74. Hypoxemia in Partial Anomalous of Pulmonary Venous Return With Sinus Venosus Atrial Septal Defect

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Body

Background: Severe pulmonary arterial hypertension associated congenital heart disease (CHD) and Eisenmenger's syndromes (ES) were difficult to differentiate.

Case: A 30-year-old woman presented with progressive exertional dyspnea and orthopnea (The New York Heart Association functional class III (NYHA FC IV)). Chest X-ray (CXR) (Figure 1.1). Transthoracic echocardiogram showed Left ventricular ejection fraction 64% with markedly dilated of right heart with LV D-shape (Figure 1.2). Right heart catherization (RHC) was performed.

Discussion: RHC results showed oxygen step up at the right atrium. The resting oxygenated blood in aorta was 84%, Qp:Qs = 0.46, pulmonary vascular resistance = 10 Wood. After 100%oxygenation, the blood was 100% oxygenated, Qp:Qs =1.32, pulmonary vascular resistance =10 Wood. It could be implied that pulmonary vasodilator minimized the right to left shunt. Partial anomalous of pulmonary venous return with sinus venosus of atrial septal defect was confirmed by cardiac imaging (Figure1.3,1.4). CXR was not compatible with ES even though high pulmonary vascular resistance. The patient was treated with Sildenafil. 2 weeks later, her symptoms were improved (NYHA FCII), no orthopnea. We will re-RHC in 6 months for further planning whether shunt correction required.

Pulmonary hypertension can present with orthopnea that improved with Sildenafil. Not all of CHD with hypoxemia was ES. CXR was the clue to differentiate severe pulmonary arterial hypertension in CHD from ES in order to give the patient a chance for surgical correction.

