Selective coronary angiography in primary myocardial disease¹

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Coronary angiography was performed in 16 patients with a primary myocardial disease, Bantu cardiomyopathy. Two patients had discrete coronary occlusion, presumed embolic from ventricular mural thrombus. There was no evidence of coronary atheroma and all the vessels and their proximal intramural branches were large. The total cross-sectional area of the proximal coronary arteries correlated with left ventricular muscle mass. The coronary angiograms were also used to study left ventricular function.

Primary myocardial disease is common in the South African Bantu. The aetiology is unknown, though excessive ingestion of alcohol and toxins, thiamine and tryptophane deficiency, viral infections, and pregnancy have all been incriminated (Gillanders, 1951; Higginson, Gillanders, and Murray, 1952; Becker, Chatgidakis, and Van Lingen, 1953; Grusin, 1957; Burch and Walsh, 1960; Seftel and Susser, 1961; Smith, 1970; Schrire, 1971). In Jamaica a few patients with primary myocardial disease have an abnormality of the smaller coronary arteries which may be responsible for the myocardial fibrosis and poor left ventricular function (Stuart and Bras, 1971; Campbell *et al.*, 1971). Coronary blood flow has been studied but found to be normal in primary myocardial disease (Brink and Lewis, 1967).

Cardiomyopathy also occurs in Caucasian patients. Their major coronary arteries are normal but it has been suggested that the blood supply to the myocardium is inadequate and that this limits compensatory hypertrophy and may be responsible for the poor prognosis (Oakley, 1971).

Ischaemic heart disease is uncommon in the South African Bantu though isolated cases occur in subjects who are hypertensive, diabetic, or exposed to a Western diet. The electrocardiographic pattern seen in these patients may be indistinguishable from primary myocardial disease.

Selective coronary arteriography was performed in 16 patients with Bantu primary myocardial disease. The investigation was undertaken as a diagnostic procedure to exclude coronary artery disease,

to study the morphology of the coronary arteries, to relate their anatomy to the ventricular hypertrophy, and to use the arteriogram to study ventricular dynamics. The patients did not understand English so the procedures were explained to them in their own language; they understood that the investigation was not expected to contribute to their treatment or to be of special benefit to them.

Patients

Sixteen patients were studied. Their clinical, electrocardiographic, and radiological features are given in Table 1 and summarized in Table 2, and the haemodynamic features shown in Table 3. The basic abnormality was left ventricular dysfunction and more severely affected patients showed increasing degrees of cardiac enlargement, a loud third heart sound, functional mitral incompetence, pulmonary arterial hypertension, and cardiac failure (Gotsman, van der Horst, and Winship, 1971a; Gotsman *et al.*, 1971b). Two patients were thought to have coronary embolism because the electrocardiograms showed evidence of transmural myocardial infarction; isolated obstruction of a single coronary artery was subsequently confirmed by coronary angiography, and there was no evidence of atheroma elsewhere in the coronary circulation. One patient had overt pulmonary thromboembolism.

Methods

The patients were assessed on the basis of standard clinical, electrocardiographic, and radiological criteria. Right and left heart catheterization was performed using the mid-chest level as the zero reference for pressures. Cardiac output was measured by the direct Fick method. Selective coronary arteriography was carried out by either the Sones or the Judkins method in multiple oblique views (Sones and Shirey, 1962; Judkins, 1967;

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TABLE I The patients

	Age (yr)	Sex	Dis- ability*	Electrocardiogram	Heart volume†	Systolic time intervals (PEP/LVET)‡
<i>Case No.</i>						
1	50	M	4	Anterolateral infarction	1000	—
2	48	M	4	Anteroseptal infarction	1050	0.65
3	27	M	1	Left anterior hemiblock; left ventricular hypertrophy	460	0.50
4	47	M	1	Left anterior hemiblock; normal voltage	515	0.41
5	72	M	3	Left ventricular hypertrophy	1410	0.44
6	50	M	2 (b)	Left anterior hemiblock; left ventricular hypertrophy	830	0.31
7	36	F	3	Left ventricular hypertrophy	—	1.00
8	60	M	3	Left bundle-branch block	610	0.46
9	65	M	2 (a)	Left anterior hemiblock; left ventricular hypertrophy	560	0.46
10	65	M	3	Incomplete RBBB + left anterior hemiblock	640	0.64
11	44	M	2 (b)	Left anterior hemiblock; left ventricular hypertrophy	935	0.82
12	40	M	4	Left anterior hemiblock; left ventricular hypertrophy	940	—
13	48	M	3	Left anterior hemiblock; left ventricular hypertrophy	890	0.79
14	55	M	4	Left anterior hemiblock; left ventricular hypertrophy	955	0.94
15	34	M	4	Left anterior hemiblock; left ventricular hypertrophy	775	0.61
16	41	M	4	Atrial fibrillation; incomplete RBBB; left ventricular hypertrophy	1478	—
<i>Normal Subjects</i>						
A	27	F		Normal	274	
B	39	F		Normal	446	
C	39	F		Normal	412	
D	34	F		Normal	320	

* Disability = New York Heart Association (1964) grading at time of study.

