Two-dimensional and Doppler Echocardiographic Features of Left Circumflex Coronary Artery to Right Ventricle Fistula: Case Report and Literature Review

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SUMMARY. We report the case of a seven-month-old infant with a clinical diagnosis of patent ductus arteriosus whose two-dimensional and Doppler echocardiographic examinations were consistent with a coronary artery fistula. At angiography, a left circumflex coronary artery to right ventricle fistula was diagnosed. Echo-Doppler techniques for the noninvasive diagnosis of this lesion are discussed and the literature is reviewed.

KEY WORDS: Coronary artery fistula — Doppler ultrasound — Two-dimensional echocardiography

Congenital coronary artery fistula is a rare defect reported to occur in one in 50,000 patients with congenital heart disease and 1 in 500 patients undergoing coronary arteriography [19]. In the presence of a significant shunt, the physical examination may be nearly identical to that of a patient with a patent ductus arteriosus except for subtle differences in the location of the murmur [4]. Visualization of coronary arteries by two-dimensional echocardiography coupled with additional information obtained from the Doppler examination provides an excellent technique for the noninvasive diagnosis of coronary artery fistula. We report the two-dimensional echocardiographic and Doppler ultrasound findings in an infant with a left circumflex coronary artery to right ventricle fistula.

Case Report

A 2.6-kg baby boy was born following an uncomplicated term pregnancy. A murmur was first heard at six months of age. The clinical diagnosis of a patent ductus arteriosus was made and cardiac catheterization was recommended. The child was first seen in our cardiology outpatient clinic at seven months of age because the parents sought a second opinion on the diagnosis and recommendation for cardiac catheterization. Review of the past medical history showed normal growth and development and an absence of cardiovascular symptoms. There was no family history of congenital heart disease.

On physical examination, the patient’s length was 70.5 cm (50%-75%), his weight was 8.16 kg (50%), and his head circumference 45.5 cm (75%). The heart rate was 152/min and regular, respiratory rate 36/min, blood pressure in the right arm 108/50 and in the right leg 110 systolic. He was acyanotic. Other pertinent cardiovascular findings included mildly hyperactive peripheral pulses, absence of a palpable thrill, and mildly hyperactive apical impulse. There was a physiologically split second heart sound with a normal pulmonic closure sound and a prominent apical third heart sound. A grade-3/6 continuous machinery murmur was well heard at the third left intercostal space and left upper sternal border but was not well heard over the back. A grade-2/4 middiastolic rumble was heard at the apex. The lungs were clear to auscultation and the liver edge was palpable 2 cm below the right costal margin.

Noninvasive studies done at the initial clinic visit included a transcutaneous $\text{Po}_2$ measurement of 79 mmHg. A chest x-ray film showed moderate cardiomegaly with a mild increase in pulmonary arteriolar markings and an enlarged thymus. The electrocardiogram showed sinus tachycardia at a rate of 150/min, a QRS axis of +60°, probable left atrial enlargement, and left ventricular hypertrophy of the volume overload type.

We advised the parents that we agreed with the diagnosis, and the infant was admitted for surgery. Preoperative echocardiographic evaluation showed left atrial and left ventricular volume overload. A patent ductus arteriosus could not be imaged directly or proven to exist by Doppler examination. An area of echocardiographic dropout was imaged in the apical portion of the ventricular septum in several views and was interpreted as representing a muscular ventricular septal defect (Fig. 1). The Doppler exam showed disturbed systolic flow in the apex of the
right ventricle. Due to the discrepancy between clinical and echocardiographic diagnoses, a cardiac catheterization was performed. There was an increase in oxygen saturation in the right heart from 71% in the superior vena cava to 83% in the left pulmonary artery. Left heart oxygen saturations were normal. Right and left heart pressures were normal. The pulmonary-systemic flow ratio was 2.0:1. A left ventricular cineangiogram showed massively enlarged left main and left circumflex coronary arteries. The left circumflex coronary artery penetrated the cardiac apex and the apical portion of the ventricular septum, thus communicating with the right ventricle (Fig. 2). No ventricular septal defect or patent ductus arteriosus was present. Because of the catheterization findings, a repeat echocardiographic examination was obtained at the time of catheterization. The enlarged left main and circumflex coronary arteries were well seen in the subcostal four-chamber view on the surface of the left ventricle (Fig. 3). The area of echocardiographic dropout, which was interpreted as being a muscular ventricular septal defect, was confirmed by cineangiography and aortic root saline contrast injections to be the point of entry of the fistula through the ventricular septum into the right ventricle. A Doppler recording obtained just opposite the site of entry of the coronary artery fistula into the right ventricle showed disturbed flow beginning in mid-systole and extending into early diastole (Fig. 4). Doppler examination of the descending thoracic aorta showed normal laminar systolic flow and no significant diastolic flow. Doppler examination of the ascending aorta showed retrograde diastolic flow (Fig. 5). Doppler examination of the left ventricular outflow showed no evidence of aortic insufficiency.

The child was taken to surgery where the fistula was closed at its entry into the myocardium on the diaphragmatic surface of the right ventricle using three pledgeted horizontal mattress sutures and proximal and distal ligatures. Postoperatively, the pul-