Doppler echocardiographic features of coronary arteriovenous fistula

Complementary roles of cross sectional echocardiography and the Doppler technique

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SUMMARY The Doppler echocardiographic features of coronary arteriovenous fistula were investigated in eight patients with left or right coronary arteriovenous fistulas who had a continuous heart murmur in the upper precordial area and whose diagnoses were confirmed by coronary angiography. In four patients the dilated lumen of the coronary arteriovenous fistula was visualised by cross sectional echocardiography. Of these, three showed abnormal unidirectional continuous flow signals with broad velocity spectra in the fistula. Abnormal, powerful, unidirectional or bidirectional continuous Doppler signals were detected in part of the pulmonary artery in two of the eight patients, in part of the right ventricle in two, and in part of the right atrium in one; these signals were interpreted as indicating shunt flow. Although the opening of the fistula was difficult to visualise by cross sectional echocardiography, the pulsed Doppler technique helped identify the site in patients with dilatation of the coronary artery. In the remaining three patients with a small shunt no abnormal findings were obtained with cross sectional echocardiography or the Doppler technique. The size of the fistula below which no abnormal findings may be obtained by Doppler echocardiography still needs to be determined.

The first step in the clinical diagnosis of coronary arteriovenous fistula is the finding of a continuous heart murmur in the upper precordial area. To confirm the diagnosis of this anomaly coronary angiography is needed. With recent advances in cross sectional echocardiography, the root of the coronary artery may now be imaged non-invasively,¹⁻³ and dilatation of the root of the coronary artery has been reported in cases of coronary arteriovenous fistula.^{4,5} Even with cross sectional echocardiography, however, the site of the shunt—that is, the opening of the fistula—cannot always be detected.

In the present study we attempted to identify the coronary arteriovenous fistula, detect the shunt site, and determine the haemodynamic conditions inside the fistula using cross sectional echocardiography and the Doppler technique.

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Patients and methods

Eight patients (five men, three women; age range 22-64 years) with a coronary arteriovenous fistula were examined. They were referred to the National Cardiovascular Center for cardiac catheterisation and angiocardiology because of a continuous heart murmur in the upper precordial area, although their exertion capacity was satisfactory. Coronary angiography showed that the continuous heart murmur originated from a coronary arteriovenous fistula.

DOPPLER TECHNIQUE

The equipment used included two commercially available cross sectional echocardiographs, a Toshiba SSH 11A/SDS 10A and a SSH 40A/SDS 21A, which incorporated a pulsed Doppler flowmeter in an ultrasonic wide angle phased array system. The ultrasonic frequency used was 2.4 MHz and the pulse repetition rates were 4 and 6 kHz. The sample volume was tear-drop shaped and its size was 4 mm in width

Coronary arteriovenous fistula by Doppler echocardiography

Table Doppler echocardiographic and angiographic findings

Case No	Age (yr)	Sex	Dilated coronary artery by cross sectional echocardiography	Abnormal Doppler signal		Shunt site by angiography
				Coronary fistula	Heart chamber	
1	21	M	+	+	+	RV
2	32	F	+	+	+	RA
3	54	M	+	—	+	PA
4	64	F	—	—	+	PA
5	45	M	+	+	+	RV
6	28	F	—	—	—	PA
7	38	M	—	—	—	PA
8	53	M	—	—	—	PA

+, present; —, absent; RV, right ventricle; PA, pulmonary artery; RA, right atrium.

and 2 mm in depth.

The ultrasonic pulses for cross sectional echocardiography and the Doppler technique were transmitted alternately so that the echocardiographic images and Doppler searching were done simultaneously. In practice, however, the cross sectional image was recorded first followed by the Doppler signal, since when both were recorded simultaneously the pulse repetition rate had to be reduced to 2 and 3 kHz for each, which resulted in impairment of the quality of the image and the Doppler signal.

The examination was performed with the patient in the supine position. Cross sectional echocardiography and the Doppler examination were performed from the conventional parasternal, apical, and subcostal approaches.⁶ The Doppler signals were analysed with a spectroanalyser based on the fast Fourier transform; the sound spectrogram obtained by this means indicated a flow pattern composed of directional flow components with velocity spectra. The electrocardiogram, phonocardiogram, and M mode echocardiogram were displayed polygraphically with the Doppler flow pattern by a strip chart recorder. The paper speed was 50 mm/s.

