Assessment of Congenital Coronary Artery Fistulas by Transesophageal Color Doppler Echocardiography

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PURPOSE: Coronary angiography is the gold standard for imaging the coronary tree, but the relation of coronary artery fistulas to other structures, and their origin and course, may not be apparent. We evaluated the ability of multiplane color Doppler transesophageal echocardiography to identify coronary fistulas.
PATIENTS AND METHODS: Twenty-one patients with angiographically confirmed coronary artery fistulas were investigated by transesophageal echocardiography in four Italian hospitals between January 1997 and May 2001.
RESULTS: Transesophageal echocardiography correctly diagnosed fistulous connection in all 21 patients. This included 6 patients with connections from the left circumflex artery (into the right chambers of the heart in 5 patients, and into the left ventricle in 1 patient), 10 patients with a fistula arising from the left anterior descending artery or left main coronary artery (with drainage into the right ventricle or main pulmonary artery), and 5 patients with a fistula from the right coronary artery (with drainage sites in the lateral aspect of the right ventricle, the low posterior right atrium, or the superior vena cava). In 4 of the 21 patients, angiography did not identify the precise site of a fistula into the coronary sinus or right ventricle.
CONCLUSION: Color Doppler transesophageal echocardiography is useful in the diagnosis and in the precise localization of coronary artery fistulas. **Am J Med. 2002;113:127–133.** ©2002 by Excerpta Medica, Inc.

Coronary artery fistulas are found in 0.3% to 0.8% of patients who undergo coronary angiography. They are the most common congenital coronary artery anomalies that affect hemodynamic parameters (1–5). Recognition of these fistulas is clinically important, since they can be associated with an increased risk of complications, including heart failure, myocardial ischemia, infective endocarditis, atrial fibrillation, and rupture. Coronary angiography remains the gold standard for imaging the coronary tree. However, the relation of coronary fistulas to other cardiac structures is at times unclear, and their origin and course can be limited by overlap of adjacent structures (5–13). The aim of the present study was to evaluate the ability of multiplane color Doppler transesophageal echocardiography to identify these abnormalities.

METHODS

Sample

Twenty-one patients with angiographically proved coronary artery fistulas were studied by multiplane color Doppler transesophageal echocardiography in four Italian hospitals between January 1997 and May 2001. In 18 patients, transthoracic imaging was also performed. All patients had clinical examinations that included 12-lead electrocardiography. Fourteen patients underwent cardiac surgery.

Echocardiography

All images were obtained using commercially available systems equipped with color-flow imaging and transesophageal capabilities (Toshiba Power Vision 6000/8000, Toshiba Corporation, Tokyo, Japan; Hewlett-Packard Sonos 2500/5500, Hewlett-Packard Inc., Andover, Massachusetts; Vingmed System Five, Vingmed GE, Hopten, Norway). Phased array transducers (2.5/2.0 MHz and 3.5/2.7 MHz) were used to acquire transthoracic images, and a 5/3.7-MHz phased array omniplane transducer mounted to the end of a flexible gastroscope was used for acquisition of transesophageal images. The echocardiograms were recorded on videotape, and still-frame prints were made using a camera attached to the ultrasound unit.

The right and left coronary ostia were seen routinely during the transesophageal study. The luminal diameter was assessed as the distance between two walls of the coronary arteries, distal to the ostia. Dilation of a coronary artery was judged to be present when its diameter appeared greater than that of the other coronary artery and >3 mm. Coronary arteries were imaged at the level of the aortic root, initially in a short-axis view, by manipulating the transducer with a combination of probe tip flexion, side-to-side tip translation, and rotation to follow their proximal course. When a coronary artery anomaly was

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Table 1. Characteristics of Patients with Coronary Artery Fistulas (n = 21)

