

TWO-DIMENSIONAL AND DOPPLER ECHOCARDIOGRAPHIC FINDINGS OF CONGENITAL CORONARY ARTERY FISTULA*

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SUMMARY: Özkutlu S, Saraçlar M, Atalay S, Bilgiç A, Yurdakul Y, Çiçek S. (Department of Pediatric Cardiology, Hacettepe University Institute of Child Health, Ankara, Turkey). Two-dimensional and Doppler echocardiographic findings of congenital coronary artery fistula. Turk J Pediatr 34: 167-174, 1992.

Three patients with coronary artery fistula confirmed by coronary angiography and surgery were studied using two-dimensional and Doppler echocardiography. Two of these patients had left coronary artery-right ventricle fistula. The proximal dilatation of the coronary arteries was visualized in all three patients by two-dimensional echocardiography. The course of the dilated right coronary artery and left circumflex artery was demonstrated by two-dimensional echocardiography in two patients. We were able to detect the disturbed flow at the drainage site in one patient by using the conventional Doppler echocardiography.

In conclusion, it should be emphasized that two-dimensional and Doppler echocardiography can be useful methods in diagnosing coronary artery fistula. *Key words:* congenital coronary artery fistula, echocardiography.

Congenital coronary artery fistula is relatively uncommon and generally occurs as an isolated anomaly¹⁻³. The diagnosis of coronary artery fistula is established by cardiac catheterization and selective coronary angiography^{1,4}. More recently, advances in two-dimensional echocardiographic techniques have aided in identifying a dilated involved coronary artery. It is possible, however, to obtain additional information by employing pulsed Doppler echocardiography for detecting shunt flow in selected patients with coronary artery fistula⁵⁻¹¹.

We present three patients with coronary artery fistula diagnosed by two-dimensional and Doppler echocardiography which was also confirmed by angiocardiography. The diagnosis of two of these patients was also substantiated by surgery.

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Material and Methods

Of the patients referred to the Department of Pediatric Cardiology of the Hacettepe University Institute of Child Health between 1983-1990, a diagnosis of congenital coronary artery fistula was established in three using echocardiography, cardiac catheterization and selective coronary angiography (Fig. 1).

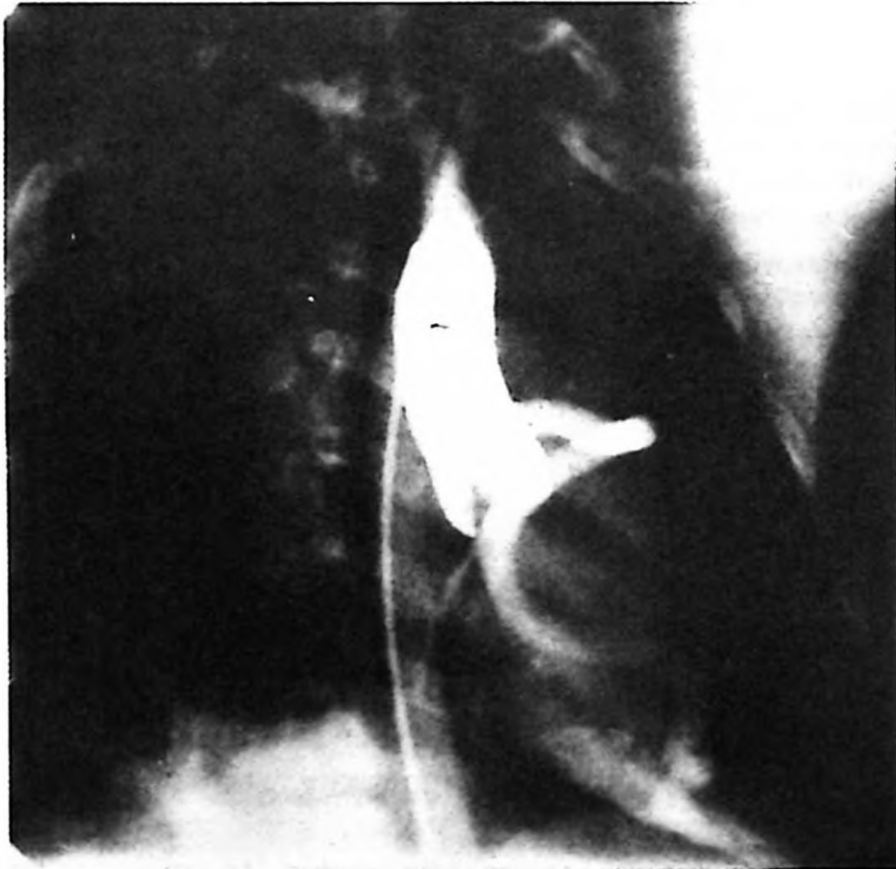


Fig. 1: Aortic root injection in the right anterior oblique position. Drainage of the left circumflex coronary artery into the right ventricular apex is seen (Case No 3).