† Heart volume = Radiographic heart volume expressed in ml/m² (Jonsell, 1939).

‡ PEP = pre-ejection phase; LVET = left ventricular ejection time (Weissler and Garrard, 1971a, b).

TABLE 2 16 patients with primary myocardial disease

Cardiac disability (N.Y.H.A. grading)	No.
Grade 1	2
Grade 2 (a)	1
(b)	2
Grade 3	5
Grade 4	6
Electrocardiogram	No.
Infarction	2
Left anterior hemiblock	10
Left bundle-branch block	1
Left ventricular hypertrophy	11

Gotsman *et al.*, 1969). The procedure was completed with selective left ventriculography in the right anterior oblique view in all patients and in the left anterior oblique view also in 7 of them.

The following features were examined on the cine-angiograms.

1) The anatomy and abnormalities of the coronary arteries.

2) Left ventricular volume and wall mass using the uni-plane method of Rackley *et al.* (1964) and Greene *et al.* (1967). Ejection fraction was calculated from the end-diastolic and end-systolic volumes.

3) Left ventricular ejection fraction was also calculated from the coronary arteriogram.

4) The relative size of the individual cardiac chambers was estimated.

5) The internal diameter of the proximal left and right coronary arteries (before their first branch) was measured and their respective and total cross-sectional areas calculated.

6) Studies were made of the movement of the coronary arteries in the atrioventricular groove and of relative shortening and folding of segments of the coronary arteries.

The diameter of the cardiac catheter was used as a basis for assessing magnification. This technique is ideal for the comparison of small measurements such as coronary artery internal diameter or ventricular wall thickness, but it may exaggerate errors in ventricular volumes. None the less, estimation of ejection fraction is independent of magnification and is therefore accurate.

The injection of contrast medium alters ventricular dynamics and though angiographic techniques have gained wide acceptance, we have reservations about their validity. The rapid injection of contrast often induced ventricular ectopic beats and though we ignored these

TABLE 3 Haemodynamic and angiographic measurements

	LVEDP (mmHg)	PAP (mmHg)	EF (%)	CI (l./min/ m ²)	SI (ml/beat/ m ²)	EDV (ml/m ²)	LV wall thickness (mm)	LV wall mass (g)	LCA (mm)	RCA (mm)	Total cross- sectional area (mm ²)
<i>Case No.</i>											
1	30	21	35	2.2	31	151	11.4	247	6.4	5.6	56.8
2	35	47	—	2.3	23	131	14.9	377	4.0	3.3	21.2
3	2	15	37	3.5	64	234	12.8	497	3.6	2.8	16.5
4	4	11	35	3.0	43	254	15.2	591	4.7	4.0	29.9
5	10	20	32	2.8	31	330	10.9	415	3.6	3.3	19.0
6	20	22	30	2.5	49	194	11.1	359	4.6	4.0	29.3
7	6	10	30	3.2	45	475	10.7	583	4.7	6.3	48.6
8	20	25	20	2.9	29	523	13.0	761	5.3	4.4	37.6
9	5	22	34	4.1	51	175	14.0	470	6.7	5.3	57.2
10	6	50	39	2.1	23	217	13.4	495	5.6	—	—
11	27	37	13	2.9	26	143	10.8	278	6.0	—	—
12	20	30	21	1.7	14	232	15.1	555	6.2	3.8	41.4
13	25	—	19	2.0	25	220	12.0	415	5.0	4.2	34.7
14	35	40	17	1.5	13	330	14.7	677	6.0	6.2	50.2
15	30	43	21	3.4	32	252	12.3	506	8.4	4.2	69.1
16	28	34	26	2.5	34	320	9.4	357	4.3	5.3	36.1
<i>Normal subjects</i>											
A	11	—	59	—	—	119	9.2	209	3.2	3.7	19.0
B	10	20	66	4.7	72	98	11.4	241	3.8	4.0	24.7
C	12	17	62	4.9	65	125	8.3	239	4.0	3.7	23.8
D	5	10	66	4.0	37	97	9.4	272	3.9	4.7	29.5

LVEDP = left ventricular end-diastolic pressure; PAP = mean pulmonary arterial pressure; EF = ejection fraction; CI = cardiac index; SI = stroke index; EDV = left ventricular end-diastolic volume; LCA = left coronary artery internal diameter; RCA = right coronary artery internal diameter; total cross-sectional area = total cross-sectional area of left and right coronary arteries.

and the immediate post-ectopic beats, premature ventricular contractions were responsible for ventricular alternans.

