The Turkish Journal of Pediatrics 34:175-178, 1992

CONGENITAL CORONARY ARTERIOVENOUS FISTULA IN A TEN-YEAR-OLD BOY WITH ANGINA PECTORIS*

*Talat Cantez MD***, Ümrah Aydoğan MD***, Enver Dayıoğlu MD****
*Ertan Onursal MD*****, Ilmay Bilge MD******

SUMMARY: Cantez T, Aydoğan Ü, Dayıoğlu E, Onursal E, Bilge I. (Departments of Pediatrics and Cardiovascular and Thoracic Surgery, İstanbul University Faculty of Medicine, İstanbul, Turkey). Congenital coronary arteriovenous fistula in a ten-year-old boy with angina pectoris. Turk J Pediatr 34: 175-178, 1992.

Congenital arteriovenous fistula (CAVF) is a rare cardiac lesion. Angina pectoris is uncommon in younger patients with CAVF. Fistula-related symptoms, complications of this anomaly and surgical complications have a strong correlation with the age of the patient. A ten-yer-old male patient with angina pectoris in whom the diagnosis of CAVF was established, and who, following surgical ligation recovered, is presented.
Key words: coronary artery anomalies, coronary arteriovenous fistula.

Congenital coronary arteriovenous fistula (CAVF) is one of the rare congenital heart lesions. The term CAVF describes the fistulous communications between branches of the coronary arteries and a large vessel or cardiac chamber. According to Wenger's estimate[1], the incidence of CAVF is 1 per 6,250,000. Dyspnea on exertion, fatigue and angina are the major symptoms of this anomaly. However, only nine percent of patients under the age of 20 are symptomatic[2].

In this report, we describe a ten-year-old boy with CAVF who presented with all the known symptoms associated with this anomaly.

Case Report

A ten-year-old boy was admitted to the İstanbul University Faculty of Medicine Children's Hospital with a history of dyspnea, angina on exertion, and fatigue of four years' duration.

Physical examination revealed a weight of 31.5 kg (25th-50th percentile), and height of 1.34 m (25th-50th percentile). The heart rate was 72 beats/min, with a

* From the Departments of Pediatrics and Cardiovascular and Thoracic Surgery, İstanbul University Faculty of Medicine, İstanbul.
** Professor of Pediatrics and Pediatric Cardiologist, İstanbul University Faculty of Medicine.
*** Chief Resident in Pediatrics, İstanbul University Faculty of Medicine.
**** Associate Professor of Cardiovascular and Thoracic Surgery, İstanbul University Faculty of Medicine.
***** Professor of Cardiovascular and Thoracic Surgery, İstanbul University Faculty of Medicine.
****** Resident in Pediatrics, İstanbul University Faculty of Medicine.

sinusal rhythm. The blood pressure was 110/70 mmHg. Radial and femoral pulsations were normal on palpation. Cardiac examination revealed that S_1 and S_2 were single and not accentuated. A continuous murmur was audible at the 3^{rd}-4^{th} left intercostal margin. No other abnormalities were noted.

The results of laboratory studies were within normal limits. The teleroentgenogram demonstrated that the heart was of normal size, but that the apex was slightly upturned and pulmonary vascular tracings were slightly elevated. The electrocardiogram showed no anomalies. The electrocardiogram stress test was also within normal limits. M-mode and two-dimensional echocardiography showed normal findings. Color Doppler echocardiography revealed a blood flow from the right ventricular apex into the cavity (Fig. 1). Based on these findings, a tentative diagnosis of CAVF of the right coronary artery into the right ventricle was established. Hemodynamic examination showed that the catheter passed into the left atrium through the foramen ovale. O_2 saturation in the right atrium was 79 percent, while it was 83 percent in the right ventricle, showing a four percent increment. Systolic, diastolic and end-diastolic pressures in the right ventricle were within normal limits. The Qp/Qs ratio was 1.22 and the L-R shunt was calculated as 1.38 L/min. Selective coronary artery injections showed no anomaly in the left coronary artery, but the right coronary artery showed an aneurysmal dilatation from the aortic end to the apex of the heart, where the fistula entered

