

# A RARE CASE OF CONGENITAL STENOSIS OF THE COMMON RIGHT PULMONARY VENOUS OSTIUM

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## BACKGROUND:

Pulmonary vein stenosis refers to a variety of congenital and acquired disorders characterized by narrowing of the one or more of the pulmonary veins. Pulmonary vein stenosis is challenging to diagnose and thus is associated with high mortality.

## CASE:

A 20 year-old Caucasian female with scoliosis presented with dyspnea on exertion and exercise intolerance. A transthoracic echocardiogram revealed a severely dilated right ventricle (Figure 1A) with mildly reduced systolic function and severely elevated systolic pressure (Figure 1B). Subsequent right heart catheterization confirmed severe pulmonary hypertension. Additional workup including a VQ scan and autoimmune serologies were normal. Treatment for pulmonary arterial hypertension was initiated with Sildenafil and Ambrisentan without significant improvement. Thus, cardiac CT was order for further evaluation.

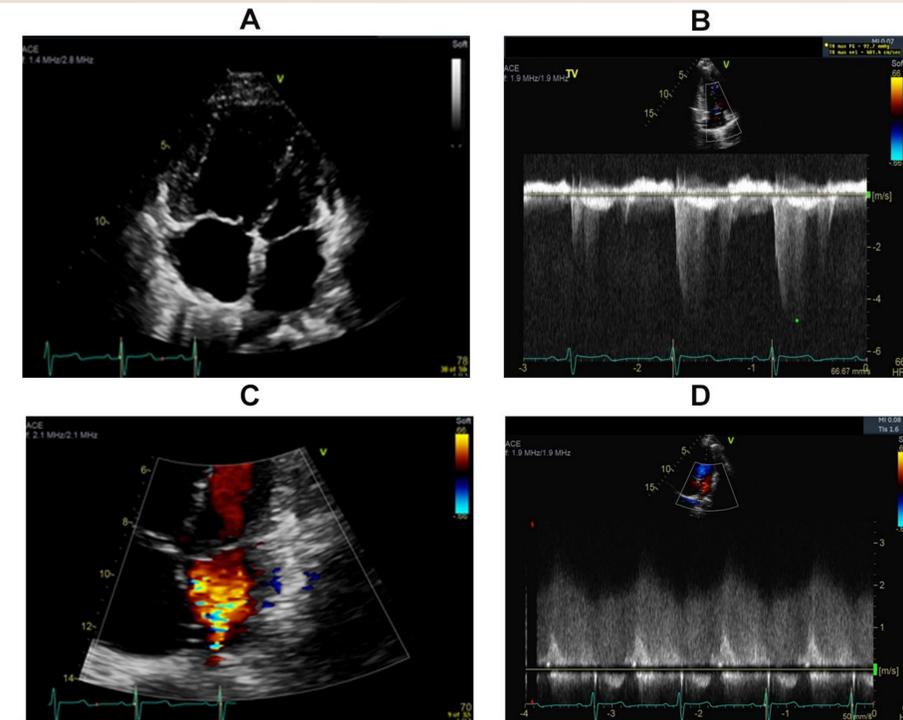


Figure 1.

- A. Apical four-chamber view showed a severely dilated right ventricle.
- B. Severely elevated right ventricular systolic pressure—peak pressure based on the tricuspid regurgitant jet was 93mmHg.
- C. Color Doppler demonstrated accelerated flow through the right pulmonary vein.
- D. Continuous Doppler revealed a maximal flow velocity of 2.8 m/sec through the right pulmonary vein.

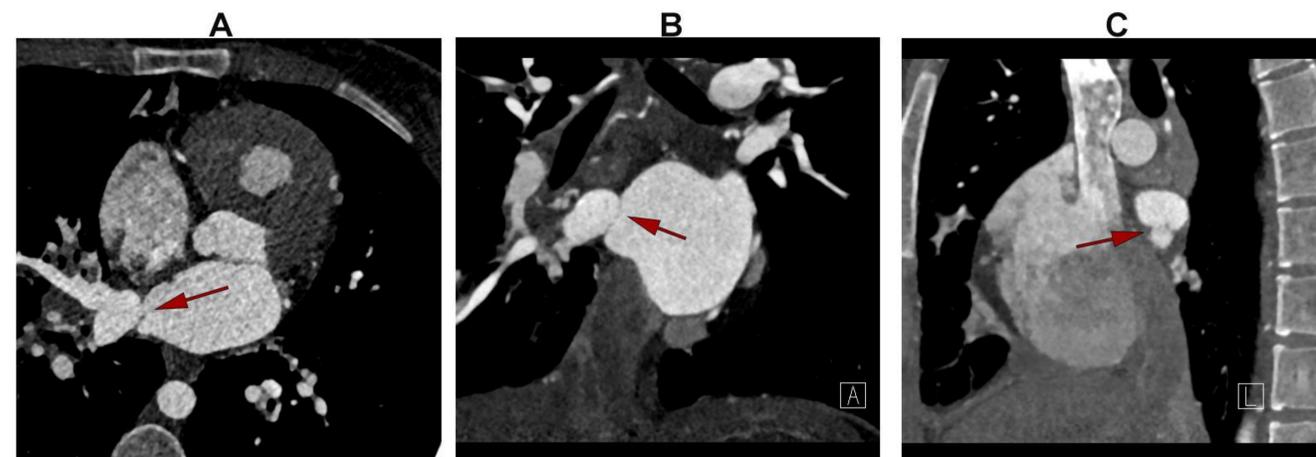


Figure 2. Red arrows pointing to the stenotic ostium of the common right pulmonary vein. A. Axial view. B. Coronal view. C. Sagittal view.

## DECISION MAKING:

Cardiac CT demonstrated right inferior and superior pulmonary veins converging to a common trunk before entering the left atrium. The ostium of the right common trunk demonstrated an incomplete membrane rendering it stenotic (Figure 2A, B, C). On further review of her echocardiogram, color and continuous Doppler (Figure 1C, D) were consistent with significantly elevated right pulmonary venous flow velocity.

**CONCLUSION:** This case describes a rare congenital stenosis of the common right pulmonary vein ostium that lead to severe pulmonary hypertension and severe right ventricular dilation.