This study was carried out between 1983 and 1990. The ages of the patients ranged between 3-5 years. Two patients had undergone surgical closure of the fistula.

Echocardiography: The echocardiographic examination included two-dimensional and Doppler studies using a Toshiba-Sonolayer-S SSH 60 A scanner with 2.5 and 3.75 MHz transducers.

Case Reports

The clinical manifestations and laboratory findings of our patients are presented in Table I. Echo findings were as follows:

TABLE I: Clinical and Laboratory Findings of the Patients

Caso No	Age (yr) sex	Murmur	Cardiac catheterization	Angiocardiology	Treatment and outcome
1 (1983)	5, F	3 rd intercostal space continuous murmur with diastolic accentuation	RVP : 35 mm Hg PAP : 25/10 mm Hg mean : 12 mm Hg Qp/Qs: 1.2	Left coronary artery- right ventricular outflow tract fistula	Surgical therapy recommended
2 (1987)	4, F	4 th intercostal space continuous murmur with diastolic accentuation	RVP : 27 mm Hg PAP : 21/7 mm Hg mean : 12 mm Hg Qp/Qs: 1.4	Right coronary artery- right ventricular outflow tract fistula	Fistula closed with patch
3 (1990)	3, F	Continuous murmur with diastolic accentuation at the apex	RVP : 30 mm Hg PAP : 20/5 mm Hg mean : 14 mm Hg There was no shunt	Left coronary artery- right ventricular apex fistula	Fistula closed with patch

RVP: right ventricular pressure

PAP: pulmonary artery pressure

Qp : pulmonary flow

Qs : systemic flow

Case 1

Two-dimensional echocardiography showed that the dimensions of the cardiac chambers were within normal limits. In the high parasternal short axis records, dilatation of the proximal portion of the left coronary artery was seen (Fig. 2). The diameter of the coronary artery on this side was 9 mm and the aortic root 24 mm. The ratio of the coronary artery diameter to the aortic root diameter was 0.37.



Fig. 2: In the parasternal short-axis view, proximal dilatation of the left coronary artery is seen (arrows).
Ao: aorta

Case 2

Echocardiographic examination revealed slight right atrial and right ventricular dilatation. Left ventricular functions were normal. The high parasternal short-axis records demonstrated dilatation of the proximal portion of the right coronary artery, which was 9 mm in diameter (Fig. 3). The aortic root was 21 mm and the ratio of the coronary artery diameter to the aortic root was 0.42. The dilated right coronary artery was demonstrated on the anterior groove of the right ventricle in the subcostal position.

