

## Noninvasive diagnosis and surgical management in total anomalous pulmonary venous return draining into the coronary sinus: Report of 2 cases

Vedide Tavlı<sup>1</sup>, Bekir Kayhan<sup>2</sup>, F. Fevzi Okur<sup>2</sup>, Mustafa Kırman<sup>2</sup>, Mehmet Tekdoğan<sup>2</sup>

Unit of <sup>1</sup>Pediatric Cardiology and <sup>2</sup>Cardiovascular Surgery, Şifa Medical Center, İzmir, Turkey

**SUMMARY:** Tavlı V, Kayhan B, Okur FF, Kırman M, Tekdoğan M. Noninvasive diagnosis and surgical management in total anomalous pulmonary venous return draining into the coronary sinus: report of 2 cases. Turk J Pediatr 2001; 43: 91-93.

Total anomalous pulmonary venous return results from nondevelopment of the common pulmonary vein, with consequent enlargement of embryonic collaterals between the lungs and the systemic veins. In this report, two patients with this anomaly draining into the coronary sinus who presented in infancy are described. One of the patients was referred because of growth and development failure and chronic constipation, while the other had tachypnea as the presenting problem. Both were diagnosed during the echocardiographic examination. Typical echocardiographic findings were a small left atrium, a sausage-shaped dilated coronary sinus receiving the pulmonary veins in the subcostal short axis, and flying seagull configuration in the subcostal long axis views. Both had a large interatrial communication. The patients underwent corrective surgery. The aim of the presentation is to emphasize the role of segmental echocardiography in the differential diagnosis.

**Key words:** total anomalous pulmonary venous return, echocardiography.

Total anomalous pulmonary venous return (TAPVR) results from nondevelopment of the common pulmonary vein, with consequent enlargement of embryonic collaterals between the lungs and the systemic veins. It comprises approximately one to two percent of all congenital heart diseases<sup>1,2</sup>. An obligatory right to left shunt either through the atrial septal defect or patent foramen ovale is usually present. Presence of an obstructive lesion in the anomalous pulmonary venous channel profoundly influences the hemodynamic state and clinical features in a case of TAPVR. The aim of the presentation was to emphasize the role of segmental echocardiography in the differential diagnosis and management of TAPVR draining into the coronary sinus<sup>3-6</sup>.

### Case Reports

#### Case 1

A 15-month-old boy (weight: 8 kg) was referred to our hospital because of growth and development failure and chronic constipation.

A grade 3/6 systolic murmur was heard best along the left third intercostal space. The second heart sound was loud and fixed split. He also had coloboma of the iris, right-sided preauricular skin tag and frontal bossing. There was cardiomegaly and prominent pulmonary trunk on his chest X-ray. Right ventricular hypertrophy was present on his echocardiography (ECG). On his 2-D echocardiography, the left atrium (LA) and left ventricle (LV) were small. The pulmonary veins were seen to collect in a common chamber (PVC) unrelated to the LA (Fig. 1). There was a 12 mm secundum type atrial septal defect (ASD). Posterior angulation in the interrogation of the four-chamber view revealed a very enlarged coronary sinus (Fig. 2). Subcostal short axis view revealed a sausage-shaped pulmonary venous trunk draining into the right atrium (RA) via the coronary sinus, which should be considered typical for this type of TAPVR (Fig. 3). Hemodynamic assessment revealed mildly elevated pulmonary artery pressure.

Pulmonary artery angiogram revealed abnormal pulmonary venous return into the RA through the enlarged coronary sinus. The LV was small. The patient was scheduled urgently for cardiac surgery. Under cardiopulmonary by pass, the repair was done from inside the RA. The coronary sinus was incised into the LA, and the entire area was covered with a pericardial patch. Both pulmonary venous return and coronary sinus blood flow were directed into the LA, creating a small obligatory right to left shunt that was well tolerated. The patient was extubated the next day and remained in the intensive care unit for one day. He was discharged within six days. He has remained well for a follow-up period of two years and markedly gained weight and height. On his latest chest X-ray, the border of the RA was less prominent and the cardiothoracic ratio within normal limits. Post-operative echocardiography revealed a larger LV volume and appropriate drainage of the pulmonary veins into the LA.

#### Case 2

A two-month-old girl presented with tachypnea. On her examination there was a 2-3/6 grade systolic murmur on the third left intercostal space. Her echocardiography revealed a large ASD, small LA and LV, and an enlarged coronary sinus receiving the pulmonary venous return which resembled a flying seagull (Fig. 4). Selective left and right pulmonary angiograms were performed. Left and right sided pulmonary venous return was to the RA via the coronary sinus. She was operated at another institution and has remained healthy.



Fig. 1. The pulmonary veins are seen to collect in a pulmonary venous trunk posterior to the small left atrium (LA) in Case 1 (arrows). ra: right atrium; rv: right ventricle; lv: left ventricle.

