

Congenital Pulmonary Vein Stenosis

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A 55 years female presented with fatigue on exertion. On transthoracic echocardiography there was a secundum atrial septal defect with left to right shunt. Transesophageal echocardiography confirmed presence of secundum atrial septal defect (Figure 1a). Left sided pulmonary vein flow was normal (Figure 1b). Right upper pulmonary vein was very small in caliber with turbulent continuous flow on colour Doppler evaluation (Figure 1c)

Congenital pulmonary vein stenosis may occur as focal stenosis at the point of entry into left atrium or as generalized hypoplasia of pulmonary vein as in this patient. Pathology may involve one or more pulmonary veins. It is almost always associated with congenital cardiac defects eg:- VSD, ASD, TOF, Tricuspid or Mitral atresia or A-V septal defects. Stenosis of multiple pulmonary veins produces pulmonary venous congestion with dyspnoea, cough & hemoptysis. Secondary pulmonary artery hypertension is a consequence. Unilateral pulmonary vein stenosis does not produce clinical symptoms due to pulmonary flow distribution to the opposite lung. In our patient symptoms were largely due to hemodynamic impact of large left to right shunt (echocardiographic QP:QS-3.2:1). Pulmonary vein stenosis could be partly contributing to symptoms. Patient could not afford cardiac CT scan. Transesophageal echocardiography with Doppler evaluation is usually enough to give anatomical diagnosis & hemodynamic information.

Grant Support

None

Conflict of Interest

None

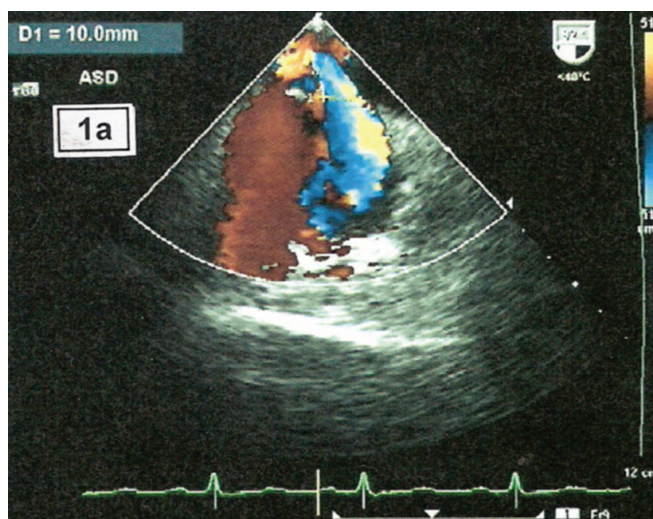


Figure 1a. Transesophageal echocardiographic bicaval view showing secundum ASD.

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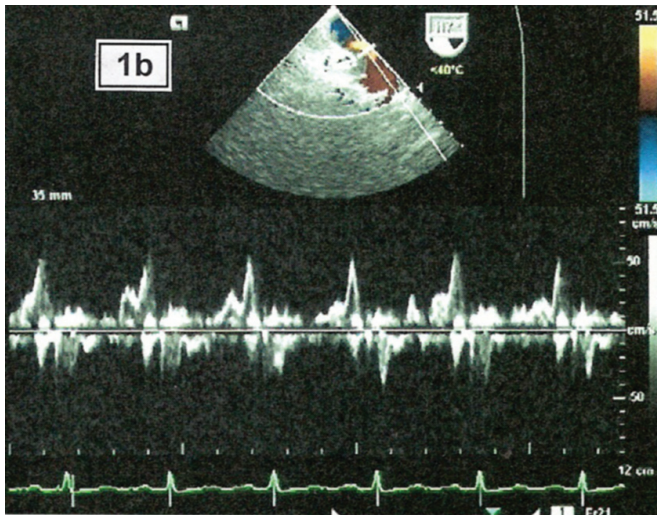


Figure 1b. Transesophageal echocardiographic view showing left sided pulmonary veins.

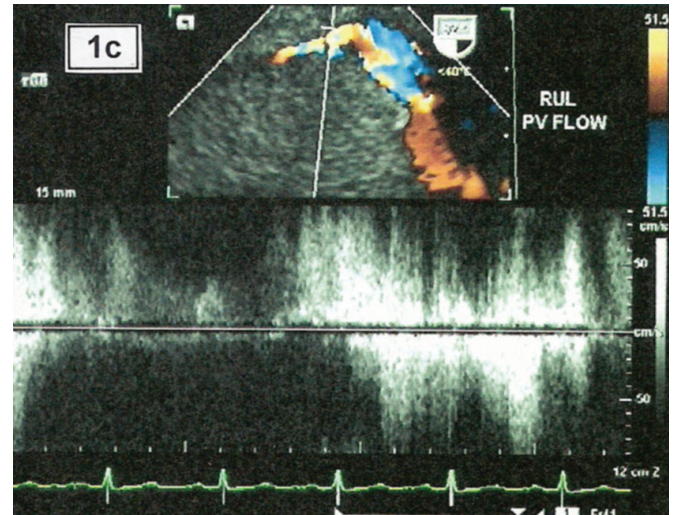


Figure 1c. Transesophageal echocardiographic view showing generalized hypoplasia of right upper pulmonary vein with continuous turbulent flow.