSD was seen. Chest CT demonstrated normal lung parenchyma, with a mediastinal abnormality shown in Figure 1.

What is the diagnosis?

Diagnosis: Partial anomalous pulmonary venous return

Partial anomalous pulmonary venous return (PAPVR) is a rare form of congenital heart disease in which one or more of the pulmonary veins drain into the right heart (right atrium, superior vena cava, inferior vena cava, or innominate vein). This condition was first described in 1739. As opposed to total anomalous pulmonary venous return (a neonatal lesion that is PDA-dependent and fatal if not recognized and surgically corrected), PAPVR can present in adults, and has been found to have an incidence of 0.4-0.7% in autopsy series.

Most cases of PAPVR are asymptomatic, and drainage of one pulmonary vein to the right heart circulation is not thought to cause any symptoms under normal conditions.

A variation of PAPVR is Scimitar syndrome, or pulmonary venolobar syndrome. This condition consists of hypoplasia of the right lung, with anomalous venous return of part or all of the right lung to the inferior vena cava. It is often asymptomatic, and related to dextrocardia or other cardiac abnormalities. It is called the scimitar syndrome after the "scimitar vein" (which has the appearance of a sword, or scimitar), in the right lower lung field (see figure 3).

Decision as to how to proceed regarding surgical repair depends upon the symptomatology of the patient and the calculated shunt ratio, which is a representation of the pulmonary to systemic flow. It can be calculated using the equation:

> qp:qs= (Aortic sat-Mixed venous sat)/(Pulmonary vein sat-Pulmonary arterial sat)

(The mixed venous saturation must be a pre-shunt value for calculation, and pulmonary venous saturation can be estimated to equal aortic saturation.)

Asymptomatic patients should be treated conservatively, as many will never develop symptoms. Those with a qp:qs ratio of greater than 1.8 should be considered for surgical intervention, as these patients are likely to proceed to develop symptoms of pulmonary hypertension and right heart failure.

Our patient had severely limiting symptoms (NYHA class III), and her qp:qs ratio= (94%-67%)/(94%-86%)= 3.38. She was referred to cardiothoracic surgery for evaluation, and underwent repair of PAPVR shortly thereafter. Unfortunately, she developed severe post-operative hypoxemia and hypotension, and died one day after surgery.

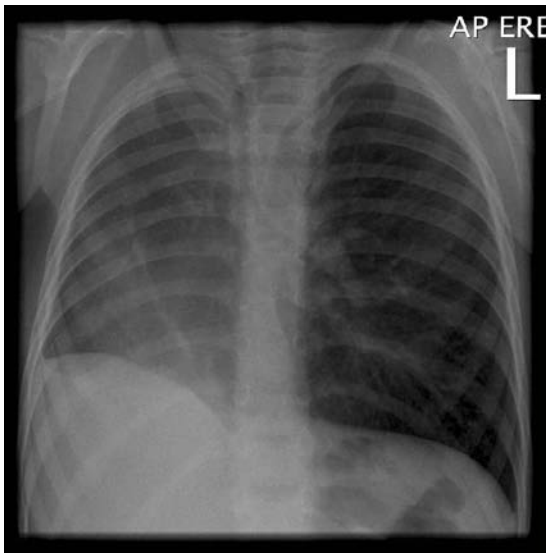

Figure 3- Scimitar syndrome, with scimitar vein noted in right lower lung field

References

Holt PD, Berdon WE, Marans Z, Griffiths S, Hsu D. Scimitar vein draining to the left atrium and a historical review of the scimitar syndrome. Pediatr Radiol 2004: 34: 409-413.

Healey JE. An anatomic survey of anomalous pulmonary veins; their clinical significance. J Thorac Surg 1952: 23: 433-444.

Wilkinson JL. Congenital heart disease: hemodynamic calculations in the catheter laboratory. Heart 2001: 85: 113-120.

Van Meter C Jr, LeBlanc JG, Culpepper WS 3rd, Ochsner JL. Partial anomalous pulmonary venous return. Circulation 1990: 82 (5 Suppl): IV195-198.