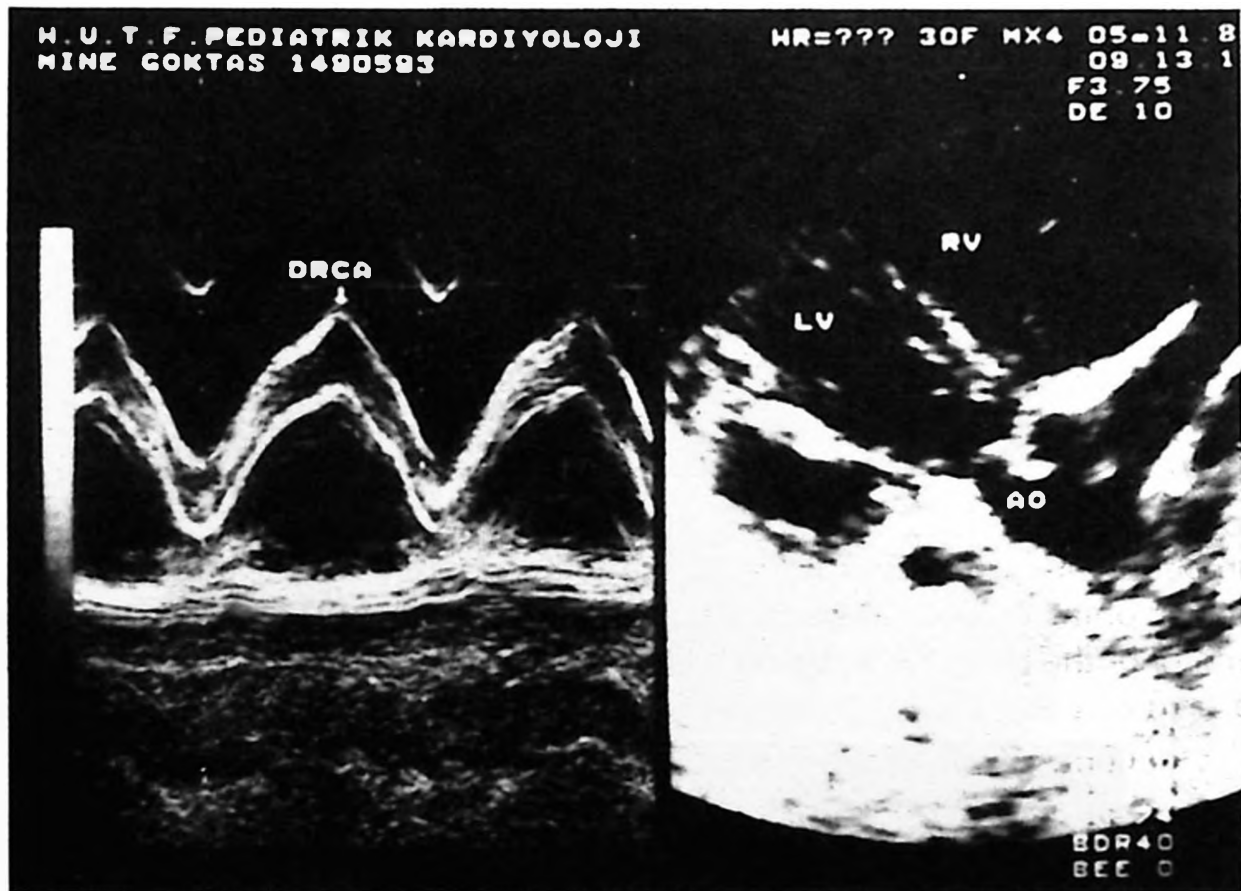


Fig. 3: Dilated right coronary artery (arrow) is demonstrated by two-dimensional (right) and M-mode echocardiography (left).

Case 3

Echocardiographic studies revealed that the diameters of the cardiac chambers and left ventricular functions were within normal limits. The proximal dilatation of the left coronary artery was demonstrated in the high parasternal short axis view, which was 7 mm in diameter. The ratio of the left coronary artery diameter to the aortic root diameter was 0.41.

The complete course of the circumflex coronary artery was visualized on the right ventricle from the apical four-chamber view (Fig. 4).

Doppler echocardiography revealed a continuous disturbed flow with diastolic accentuation in the right ventricular cavity near the apex (Fig. 5).