Four premenopausal European women who had consulted several physicians in the city on account of chest pain suggesting ischaemic heart disease were also investigated by the same methods. They understood the nature of the investigation and assented to it. In each instance no evidence of coronary heart disease was found and these subjects could therefore be regarded as normal controls. The results were of therapeutic value to the patients and gave reassurance to the physicians referring them.

Results

In 14 patients, the right coronary artery was dominant, terminating as the posterior interventricular branch.

Two patients had obstruction of a coronary artery: one had abrupt obstruction of the right coronary artery immediately beyond its marginal branch and the distal vessel filled through collaterals. The second patient had an abrupt obstruction of the left anterior descending artery (Fig. 1 and 2). Neither had obvious coronary atheroma. This does not exclude underlying atherosclerosis but the

severe generalized left ventricular hypokinesis was in keeping with primary myocardial disease, and embolism from thrombus overlying the ventricular endocardium was a likely source.

The coronary arteries and their proximal intramural branches were unusually large and dilated and were stretched over a voluminous ventricle like the tentacles of an octopus (Fig. 3). In a normal subject these vessels are tortuous and the tortuosity increases during systole. Our patients had a large dilated ventricle with a low ejection fraction and little change in size during systole (Fig. 4a and b). The vessels were elongated and lacked the normal tortuosity and, though they had an attenuated appearance, measurement revealed that they were in fact much larger than normal. The intramural coronary arteries were enlarged in contrast to normal angiograms where only a few vessels are seen. This suggests either unusual dilatation or an increase in blood flow. The smaller branches could not be studied by this technique as the resolving power of the radiological system was inadequate.

The coronary arteriograms showed a three-dimensional view of the cardiac skeleton and relative displacement of the vessels provided information about the size of the cardiac chambers. In the

FIG. 1 Left coronary angiogram in the right anterior oblique view of the patient with left anterior descending artery obstruction.

FIG. 2 Precatheterization electrocardiogram from the same patient showing the features of anteroseptal infarction.

left anterior oblique view, the circumflex, and the right coronary arteries showed the size and position of the left and right atrioventricular rings. Patients with pulmonary arterial hypertension and right ventricular dilatation had a dilated tricuspid annulus (Fig. 5). With left ventricular dilatation the angle at

the division of the left coronary artery into its anterior descending and circumflex branches was increased and the circumflex artery was splayed over the large left ventricle (Fig. 6).

Kinetic studies of arterial movement provide interesting information about the changes in atrio-

FIG. 3 Left coronary arteriogram in the right anterior oblique view showing large, dilated, elongated arteries extending around the voluminous left ventricle and stencilling its outline. Intramural branches are well seen. There is loss of tortuosity in these vessels.

ventricular volumes. In the right anterior oblique view the left coronary artery stencils the outline of the left ventricle, and its circumflex branch shows the mitral annulus in profile. If allowance is made for left ventricular wall thickness, the left coronary artery outlines left ventricular end-diastolic and end-systolic volumes and so demonstrates the ejection fraction. Moreover, left ventricular function is not influenced by the delivery of a large volume of contrast medium into the left ventricle which alters the preload. Study of left circumflex artery movement in this view showed that diastolic events were important, and Fig. 7 shows the contributions of the rapid filling phase and atrial systole to ventricular filling. The rapid filling phase has a distinct halt at the limit of ventricular compliance. This phenomenon was most marked in ill patients who had large dilated left ventricles and corresponded with the clinically audible third heart sound. Ventricular movement during systole was also clearly identified

at the atrioventricular ring. It was slow and the rate of ventricular filling during the two phases of diastole was almost as great as the rate of myocardial fibre shortening during ventricular contraction and ejection. The right coronary artery showed movement of the right atrioventricular groove in the same right anterior oblique view.