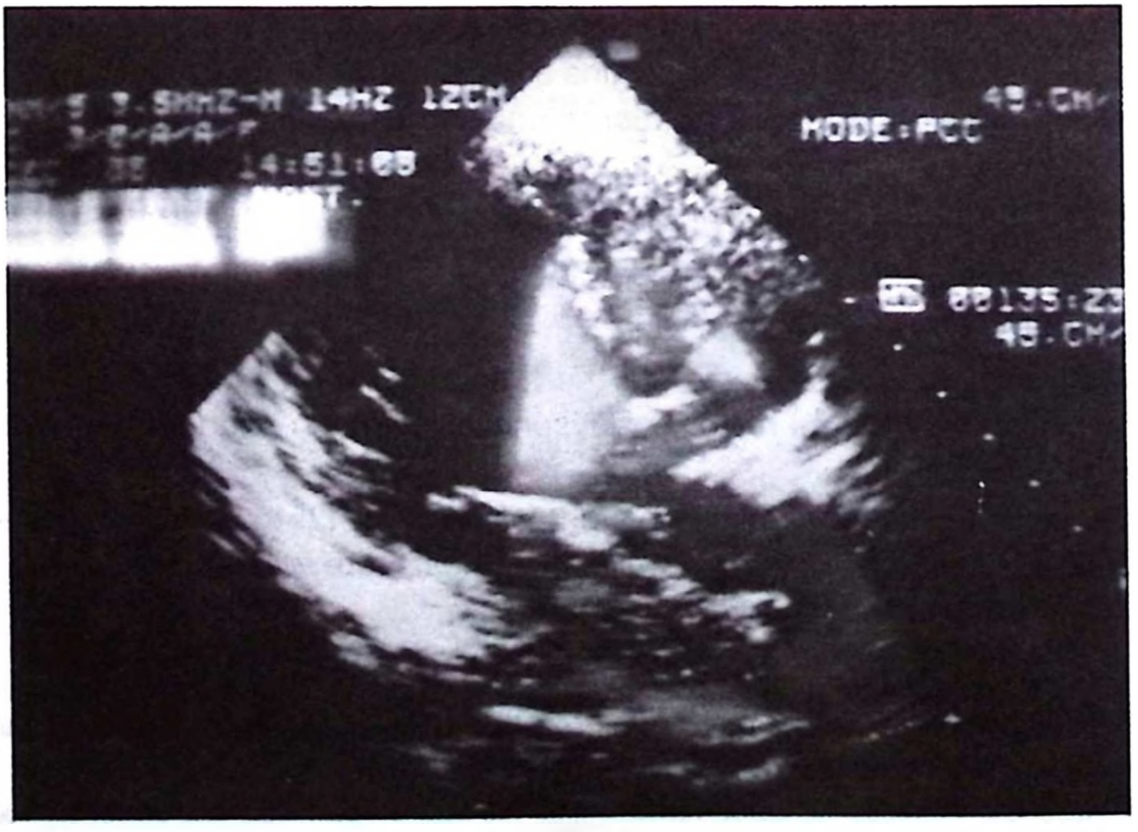

Fig. 1: Turbulent flow from the apex of the RV on color Doppler echocardiography.

the right ventricle (Fig. 2). The distal end of the coronary artery was too puny. Cardiac cineangiography demonstrated no other anomaly. The fistulous communication was surgically ligated, resulting in the disappearance of all complaints.

