Dual isotope stress testing in congenital atresia of left coronary ostium

Applications before and after surgical treatment

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SUMMARY A 38-year-old woman presented with an 11-year history of angina pectoris. Coronary arteriography disclosed a large right coronary artery which filled the entire left coronary tree retrogradely. The left main coronary artery ended blindly and was not connected to the aortic root. There were no atherosclerotic lesions in any vessel. Exercise thallium-201 scintigrams showed a perfusion defect in the anterior region of the left ventricle and exercise first pass radionuclide ventriculography showed anterior hypokinesis of the left ventricle with an ejection fraction of 54 per cent, compared with 60 per cent at rest.

An aortocoronary saphenous vein graft was constructed to the left coronary artery. Four months after operation the patient is free from symptoms. Repeat thallium scintigrams were normal. Exercise radionuclide ventriculography after operation disclosed no wall motion abnormality, and ejection fraction on exercise was 70 per cent.

The mechanism of angina in this patient is unclear but may have been related to the abnormal timing of delivery of blood to the left ventricular myocardium. Dual radionuclide stress testing showed abnormalities of perfusion and wall motion associated with this anomaly, and the reversal of these abnormalities after operation. This non-invasive approach may be useful in the assessment of the physiological significance of coronary anomalies and of the value of corrective surgery.

Single coronary artery and origin of both coronary arteries from the same sinus of Valsalva have generally been regarded as minor congenital abnormalities of little clinical significance. In the absence of associated severe congenital cardiac lesions the anomalies are compatible with longevity.

The widespread use of coronary arteriography has led to the more frequent recognition of these anomalies and an appreciation of their potential clinical significance. There are a number of case reports of sudden death in patients with single coronary artery or anomalous aortic origin of the coronaries. In addition, myocardial infarction and angina pectoris have been described in conjunction with certain anomalies. Myocardial perfusion scintigraphy with thallium-201 and radionuclide ventriculography have proved valuable in the non-invasive assessment of the physiological changes occurring with stress in patients with atherosclerotic coronary artery disease.

In this paper we describe the applications of these techniques in the detection of abnormal physiology in a patient with atresia of the left coronary ostium, and the reversal of the abnormalities after surgical treatment.

Case report

A 38-year-old Caucasian housewife was admitted to hospital complaining of diarrhoea, abdominal pain, malaise, nausea, and vomiting. She had previously attended another hospital 19 years ago with similar complaints and a diagnosis of ulcerative colitis had been made, but she defaulted from follow-up. On systematic inquiry she admitted to anginal chest pain for the past 11 years, and as a child she had frequent ‘stitch-like’ chest pains on exertion. She
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had been seen at another hospital 10 years previously with chest pains, and had a positive exercise electrocardiogram. At that time her oral contraceptives were withdrawn and anticoagulant treatment was started. This was discontinued two years later.

On her current admission she was referred to the cardiac department for assessment. On examination she was in sinus rhythm and normotensive. Venous pressure was normal. On auscultation there was a mid-systolic click and late systolic murmur at the apex suggesting prolapse of the posterior cusp of the mitral valve. Electrocardiogram and chest x-ray film were within normal limits. An echocardiogram failed to show a systolic prolapse of a mitral cusp. During exercise testing the patient developed chest pain after a total of 6780 joules on the bicycle ergometer. Exercise electrocardiogram (Fig. 1) was positive with 2 mm ST segment depression. In addition, there was variable voltage alternation of the T wave.15

At cardiac catheterisation, intracardiac pressures were normal. Left ventricular angiogram was normal with no prolapse of the mitral valve. At coronary arteriography, the left coronary ostium could not be found. The right coronary arteriogram (Fig. 2) showed a large calibre vessel, with the posterior descending artery continuing around the apex and filling the anterior descending directly retrogradely. The remainder of the left coronary tree was opacified via the left anterior descending. The left main stem terminated blindly and was not connected to the aorta. The large apical vessel connecting the posterior descending to the anterior descending is best seen in Fig. 2b.

Simultaneous aortogram and right coronary arteriogram confirmed that there was no connection between aorta and left main coronary. No obstructive lesions of the coronary tree were present.

In an attempt to decide whether this coronary anomaly per se could produce abnormal perfusion or wall motion, exercise thallium-201 scintigraphy and exercise first pass radionuclide ventriculography were performed. For stress thallium imaging,
exercise was carried out upright and 2 mCi of the tracer were injected at the onset of angina pectoris. Exercise was continued for a further 60 seconds. Images were obtained with an Ohio nuclear gamma camera and a high resolution parallel-hole collimator, in four standard projections. Five hundred thousand counts per view were collected with a window extending from 65 to 90 KeV (mercury x-ray peaks). The camera was linked on-line to a Varian 620L computer. Images were processed by background correction and analysed according to the criteria of Lenaers et al.13 with perfusion defects regarded as those areas with less than 75 per cent of the maximum intramyocardial counts. Imaging was repeated four hours later at rest, after redistribution of the tracer. Fig. 3a and 3b are anteroposterior images on exercise and at rest. Fig. 3a shows a large perfusion defect anteriorly with normal perfusion inferiorly. At redistribution the defect had disappeared with only some thinning at the apex which is a normal variant.13

First pass radionuclide ventriculography was carried out in the right anterior oblique projection using a computerised multicrystal gamma camera (Baird-Atomic, System 77) as previously described.14 Exercise was supine, and the ventriculograms were performed at angina and after a 20-minute rest period. This technique has shown to be sensitive for detecting falls in ejection fraction and abnormal wall motion on exercise in patients with obstructive coronary artery lesions.11 Fig. 4a shows superimposed end-diastolic and end-systolic perimeters from rest (left-hand) and exercise studies, respectively. Ejection fraction measured by count rate changes was 60 per cent at rest and 54 per cent on exercise, with exercise-induced hypokinesia anteriorly.