The internal diameter of the proximal coronary arteries and the sum of their cross-sectional areas are shown in Table 3 and the relation to left ventricular muscle mass is shown in Fig. 8. Four normal subjects are shown for comparison. In general, the total cross-sectional area of the proximal coronary arteries is related to muscle mass, but patients with mild primary myocardial disease had an increase in muscle mass without a corresponding increase in coronary arterial size. The patient with obstruction of the anterior descending left coronary artery had a normal mass, but large proximal vessels. It is possible that left ventricular wall thickness was

FIG. 4 (a and b) Serial frames from the left coronary arteriogram (right anterior oblique view) of a 36-year-old African woman with primary myocardial disease. The magnification is the same in these two pictures. There has been very little change in position of the arteries from end-diastole (a) to end-systole (b) indicating a low left ventricular ejection fraction. Most movement has involved the circumflex artery in the left atrioventricular groove. There is little change in the tortuosity of the vessels between end-systole and end-diastole.

measured in a fibrotic area and that the left ventricular muscle mass was therefore underestimated.

The left ventriculogram showed a dilated left ventricle with an increase in end-diastolic volume and a low ejection fraction. The details are summarized in Table 3. Similar values for ejection fraction were calculated from the coronary arteriograms. The two patients with infarction showed local areas of dyskinesis. Another patient with left bundle-branch block showed asynergy. Mild to moderate mitral incompetence occurred in 10 patients.

A right atrial angiogram was made in the patient who had right coronary artery obstruction. This showed poor contraction of a dilated right atrium with mass transfer of contrast medium into the right ventricle during the rapid filling phase, a clear gradient in contrast density between atrium and ventricle in diastole as a consequence of a high ventricular end-diastolic pressure, and large apical areas of stagnation in the ventricle, implying right ventricular infarction.

Aortography has been used to study coronary arteries. We found this technique inadequate and have not included such patients in our study. None the less, aortography provides a useful indication of cardiac output and ejection fraction in primary myocardial disease, with extremely slow clearance of contrast medium from the ascending aorta in the more severe cases. Similarly when the stroke output is low, contrast clears slowly from the coronary vessels in selective coronary angiography.

Discussion

Coronary arteriography was undertaken in 16 patients with primary myocardial disease to exclude coronary atheroma. Ten of the 16 patients had left anterior hemiblock and one had left bundle-branch block presumably as manifestations of myocardial fibrosis. Eleven patients had evidence of left ventricular hypertrophy on electrocardiograms, often with a long narrow posteriorly directed spatial

FIG. 5 Aortogram in the left anterior oblique view of a patient with severe pulmonary arterial hypertension and right ventricular involvement. The right coronary artery in the right atrio-ventricular groove demonstrates a dilated tricuspid annulus.

vector loop simulating myocardial infarction. Only 2 patients showed coronary artery obstruction and in both the lesion suggested embolism rather than coronary atheroma. None showed evidence of diffuse coronary disease (Raftery, Banks, and Oram, 1969; Richardson and Gotsman, 1971). Furthermore, our experience with coronary arteriography in other Bantu subjects has shown that atheroma is extremely rare in this racial group.

The coronary arteriogram provided a three-dimensional view of the skeleton of the heart. Though this fact is self-evident, it has not been used widely to study ventricular size and function (McDonald, 1970; Kong, Morris, and McIntosh, 1971).

The coronary arteries were dilated and the total cross-sectional area of the proximal vessels correlated with left ventricular wall mass. If the velocity of coronary arterial flow is unchanged, then compensatory dilatation of the arteries maintains blood

flow to the augmented muscle mass. Angiographic left ventricular muscle mass measures the total volume of the left ventricular wall but may not indicate the actual mass of actively contracting sarcomeres. It is possible that part of the increase in wall volume is the result of an increase in mitochondrial size, intra- and intercellular oedema, and the presence of excess fibrous tissue.

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FIG. 6 This left coronary angiogram in the left anterior oblique view shows a large angle at the bifurcation of the left coronary artery into its anterior descending and circumflex branches, with splaying of the circumflex branch around a dilated mitral annulus. Pleural effusions are seen in the horizontal and oblique interlobar fissures.

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FIG. 7 This analysis of movement of the circumflex artery as seen in the right anterior oblique view shows the importance of diastolic events in the cardiac cycle in primary myocardial disease. RFP=rapid filling phase; AFW=atrial filling wave. The most rapid movement of this vessel is clearly seen to occur during diastole in this patient. Percentage movement compares the position of a fixed point on the vessel studied to its position at end-diastole (0%) and end-systole (100%).

FIG. 8 The total cross-sectional area of the coronary arteries is related to LV muscle mass. If Case 1, with extensive anteroseptal infarction secondary to coronary embolism, is excluded, then the two parameters are related ($n=17$; $r=0.50$; $P<0.05$).

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