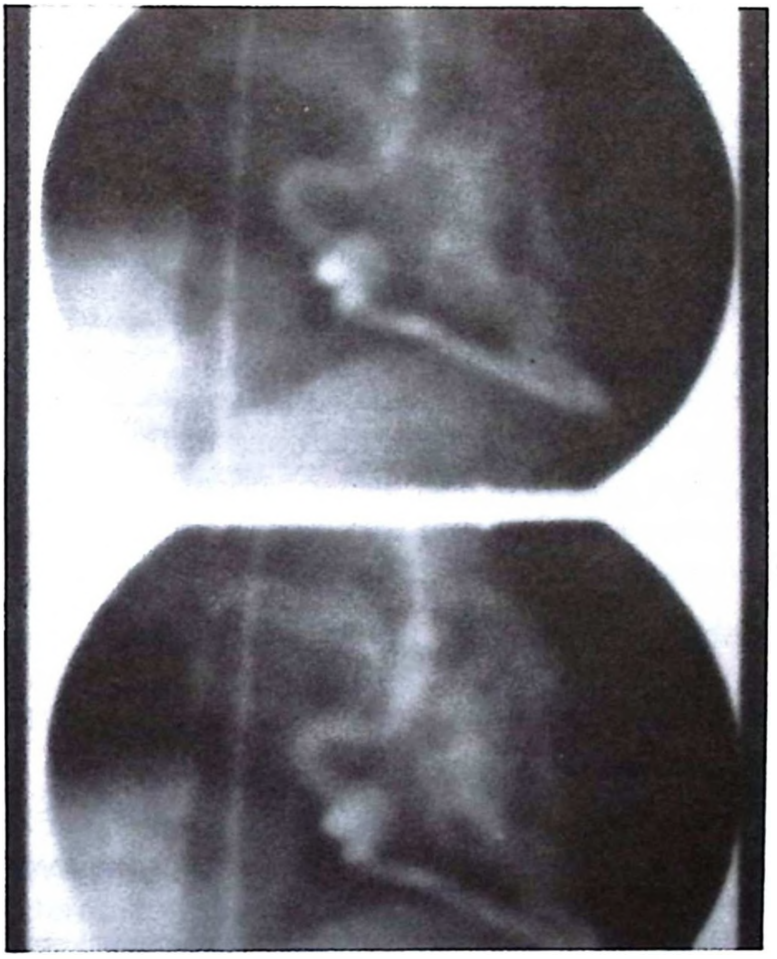

Fig. 2: Dilated right coronary artery and visualization of the right ventricular cavity by the epical fistula.

Discussion

Since its initial description in 1865, about 350 patients with CAVF have been reported[2-9]. Embryologically, these fistulas seem to represent persistent junctions of the primordial sinusoidal circulation. They vary from a single vessel, frequently tortuous, with a single origin and a single termination, as was the situation in this patient, to complex, worm-like aneurysmal cavities in which blood may stagnate, clot and calcify[10]. Aneurysmal dilatation results from a structural weakening of the vessel wall due to increased blood flow through the fistula[11].

More than half of CAVF patients of all ages are clinically asymptomatic. They show normal stress tolerance despite their moderate volume overloads. According to some authors, the mean shunt does not differ significantly from

those with or without symptoms or complications, especially in patients under the age of twenty[2,9]. Liberthson et al[2], in their review of 186 patients reported in the literature, grouped the patients according to age. Among the 88 patients over the age of twenty, 67 percent had preoperative symptoms of CAVF-related complications such as congestive heart failure, subacute bacterial endocarditis, myocardial infarction, fistula rupture or a fatal outcome. In contrast, the frequency of symptoms and CAVF-related complications in the younger group was only 19 percent. Only three percent of the younger patients had symptoms peculiar to angina pectoris. The frequency of angina pectoris in younger patients is also uncommon in later reports[4,7-9]. In these patients, angina pectoris is attributed to insufficient blood flow to the distal end of the fistula (coronary steal)[10].

In view of the fact that there is a high incidence of fistula-related late symptoms/complications, increased morbidity and mortality associated with surgical intervention in older patients and the low incidence of spontaneous closure, fistula ligation was attempted in this patient. The fistulous communication was ligated by two transfixion sutures of nonabsorbable material, without resorting to cardiopulmonary bypass. Shortly after the operation, the patient's complaints disappeared, and no complications related to the surgery occurred.

REFERENCES

1. Wenger NK. Rare causes of coronary artery disease. In Hurst JW (ed). *The Heart* (4th ed). New York: McGraw, 1978, pp 1348-1349.
2. Liberthson RR, Sagar K, Berkoben JP, et al. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. *Circulation* 59: 849, 1979.
3. Kirklin JW. Congenital anomalies of the coronary arteries. In Kirklin JW (ed). *Cardiac Surgery*. Willey Medical Publication, 1986, pp 945-955.
4. Aytaç A, Bilgiç A, Olga R, et al. Congenital right coronary artery-right atrial fistula. *Jpn Heart J* 21: 409, 1980.
5. Tayyareci G, Kalangoz D, Çağıl A, et al. Koroner arter fistülü. *Türk Kardiol. Der. Ar.* 18: 209, 1990.
6. Okay T, Özdemir M, Sancaktar O. Bilateral koroner arter-pulmoner arter fistülü olgusu. *Türk Kardiol. Der. Ar.* 17: 104, 1989.
7. Özer S, Saraçlar M, Yurdakul Y. Duktus arteriosus açıklığı ile birlikte sol koroner arterin sol atriyuma fistülü. *Türk Kardiol. Der. Ar.* 17: 43, 1989.
8. Özer S, Özkutlu S, Sarıoğlu T, et al. Konjenital koroner arter fistülleri. *Çocuk Sağlığı ve Hastalıkları Dergisi* 28: 135, 1985.
9. Urrutia-S CO, Falaschi G, Ott DA, Cooley DA. Surgical management of 56 patients with congenital coronary artery fistulas. *Ann Thorac Surg* 35: 300, 1983.
10. Lurie PR. Anomalies of the coronary arteries. In Anderson RH, Macartney FJ, Shinebourne EA, Tynan M (eds). *Padiatric Cardiology*, Vol. 2. Edinburgh: Churchill, 1987, pp 1073-1085.
11. Rittenhouse EA, Doty DB, Ehrenhaft JL. Congenital coronary artery-cardiac chamber fistula. Review of operative management. *Ann Thorac Surg* 20: 468, 1975.