Patient No.	Age (years)/ Sex	Symptoms	Cardiac Murmur	Electrocardiogram	Cardiomegaly on Chest Radiograph	Ratio of Pulmonary to Systemic Flow*	Other Cardiac Diagnoses	Surgery
1	61/M	Dyspnea	Systolic	Normal	−	1.7		+
2	47/M	Angina	None	ST-T wave abnormality	+	1.5		+
3	35/F	None	Systolic	Normal	−	1.1		−
4	41/F	None	Systolic	Normal	−	1.2		−
5	16/F	None	Continuous	Normal	+	1.9		+
6	36/M	Angina	None	Normal	−	1.6		+
7	12/M	None	Continuous	Left ventricular hypertrophy	−	1.2	Coarctation of aorta	+
8	29/F	None	Continuous	Normal	+	1.3		−
9	52/M	None	None	Normal	−	1.3		−
10	32/M	Dyspnea	None	ST-T wave abnormality	−	1.5		+
11	22/M	Endocarditis	Continuous	ST-T wave abnormality	−	1.4	Bicuspid aortic valve	+
12	29/F	None	Continuous	Normal	+	1.8		+
13	61/M	Dyspnea	None	Right-axis deviation	−	1.7	Atrial septal defect	+
14	18/M	None	None	Normal	−	1.5		+
15	44/M	None	Continuous	Normal	+	1.2		−
16	16/M	None	Continuous	Normal	−	1.4		−
17	54/M	Dyspnea	None	ST-T wave abnormality	+	1.5		+
18	53/M	Tachyarrhythmia	None	ST-T wave abnormality	−	1.3		+
19	32/F	None	Continuous	Normal	−	1.5		−
20	29/M	Dyspnea	Continuous	Right-axis deviation	+	1.9	Atrial septal defect	+
21	54/M	Dyspnea	None	ST-T wave abnormality	+	1.7		+

* As determined by cardiac catherization.
F = female; M = male.

identified, the origin, course, and drainage site were examined. Color flow imaging was used to aid location of the course of a tortuous coronary fistula, as well as the site of fistula drainage.

The diagnosis of coronary artery fistula was made independently by echocardiography in all patients before coronary arteriography results were obtained.

Coronary Arteriography

Coronary angiography was performed using the standard Judkins technique. Coronary injections were performed with hand injections of 5 to 10 mL of contrast in multiple projections. The film framing rate was 30 frames per second.

RESULTS

The clinical features and the transesophageal echocardiographic findings in these patients are shown in Tables 1 and 2. Eleven patients had no symptoms, and the coronary fistulas were incidental findings or discovered because of heart murmurs; 10 patients were identified because of myocardial ischemia, heart failure, or endocarditis. The coronary fistulas were also discovered incidentally during transesophageal echocardiography in 14 patients, or following an abnormal transthoracic echocardiographic study in 7 patients.

Transesophageal echocardiography correctly diagnosed fistulous connection from the left circumflex artery into the right ventricular outflow tract or main pulmonary artery in 3 patients, into the coronary sinus near its opening into the right atrium in 2 patients, and into the left ventricle in 1 patient. A fistula arising from the left anterior descending artery or left main coronary artery was observed in 10 patients, with drainage into the right ventricle or main pulmonary artery. In 5 patients, the origin of the fistula was from the right coronary artery;

Table 2. Transesophageal Echo Doppler Findings in Patients with Coronary Artery Fistulas

Patient No.	Transesophageal Echocardiography Fistula Origin	Coronary Size (mm)	Turbulent Flow Pattern	Drainage Site	Transthoracic Echocardiography	Coronary Angiography
1	Right coronary artery	8	Continuous	Right atrium	Negative	+
2	Left anterior descending artery	4	Continuous	Main pulmonary artery	Negative	+
3	Left main	12	Continuous	Right ventricle	Negative	Drainage site not identified
4	Right coronary artery	5	Continuous	Right ventricle	Not performed	+
5	Left anterior descending artery	10	Continuous	Main pulmonary artery	Suggestive	+
6	Left anterior descending artery	13	Continuous	Right ventricle	Negative	+
7	Left circumflex coronary artery	7	Continuous	Right ventricular outflow tract	Negative	+
8	Left circumflex coronary artery	11	Continuous	Left ventricular	Suggestive	+
9	Left main	8	Continuous	Main pulmonary artery	Not performed	+
10	Left anterior descending artery	4	Diastolic	Main pulmonary artery	Negative	+
11	Left circumflex coronary artery	5	Continuous	Coronary sinus	Negative	Drainage site not identified
12	Right coronary artery	19	Continuous	Right ventricle	Suggestive	+
13	Left anterior descending artery	7	Diastolic	Right ventricle	Negative	+
14	Right coronary artery	8	Diastolic	Superior vena cava	Suggestive	+
15	Left circumflex coronary artery	9	Continuous	Coronary sinus	Not performed	+
16	Left anterior descending artery	11	Continuous	Right ventricle	Suggestive	Drainage site not identified
17	Left main	10	Continuous	Main pulmonary artery	Negative	+
18	Left circumflex coronary artery	4	Continuous	Main pulmonary artery	Negative	+
19	Left circumflex coronary artery	7	Diastolic	Main pulmonary artery	Suggestive	+
20	Right coronary artery	8	Continuous	Right ventricle	Negative	Drainage site not identified
21	Left anterior descending artery	9	Continuous	Right atrium	Negative	+

drainage sites were the lateral aspect of the right ventricle, the low posterior right atrium, or the superior vena cava.