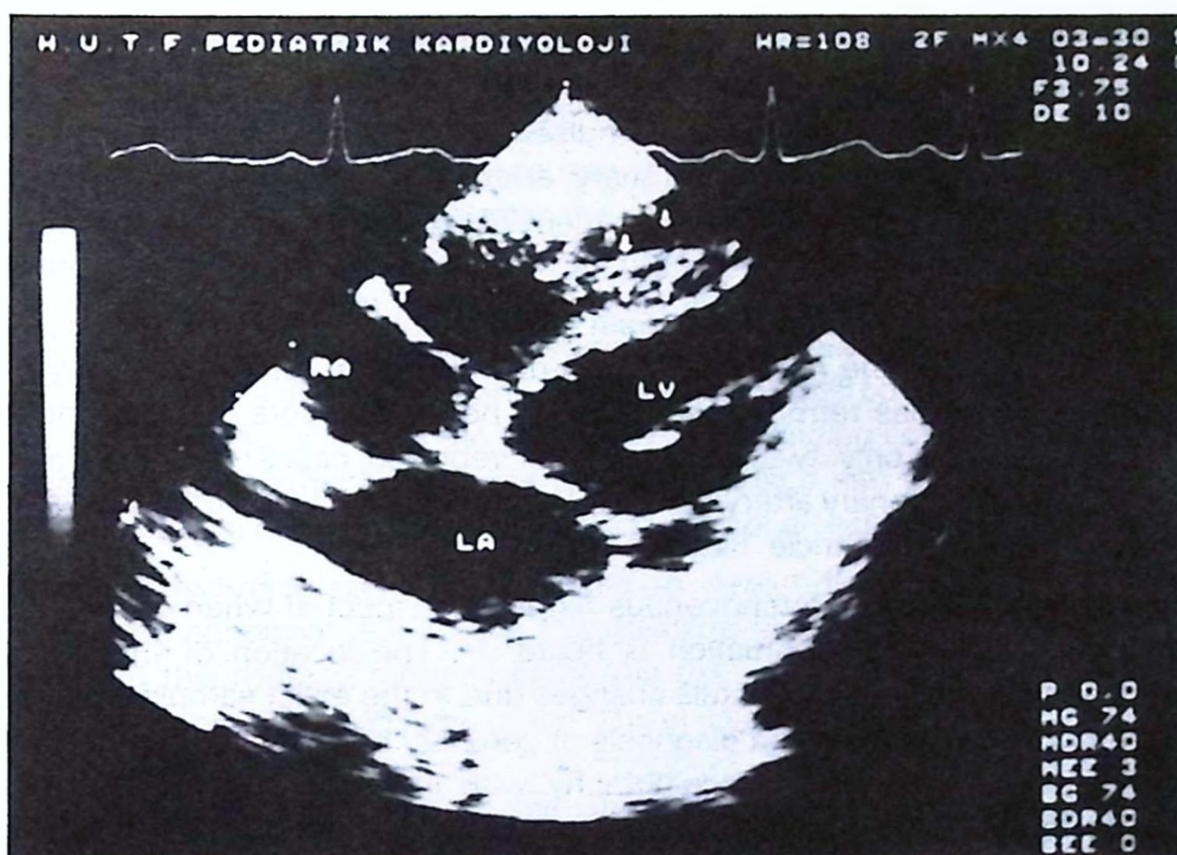


Fig. 4: Complete course of the left circumflex coronary artery (arrows) is demonstrated.

RA: right atrium
T : tricuspid valve

LA: left atrium
LV: left ventricle

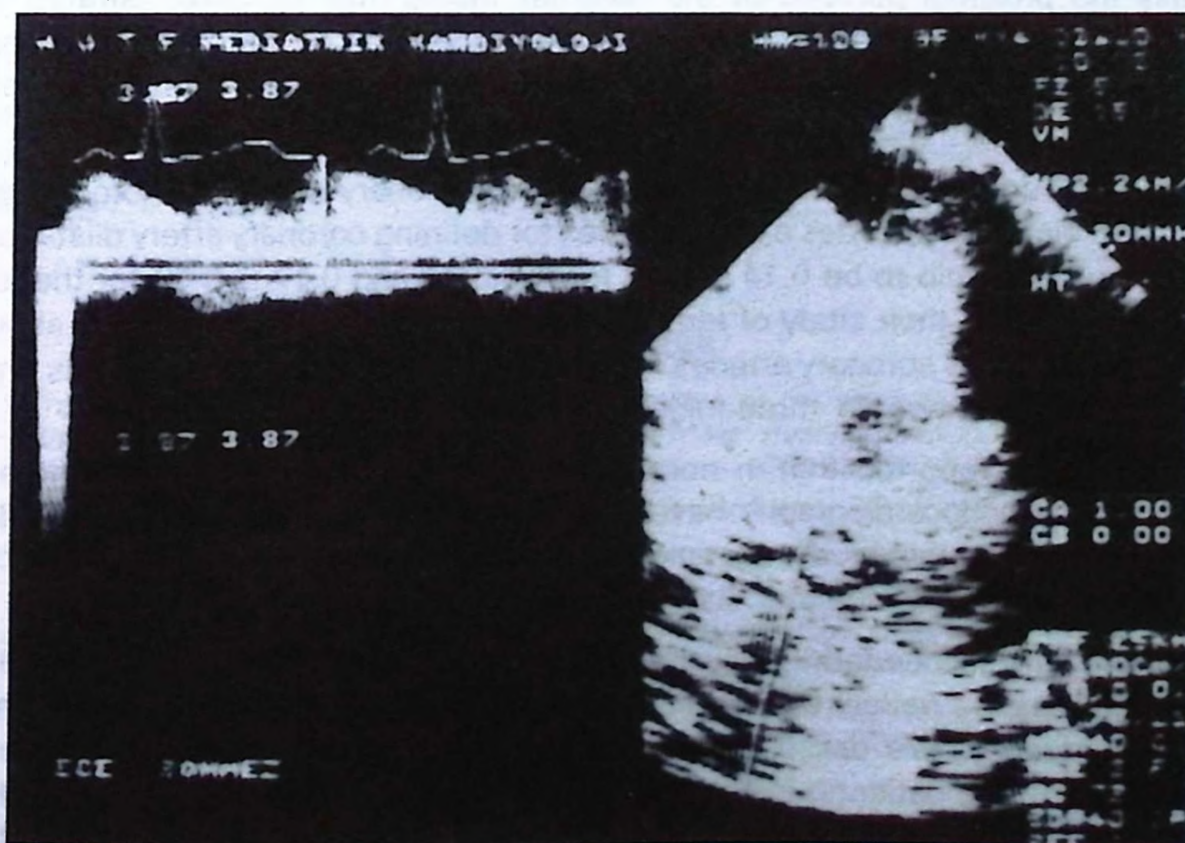


Fig. 5: Continuous disturbed flow with diastolic accentuation (left) demonstrated near apex by Doppler echocardiography.