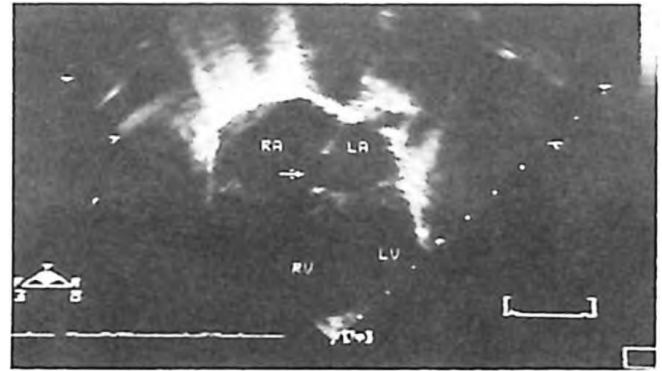


Fig. 2. An enlarged coronary sinus is seen in the posterior angulation of the four-chamber view in Case 1.



Fig. 3. Subcostal short axis view in Case 1 showing typical sausage-shaped dilated coronary sinus.



Fig. 4. Subcostal four-chamber view in Case 2. The flying seagull configuration is seen, with the wings being the pulmonary veins and the remainder being the enlarged coronary sinus receiving the pulmonary veins.

## Discussion

In the classification by Darling and co-workers<sup>2</sup>, TAPVR anomalies are divided into four types: supracardiac (45%), cardiac (25%), infracardiac (25%) and mixed (5%)<sup>3</sup>. The frequency of TAPVR drainage into the coronary sinus was 18 percent. TAPVR can also be classified by the presence or absence of venous obstruction<sup>1-3,7</sup>. Although obstruction is very common in the infracardiac types, it is less likely to be seen in the other types. The treatment of TAPVR is surgical drainage<sup>7,8</sup>. The timing of surgery should be determined by the presence or absence of pulmonary venous obstruction. Because there is no possibility of spontaneous resolution of obstructed TAPVR, the diagnosis alone is an indication for surgery. Early mortality during the first year of life has been 8-11 percent; late mortality was reported as 2-3 percent<sup>7</sup>. Both of the cases were operated once the diagnoses were made with very good early and short-term outcomes.

Segmental echocardiography performed in the subcostal long and short axes, and apical four-chamber and parasternal views with the aid of Doppler echocardiography should allow for the accurate noninvasive diagnosis of the infracardiac form of TAPVR<sup>4-6</sup>. Ostium primum type ASD should be differentiated from an enlarged coronary sinus, which is best interrogated in the posterior angulation of the four-chamber view. A small left atrium with the pulmonary veins draining into the typical sausage-shaped dilated coronary sinus is readily identified, particularly in the subcostal short axis view (Fig. 3). Slightly posterior and oblique angulation of the transducer in the acquisition of the subcostal four-chamber view

may reveal a typical flying seagull configuration with the wings being the pulmonary veins and the remainder being the enlarged coronary sinus receiving the pulmonary veins (Fig. 4).

In conclusion, following the course of the pulmonary veins through orthogonal planes and the appearance of a relatively small LA and LV should alert the physician for the definitive noninvasive diagnosis of TAPVR to the coronary sinus.

## REFERENCES

1. Delisle G, Ando M, Caldre AL, et al. Total anomalous pulmonary venous drainage to the coronary sinus. *Am Heart J* 1976; 91: 99-122.
2. Darling RC, Rothney WB, Craig DM. Total anomalous pulmonary venous drainage into the right side of the heart. *Lab Invest* 1978; 44: 1854-1859.
3. Karademir S, Bilgiç A, Özkutlu S, Öztunç F. Portal sisteme drenaj gösteren total anormal pulmoner venöz dönüş anomalisi. *Türk Kardiyol Dem Arş* 1990; 18: 214-216.
4. Pickhoff AS, Sequeria R, Ferrer PL, et al. Pulsed Doppler echocardiographic findings in total anomalous pulmonary venous drainage to the coronary sinus. *Cathet Cardiovasc Diagn* 1980; 6: 247-254.
5. Snider AR, Ports TA, Silverman NH. Venous anomalies of the coronary sinus: detection by M-mode, two-dimensional and contrast echocardiography. *Circulation* 1979; 60: 721-727.
6. Orsmod GS, Ruttenberg HS, Bessinger FB. Echocardiographic features of total anomalous pulmonary venous connection to the coronary sinus. *Am J Cardiol* 1978; 41: 597-601.
7. Stark J. Anomalous pulmonary venous return and cor triatriatum. In: Stark J, De Leval M (eds). *Surgery for Congenital Heart Defects* (3<sup>rd</sup> ed). Philadelphia: WB Saunders; 1994: 319-327.
8. Kınoğlu B, Sanoğlu T, Türkoğlu H, et al. Total anormal pulmoner venöz dönüşlerde ameliyat sonrası erken ve geç dönem sonuçlar. *Türk Kardiyol Dem Arş* 1996; 24: 366-370.