Results

In three of the seven patients with a left coronary arteriovenous fistula cross sectional echocardiography showed a round echo-free area with a diameter of about 1–2 cm just left of the aortic root in the short axis view at the level of the aortic valve (cases 1, 2, and 3; Table). In one patient (case 5) with a right coronary arteriovenous fistula a round echo-free area was also seen on cross sectional echocardiography where the vessel branched off the aorta. As in previous reports⁴ of the echocardiographic features of coronary arteriovenous fistula the round echo-free area was interpreted as being a dilatation of the root of the left coronary artery or the right coronary artery. In three of the four patients with this feature unidirectional Doppler signals of blood flow were detected in the dilated lumen (cases 1, 2, and 5).

In two cases (cases 1 and 2) powerful abnormal Doppler signals with bidirectional components with broad velocity spectra indicating disturbed blood flow were detected continuously in a heart chamber during one cardiac cycle suggesting shunt flow. In a further three cases the abnormal Doppler signals with unidirectional components with broad velocity spectra were detected continuously in the pulmonary artery (cases 3 and 4) and the right ventricle (case 5). These findings also suggest shunt flow. In the three patients (cases 6, 7, and 8) with a small shunt no positive finding was obtained by cross sectional Doppler echocardiography.

CASE REPORTS

Case 1

In a 21 year old man an echo-free area (2 cm in diameter) was imaged at the left side of the aorta in the short axis view in the third intercostal space by the left parasternal approach (Fig. 1). This area communicated with the aorta and was considered to be the dilated left coronary artery on the basis of its anatomical relations. This abnormal luminal dilatation ran towards the apex of the heart and could be traced to the part of the anterior wall of the left ventricle near the interventricular septum. Nevertheless, there was no clear image of the site of the shunt.

In the dilated lumen unidirectional Doppler signals were recorded indicating continuous flow during systole and diastole. This flow pattern showed a unidirectional broad velocity spectrum and the flow direction was interpreted as being from the base to the apex of the heart. The highest component of the instantaneous flow velocity spectrum was generally higher in diastole than in systole with its maximum in late diastole (Fig. 1). In this patient, powerful bidirectional continuous Doppler signals were also detected in the right ventricular outflow tract during systole and diastole. Coronary angiography confirmed that the aneurysmal dilated fistula ran from the main trunk of the left coronary artery to the anterior wall of the left ventricle. The distal end of the fistula then became narrow and opened into the right ventricular outflow

Fig. 1 Cross sectional and Doppler echocardiograms (case 1): (a) An echo-free area of about 2 cm in diameter is present at the left side of the aorta in the aortic short axis view at the level of the sinus of Valsalva. The white line shows the beam direction in which the Doppler signals were recorded. The white arrow indicates the site of the sample volume for the Doppler signal. (b) An echo-free area is present in the anterior wall of the left ventricle in the short axis view of the left ventricle. (c) The Doppler signal in this echo-free area showed a unidirectional wide band velocity spectrum pattern; the maximum velocity of this abnormal flow is seen in end diastole. (d) The Doppler signal obtained by another system (Hitachi Medico EUD-4Z) showed the same pattern. AO, aorta; LV, left ventricle; RV, right ventricle; SV, sample volume; PCG, phonocardiogram.

Fig. 2 Cross sectional and Doppler echocardiograms and coronary angiogram (case 1): (a) The sample volume set in the outflow of the right ventricle (white arrow). (b) Powerful, bidirectional continuous abnormal Doppler signals were recorded in the outflow tract of the right ventricle, the sampling sites of which are shown in (a). (c) An angiogram shows: a coronary arteriovenous fistula with an aneurysmal dilatation at the proximal end; the distal end opens into the right ventricle outflow tract (white arrow). LV, left ventricle; RV, right ventricle.

tract (Fig. 2).

Case 2

In a 32 year old woman cross sectional echocardiography showed a dilated coronary lumen (cavity) at the origin of the left coronary artery (Fig. 3). The dilated lumen ran behind the ascending aorta and on the posterior wall of the heart. It was evident from the subcostal approach that this dilated lumen continued behind the right atrium and had a diameter of about 2 cm. Doppler signals at this site showing a mild turbulent unidirectional non-pulsatile blood flow were detected continuously during systole and diastole, and the flow direction was interpreted as being from the

aorta to the periphery (Fig. 3). On the cross sectional echocardiogram the dilated lumen at the rear of the right atrium appeared to open into the right atrium, though this was not certain. When the Doppler sample volume was set at this site of possible opening, powerful bidirectional continuous signals were detected. These abnormal signals were detected spreading from this site into the right atrium. Coronary angiography confirmed that a dilated coronary fistula about 2–3 cm in diameter arose at the origin of the left coronary artery, wound around behind the heart, and opened into the right atrium from the right posterior direction (Fig. 4).