In most patients, the involved coronary arteries were markedly dilated and tortuous in appearance (Figures 1 to 4). The diameters of the involved coronary arteries ranged from 4 to 19 mm (mean, 7.1 $\pm$ 4.3 mm). The left anterior descending artery and the left main coronary artery were dilated in 8 patients and mildly dilated in 2 patients (with drainage into the main pulmonary artery). The right coronary artery was dilated in 5 patients, and the left circumflex artery was dilated in 5 patients and mildly dilated in 1 patient. In all patients, color flow Doppler mapping allowed determination of the location of drainage as turbulent flow in the receiving chamber. In 4 patients, angiography did not identify the precise site of a fistula into the coronary sinus or the right ventricle (Table 2).

Nine patients had enlargement of the left cardiac chambers, 3 patients had enlargement of all chambers, and 9 patients had normal chambers. Four patients had associated congenital heart diseases (atrial septal defect, coarctation of the aorta, or bicuspid aortic valve). The

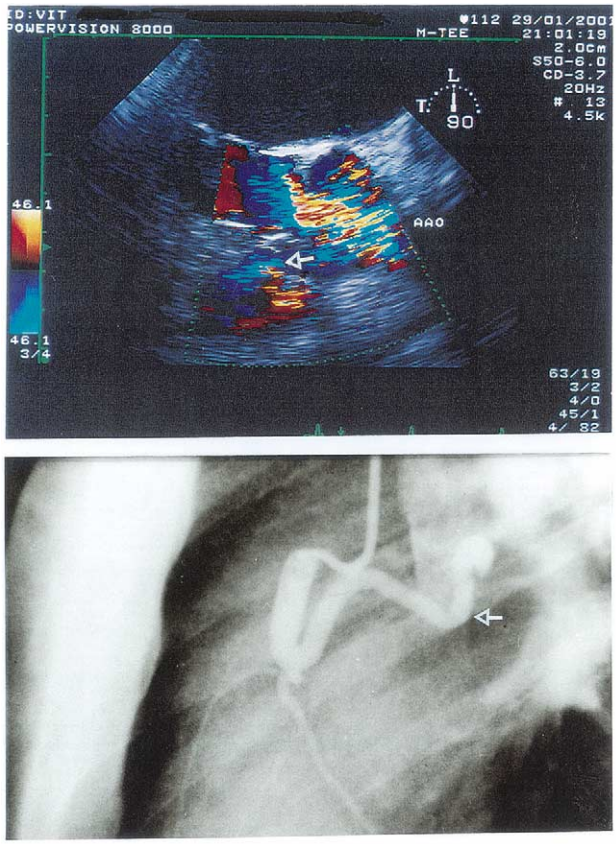

Figure 1. Multiplane transesophageal echocardiographic and coronary angiographic recordings from patient 1, showing a large right coronary artery fistula (**arrow**) arising from the aortic root and directed to the right atrium. AAO = ascending aorta.

mean (± SD) time from esophageal intubation to completion of the echocardiographic procedure was 6.5 ± 1.3 minutes (range, 5 to 13 minutes). No patient experienced a complication or adverse outcome because of esophageal intubation.

DISCUSSION

Coronary artery fistulas are difficult to detect. They are usually congenital, but may occur after trauma, such as endomyocardial biopsy (14,15). Children with this condition are usually asymptomatic. Adults may present with symptoms of fatigue, dyspnea, orthopnea, angina, or atrial arrhythmias, and with signs of heart failure, pulmonary hypertension, or endocarditis (16–23). A continuous murmur is often present and is highly suggestive of a coronary artery fistula. The differential diagnosis includes persistent ductus arteriosus, aortopulmonary window, pulmonary arteriovenous fistula, and ruptured sinus of Valsalva aneurysm.

Imaging techniques, all of which have limitations, are needed to plan surgical or percutaneous interventions. Although the course and drainage sites of anomalous coronary arteries can be visible using transthoracic echocardiography, imaging is more successful in children, in whom the acoustic window is optimal. Limitations of transthoracic echocardiography have been described in patients with coronary artery fistulas and in patients with anomalous coronary vessels (6–8). Transthoracic color Doppler echocardiography may not detect flow in the distal site of a coronary artery fistula.

The advantages of transesophageal echocardiography include the proximity of the transducer to the area of interest, as well as the use of higher-frequency transducers that provide better spatial resolution and assessment of detail (10–13). The multiplane device is also capable of displaying the coronary artery in any plane and providing a panoramic view of the fistula's origin, course, and drainage site. Most of our patients had continuous turbulent flow at the site of drainage visualized by color Doppler flow mapping, but occasionally the flow was only visible during diastole. During isovolumic contraction, an interruption of the flow within the fistula can occur because of an increase in intramyocardial vascular resistance and a rapid decrease of the gradient between the aorta and the left ventricle (8).

