Total Anomalous Pulmonary Venous Connection

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23 years female presented with breathlessness and palpitation for last 10 years. She also had marked kyphoscoliosis.

Her echocardiography showed dilated right atrium(RA) and right ventricle(RV). There was ASD secondum, of 13 mm in size. Pulmonary veins were not seen opening into left atrium. There was a large venous channel, in which all pulmonary veins drained and this channel was opening into RA near the opening of superior vena cava (Figures 1, 2). On contrast echocardiography, there was right to left shunt through ASD (Figure 3).

Total anomalous pulmonary venous connection (TAPVC) is a rare but heterogeneous anomaly, accounting for ≈1% to 3% of congenital heart disease cases.¹ Historically, TAPVC has led to a high mortality rate of ≈80% in the first year of life without intervention.² In 1959, Darling and associates proposed a classification, also based on the anatomy of the anomalous connection.³ Four types were identified: type 1, anomalous connection at the supracardiac level; type 2, anomalous connection at the cardiac level; type 3, anomalous connection at the infracardiac level; and type 4, anomalous connection at two or more of the above levels. Treatment is surgical correction. The goal of surgery is to redirect all pulmonary veins to the left atrium through wide and nonrestrictive connection.⁴

Our patient had type 2 TAPVC and underwent successful surgical repair.

References

3. Darling RC, Rothney WB, Craig JM. Total pulmonary venous drainage into the right side of the heart: report of 17 autopsied cases not associated with other major cardiovascular anomalies. Lab Invest 1957; 6:44–64.