Fig. 3 Cross sectional and Doppler echocardiograms (case 2): (a) In the aortic short axis view a dilated coronary lumen of about 1.5 cm in diameter was imaged at the origin of the left coronary artery. (b) The Doppler flow pattern in the luminal dilatation showed a unidirectional continuous flow with a wide band velocity spectrum. (c) The dilatation was imaged behind the right atrium in the view from the right edge of the sternum. (d) The same continuous Doppler flow pattern was recorded at the sampling site (white arrow in (c)). AO, aorta; PA, pulmonary artery; RA, right atrium; LA, left atrium.

Fig. 4 Cross sectional and Doppler echocardiograms and coronary angiogram (case 2): (a) In this view the luminal dilatation was imaged behind the right atrium. (b) In the Doppler recording powerful, continuous Doppler signals were recorded in the right atrium. (c) In the coronary angiogram a dilated fistula at the origin of the left coronary artery winds around behind the heart and opens into the back of the right atrium. RA, right atrium; LA, left atrium.

Case 3

In a 54 year old man cross sectional echocardiography showed that the main trunk of the left coronary artery appeared to be slightly dilated (Fig. 5). This dilatation was not, however, confirmed. No other abnormalities were seen on the cross sectional echocardiogram. With the Doppler method flow signals could not be detected at the trunk of the left coronary artery. A turbulent blood flow during systole and diastole was, however, detected and was localised in the anterior part of the main trunk of the pulmonary artery. There appeared to be a shunt flow to this part. Coronary angiography showed that the left coronary artery had a dilatation in its trunk, formed a cavern-shaped area in front of the pulmonary artery, and then opened

into the main trunk of the pulmonary artery (Fig. 5).

Case 4

In a 64 year old woman no dilatation was found at the origin of the left coronary artery, but a disturbed blood flow was detected in the pulmonary artery. The findings were the same as those in case 3.

Case 5

In a 45 year old man cross sectional echocardiography showed a dilatation (cavity) at the origin of the right coronary artery. This dilated vessel ran to the right anteriorly but could not be traced further by cross sectional echocardiography. The Doppler flow signals

Fig. 5 Cross sectional and Doppler echocardiogram and coronary angiogram (case 3): (a) The long axis view shows the outflow tract of the right ventricle to the pulmonary artery. The white arrow indicates the site of the sample volume for the Doppler signal. (b) A bidirectional Doppler signal with a broad velocity spectrum was recorded near the anterior wall of the pulmonary artery during systole and diastole. The turbulence appeared to be less when the sample volume was moved away from the anterior wall to the centre. (c) The coronary angiogram shows that the cavern-shaped fistula at the origin of the left coronary artery opens into the anterior part of the pulmonary artery. RV, right ventricle; PA, pulmonary artery.

in this dilated lumen showed a slightly turbulent, unidirectional continuous flow, the flow velocity of which was faster in systole than in diastole (Fig. 6). Another abnormal round echo-free area was imaged at the medial side of the right ventricle on the cross sectional image of the parasternal four chamber view, where uncertain Doppler flow signals were recorded. Unidirectional turbulent Doppler signals were detected continuously in the medial part of the right ventricle. Coronary angiography and left ventriculography showed that the right coronary artery was dilated in the proximal and mid-portions and communicated with the medial part of the right ventricle

(Fig. 7).

In the other three cases no abnormal findings were found by cross sectional echocardiography and the Doppler technique. In these three cases a narrow fistula from the left coronary artery to the pulmonary artery was detected by coronary angiography.

Discussion

BLOOD FLOW IN THE CORONARY FISTULA

It has been previously reported⁴ that an abnormal round echo-free area detected near the root of the left coronary artery is considered to be the dilated coro-

Fig. 6 Cross sectional and Doppler echocardiograms (case 5): (a) An echo-free area of about 1 cm in diameter is seen in the anterior part of the aorta where it branches out from the ascending aorta. (b) The Doppler signals in this dilatation show a unidirectional turbulent flow pattern, in which the flow velocity is faster in systole than in diastole. (c) An echo-free area is seen at the medial side of the right ventricle in the parasternal four chamber view. (d) The Doppler flow signal in the echo-free area was not recorded clearly. LV, left ventricle; LA, left atrium; Ao, aorta; RV, right ventricle.