Low-pressure structures are the most common recipient sites. These include the right ventricle, right atrium, pulmonary artery, coronary sinus, and superior vena cava. Coronary artery dilatation is usual, and the degree of dilatation does not always depend on the shunt size. If the fistula arises from the distal part of a coronary artery, the artery diameter will be small, and the diagnosis can be missed. In addition, when the drainage site is into the pulmonary artery, the shunt ratio is smaller than when the fistula connects with the right atrium or right ventricle. The discovery of turbulent flow in the pulmonary artery may help in the diagnosis of a fistula when the left coronary artery is only mildly enlarged.

Although gated magnetic resonance imaging allows detailed delineation of cardiac anatomy and blood flow, and may be helpful in resolving discrepancies raised by other investigations (23–24), coronary angiography remains the gold standard for imaging the coronary arteries. However, transesophageal echocardiography might have a complementary role in patients with angiographically documented coronary artery anomalies. For example, among patients in whom angiography fails to detect the drainage site, transesophageal echocardiography may be useful. When a coronary artery fistula is present, a dilated feeder vessel with an abnormal flow pattern can be identified readily. This information may affect management, since it is important in planning the optimal surgical approach (25–27) and in determining the need for

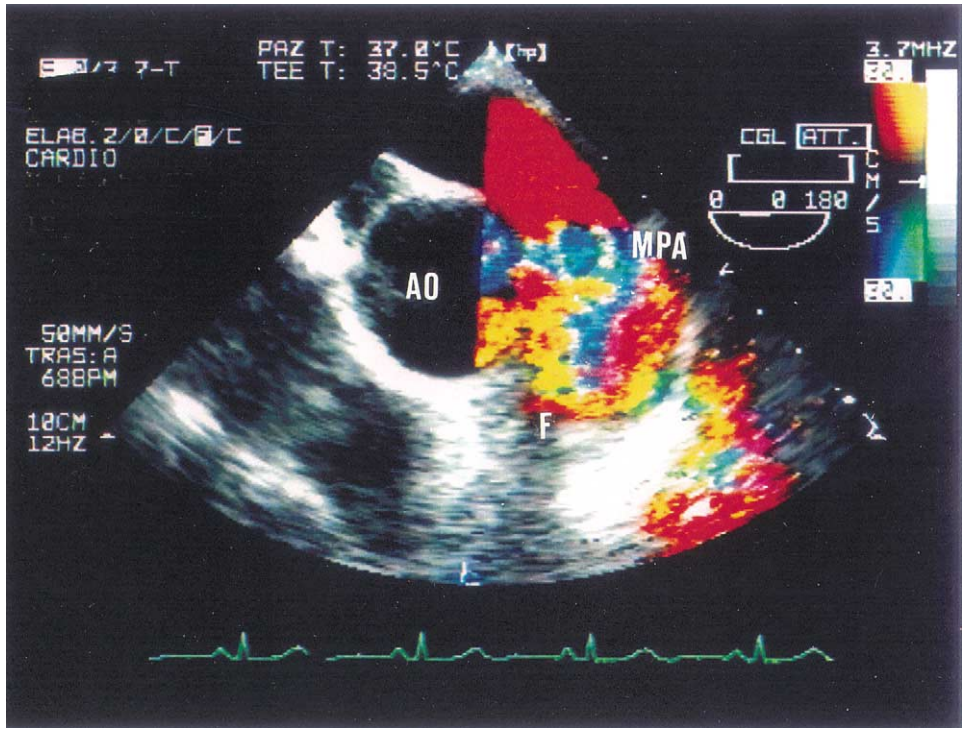

Figure 2. Multiplane transesophageal echocardiographic recording from patient 5, showing a coronary artery fistula (F) involving the left anterior descending artery with drainage into the main pulmonary artery (MPA). AO = aorta.

