

Non-obstructive supracardiac total anomalous pulmonary venous return in a 3-year-old patient who was admitted with complaint of mild cyanosis

Üç yaşında hafif siyanoz şikayetiyle başvuran hastada obstrüktif olmayan suprakardiyak tip total anormal pulmoner venöz dönüş anomali

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A three-year-old boy was admitted to the hospital with complaints of fatigue and bruising around the mouth. Physical examination revealed central cyanosis, with oxygen saturation of 85% from a finger, mild hepatomegaly and 2/6 midsystolic ejection murmur heard best along the left sternal border.

The chest X-ray was normal. Electrocardiogram demonstrated P-pulmonale, right ventricular hypertrophy, and right axis deviation. Two-dimensional echocardiography revealed dilated right-sided structures of the heart and right-to-left shunt via atrial septal defect (ASD). Moderate tricuspid valve regurgitation was detected. Further evaluation revealed a small left atrium (LA) and an echo-free space behind it (Figs. A, B). Pulmonary veins discharging into the LA could not be visualized. Cardiac catheterization was performed in order to better visualize the total anomalous pulmonary venous return (TAPVR) and for preoperative assessment. A 5-F MPA2 catheter was passed to the inferior vena cava (IVC), right atrium (RA), superior vena cava (SVC), and large left innominate vein (LIV) first through the femoral vein and then to the vertical vein (VV), and pulmonary venous confluence (PVC) was opened at that point. Blood gas samples were taken from the accessed sites. Oxygen saturation of the SVC was found as 77% and of the PVC as 95.4%. On imaging of non-ionic contrast medium given into pulmonary arteries, right and left pulmonary veins were seen to drain into the PVC, which was located behind the LA region and opened to the large LIV through the vertical vein (Videos 1, 2*; Fig. C). The SVC was seen to open to the RA, and contrast medium was seen to pass to the LA through the ASD. Medical supportive treatment was initiated and the case was referred to cardiovascular surgery.

Figures- (A) Subcostal view on echocardiography showing small LA, dilated RA, SVC, and left and right innominate vein (LIV, RIV). (B) Apical four-chamber view demonstrating vertical vein (VV) and pulmonary venous confluence (PVC). (C) Right and left pulmonary veins were seen to drain into the pulmonary venous confluence, which was located behind the left atrium region and opened to large left innominate vein through the vertical vein. *Supplementary video files associated with this presentation can be found in the online version of the journal.