Discussion

The incidence of coronary artery fistula varies between 0.26 and 0.40 percent in selected case series of congenital heart disease⁹. Coronary artery fistulas more commonly arise from the right coronary artery (50-60%) than from the left (30-40%)^{1,12,13}. It is reported that 90 percent of coronary artery fistulas drain to the venous site of the heart¹⁻³.

Thirty-nine percent of these fistulas drain into the right ventricle and 33 percent into the right atrium. The remainder (20%) drain into the pulmonary artery¹⁴⁻¹⁶. Coronary artery fistulas terminating in the left heart chambers are exceptionally rare, and constitute only two percent of all reported cases^{12,17}. Two of our patients had a left coronary artery-right ventricle fistula, while the other had a right coronary artery-right ventricle fistula.

The diagnosis of coronary arteriovenous fistula is suspected when a continuous murmur with diastolic accentuation is heard^{1,12}. The location of the murmur caused by the coronary artery fistula changes due to the exact sampling site with the chamber of drainage¹². The diagnosis of coronary artery fistula is established by cardiac catheterization and angiography with injection of contrast material either into the aortic root or selectively into the coronary arteries^{1,4,6,18,19}. More recently, diagnostic information was obtained by two-dimensional, conventional and color Doppler echocardiography⁵⁻¹¹.

Usually the proximal portions of the coronary artery may be demonstrated by two-dimensional echocardiography^{5-7,9-11}. Demonstration of the dilated coronary artery is much easier. It is necessary to know the normal ranges of coronary artery diameters in order to evaluate the coronary arteries^{5,6}.

Velvis et al⁶ demonstrated that the ratio of the coronary artery diameter to the aortic root diameter provides a reliable index for defining coronary artery dilatation. They found this ratio to be 0.14 ± 0.03 for the right and 0.17 ± 0.03 for the left coronary artery. In their study of ten patients, this ratio was 1.5 to 4 times above normal for proximal coronary arteries feeding the fistula⁶. In our patients, this ratio was found to be two to three-fold above normal.

Shakudo et al⁵ reported that in normal populations, coronary artery diameters measured by echocardiography have a diameter ranging between 3.5 and 6.0 mm. According to this study, dilatation of the coronary artery was considered to be present when the luminal diameter was greater than 6 mm⁵.

Rodgers et al⁸ succeeded in visualizing the proximal and distal portions of the coronary artery in a patient with coronary arteriovenous fistula by two-dimensional echocardiography. We demonstrated proximal dilatation of the coronary artery fistula in our three patients. In the study of Velvis et al⁶ proximal dilatation of the coronary artery fistula was seen in ten patients, as was observed in our patients.

They visualized the orifice of the coronary sinus or the right ventricle in five of their patients in whom the left circumflex coronary artery was the feeding vessel and the fistula drained into the right atrium. We visualized the proximal dilatation of the left coronary artery and complete course of the left circumflex artery in Case No 3. They did not demonstrate whether the coronary artery fistula drained from the posterior atrioventricular groove into the right atrium on the right ventricle in three of their patients in whom the right coronary artery was the feeding vessel. In our second case, the course of the dilated right coronary artery was demonstrated by two-dimensional echocardiography, but its drainage site could not be visualized. In another two cases, in which the left anterior descending coronary artery was the feeding vessel, neither the course nor the drainage site of the coronary artery fistula could be demonstrated by two-dimensional echocardiography.

Doppler echocardiography can be used to detect flow velocities in fistulous vessels and abnormal flows from the drainage site in cases of coronary artery fistula, but this method requires extensive experience, patience and skill^{5,6,8,11}. Velvis et al⁶ demonstrated a disturbed flow in dilated coronary arteries feeding the fistula in eight patients by color flow mapping. In our study, we were able to detect the disturbed flow at the drainage site in Case No 3 by using conventional Doppler echocardiography. Velvis et al⁶ and Fukuda et al¹¹ reported that this disturbed flow had occurred primarily in late systole and early diastole. We recorded a continuous disturbed flow with diastolic accentuation in Case No 3, as was reported by Kronzon et al¹⁰.

This report, along with others, demonstrates that two-dimensional echocardiography combined with Doppler echocardiography are non-invasive methods which can be employed for evaluating a coronary artery fistula and identifying its drainage site.

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