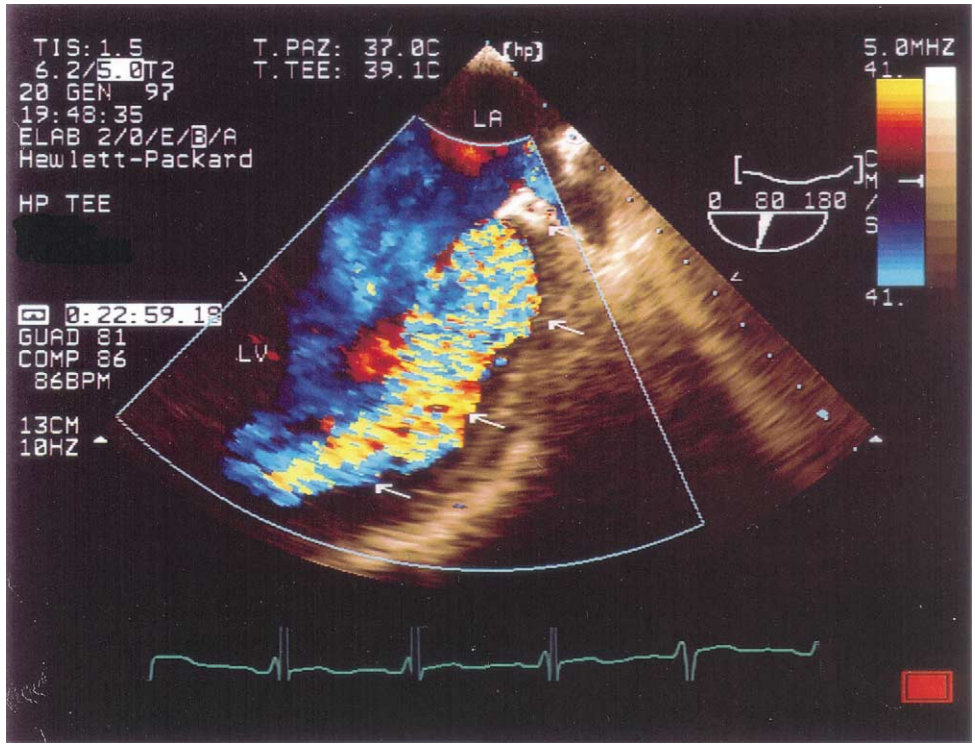

Figure 3. Multiplane transesophageal echocardiographic recording from patient 8, with the fistulous vessel (**arrows**) originating from the circumflex artery with drainage into the left ventricle (LV). LA = left atrium.

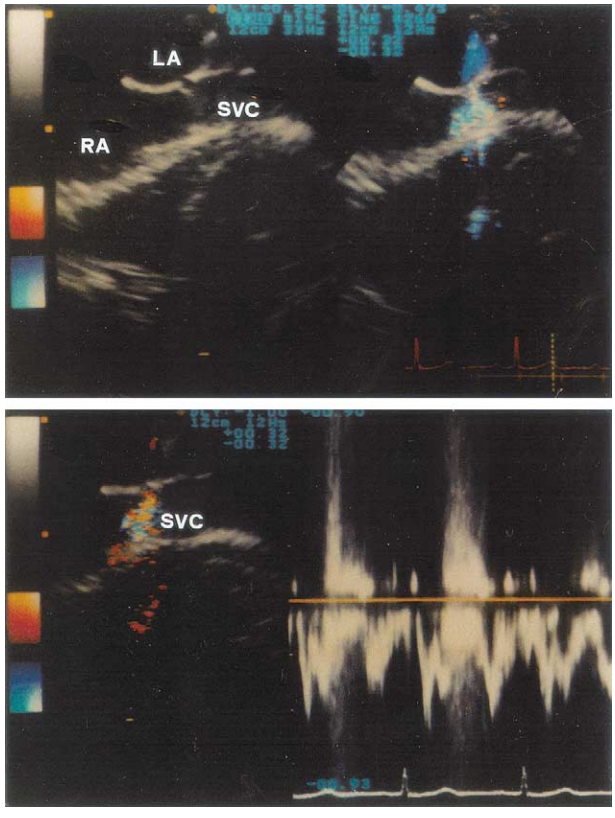

Figure 4. Multiplane transesophageal echocardiographic recording from patient 14, showing a right coronary artery fistula with drainage into the superior vena cava (SVC). Turbulent diastolic flow on spectral Doppler display is also shown. LA = left atrium; RA = right atrium.

cardiopulmonary bypass. Indeed, 2 of our patients had surgical closure of a fistula without cardiopulmonary bypass.

Even if multiplane transesophageal echocardiography provides high-quality images of proximal coronary arteries and their anomalies, it has some limitations. Identifying a coronary fistula may be limited if a vessel is curvilinear along the epicardial surface, which impairs the view of distal segments unless they are dilated. Collateral vessels are often of small caliber and are not seen by transesophageal echocardiography. Finally, obstructive coronary artery disease cannot be evaluated accurately. Despite these limitations, color Doppler transesophageal echocardiography is useful not only in the diagnosis but also in the localization of coronary artery fistulas, especially those that are not identified with angiography.

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