Fig. 7 Cross sectional and Doppler echocardiograms and coronary angiogram (case 5): (a) In the parasternal four chamber view the right ventricle was enlarged on the medial side. The Doppler sample volume was set in the medial side of the right ventricle (white arrow). (b) The Doppler signals in this sample volume showed turbulent blood flow with the peak velocity occurring during systole. (c) A right coronary angiogram showed that the right coronary artery was dilated in the proximal portion, had an aneurysmal dilatation in the distal portion, and opened into the right ventricle. (d) A left ventriculogram showed the same findings as (c). RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium.

nary artery in cases of coronary arteriovenous fistula. In the present study, blood flow signals in the dilated coronary arteries were first tested for by the Doppler technique in cases in which coronary arteriovenous fistula was confirmed by coronary angiography. The blood flow signals were detected in the dilated coronary artery if the artery was dilated at least to 1 cm in diameter. The equipment we used, which incorporated a Doppler flowmeter in a wide angle phased array system, was appropriate for searching for a moving target such as the coronary artery, because the lumen could be imaged with the sample volume while simultaneously obtaining the Doppler signals. Even with this kind of equipment, however, it was difficult to image the lumen in cases with a very small lumen.

In case 1 the flow velocity in the coronary arteriovenous fistula was faster in diastole than in systole, which is similar to the normal coronary arterial flow pattern. In case 2 and case 5 the blood flow was continuous, being almost steady throughout one cardiac cycle or slower in diastole than in systole. The reason that these two kinds of flow pattern differ is due to whether or not the coronary arteriovenous fistula passes through the myocardium. If the fistula passes through the myocardium, the cavity in the myocardium is compressed in systole, and therefore the flow velocity during systole may be slower in the other non-compressed part of the fistula.

The Doppler signals obtained from the dilated coronary artery showed that the flow was slightly turbulent although unidirectional. The reason for this turbulence is probably the unusual dilatation and tortuosity of the lumen of the fistula.

SHUNTING SITE OF THE FISTULA

In general, in patients with a continuous heart murmur there is an arteriovenous shunt in or near the heart. In such patients a detailed examination of the pulmonary artery, right ventricle, right atrium, and vena cavae is necessary. In the present study, it was generally difficult to detect and identify the opening of the fistula by cross sectional echocardiography. The same problem was encountered in several previous studies.^{4,5} By the combined use of cross sectional echocardiography and the Doppler technique, however, powerful bidirectional or unidirectional continuous Doppler signals with broad velocity spectra were detected in the part of the cardiac chamber with which the fistula communicated. Such Doppler signals with broad velocity spectra are assumed to originate from blood flow turbulence due to the mixing of the shunt flow with the normal flow in the relevant chamber. These signals were easy to detect, particularly at the maximum point of the continuous heart murmur on the precordium. Therefore, when a dilatation of the coronary artery is imaged and abnormal

continuous Doppler signals detected in part of a certain chamber, there is probably a coronary arteriovenous fistula opening into part of that chamber, even though the opening of the fistula is not visualised. The diagnosis may be clarified further by interpreting the Doppler findings and the cross sectional echocardiographic findings together even though they are still not conclusive. In patients in whom the shunt was very small and the lumen of the fistula narrow, however, no abnormalities could be detected by cross sectional echocardiography and the Doppler technique. The size of the fistula below which no abnormal findings may be obtained by Doppler echocardiography still needs to be determined.

DIFFERENTIAL DIAGNOSIS

In the diagnosis of coronary arteriovenous fistula by Doppler echocardiography, it is necessary to differentiate between coronary arteriovenous fistula and other heart diseases producing a continuous murmur in the upper precordial area such as persistent ductus arteriosus or a ruptured aneurysm of the sinus of Valsalva. For this purpose, it is important to confirm first that there is a dilatation at the origin of the coronary artery. This finding is not seen in cases of persistent ductus arteriosus.⁴ In case 4 in our study, abnormal Doppler signals were detected in the pulmonary artery, although a dilatation of the coronary artery was not confirmed. This indicates that some sort of shunt flow has passed into the pulmonary artery.⁷ In this case, however, it was difficult to differentiate between a coronary arteriovenous fistula into the pulmonary artery and persistent ductus arteriosus by ultrasonic methods alone.

In cases in which continuous turbulent blood flow signals are detected in an area near the sinus of Valsalva, it is necessary to differentiate between a coronary arteriovenous fistula and a ruptured sinus of Valsalva aneurysm. However, in patients with a ruptured sinus of Valsalva, the aneurysm is often evident by cross sectional echocardiography.

Thus the present study has shown that coronary arteriovenous fistula may be diagnosed with more certainty and its shunt site identified by the combined use of cross sectional echocardiography and the Doppler technique.

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