Fig 4b shows computer derived images of regional ejection fraction, which represent in a 16-colour isocount display the relative contribution of different zones of the left ventricle to ejection
fraction. The end-diastolic perimeter is added in each case. The colour scale ranges from black, which represents the minimum counts per crystal, to yellow, which represents the maximum counts. Thus the left ventricular zones that contribute most to ejection fraction are represented by yellow and red. The resting (left-hand) image of Fig. 4b shows homogeneous contribution to ejection fraction with the yellow and red predominant in all zones. At exercise the inferior wall shows maximal regional ejection fraction but the contribution of the anteropapical segment is poor.

On the basis of these results, the patient was referred for operation, when the right coronary artery was found to be a large vessel situated in the normal position. The distal left anterior descending was abnormally large and tortuous. The aorta was opened to assess the left coronary ostium. There was no left coronary orifice, but a blind dimple in the normal ostial position. An aorto-left anterior descending coronary artery saphenous vein bypass graft was constructed. Postoperative recovery was uneventful.

Two months later the stress testing was repeated. Exercise testing produced fatigue but no chest pain at a heart rate of 145 beats per minute and the exercise electrocardiogram showed no ischaemic changes (Fig. 1). Repeat stress thallium scintigraphy showed no exercise-induced perfusion abnormality (Fig. 3c).

Radionuclide ventriculography was carried out at the point of exercise-induced fatigue. Fig. 5a shows superimposed end-diastolic and end-systolic perimeters at rest (left-hand image) and exercise. No hypokinesis was demonstrable, and ejection fraction on exercise was 70 per cent. Resting ejection fraction was unchanged. Fig. 5b shows the regional ejection fraction images postoperatively; the right-hand image indicates a return to normal of the regional ejection fraction at exercise.

After completion of these investigations, coronary arteriography was repeated. Selective right coronary injection did not result in retrograde filling of the left coronary tree (Fig. 6a) and the large apical connecting vessel shown in Fig. 2 was not visible. The left anterior descending graft was shown to be patent and filled the left coronary system (Fig. 6b). Four months after operation the patient remains free from symptoms.

Discussion

Congenital abnormalities of the coronary arteries are among the rarest of cardiac conditions that may have practical clinical significance. Despite some assertions that coronary anomalies are of only minor importance, it has become increasingly evident that a variety of syndromes, from angina pectoris to sudden death, may be associated with abnormal origin of one or more coronary arteries. In many instances symptoms have been attributed to the abnormal course of the aberrant vessel between the aorta and pulmonary artery, or to the acute angulation of the vessel with the aorta so that the coronary orifice is slit-like. The case reported here is rare even among the spectrum of coronary anomalies, inasmuch as two normal coronary arterial systems were present. Mullins et al. described a 10-year-old child with left coronary ostial atresia and retrograde filling of the left system via collaterals from the right coronary. They drew a distinction between this and the expected anterograde filling of single coronary artery branches. Their patient had clinical and electro-
cardiographic evidence of myocardial ischaemia with a positive exercise electrocardiogram, which reverted to normal after saphenous vein grafting. Jokl et al.\(^5\) described sudden death in a young woman whose entire left ventricular blood supply came from the posterior descending branch of the right coronary, no trace of the left coronary being found. Despite the potentially catastrophic events that can complicate congenital coronary anomalies, the opportunities to study cardiac physiology during stress have been few. Chaitman et al.\(^7\) showed abnormal lactate production during pacing and reduced coronary blood flow during exercise in some patients with anomalous origin of the left coronary. Pachinger et al.\(^8\) reported deterioration of left ventricular function during pacing-induced angina in another patient.

More recently, thallium-201 perfusion scintigraphy has been applied to the diagnosis of anomalous left coronary artery in the young\(^9\) and in the serial assessment of myocardial perfusion in cases where the left coronary artery arises from the pulmonary artery.\(^8\) This report extends the use of radionuclide techniques to a more unusual coronary anomaly. The defect in anterior perfusion on exercise was coupled with a localised anterior contraction abnormality.

The mechanism of angina in this patient is unclear. One cannot postulate any of the mechanical possibilities previously mentioned such as acute angulation of the coronary with the aorta. It is possible that abnormal timing of delivery of blood to the left coronary territory contributed to angina.

The long path taken by blood destined for the left coronary territory may have led to the arrival of the blood in systole, rather than in diastole as is usual. Systolic coronary compression during exercise may have reduced the available blood supply to the anterior wall of the left ventricle to a critical level.\(^21\)

The relief of symptoms after the surgical creation of a two coronary system was given objective support by the postoperative demonstration of disappearance of electrocardiographic, perfusion, and wall motion abnormalities. As in the case of Mullins et al.\(^18\) improvement was associated with a patent graft and absence of retrograde flow into the left coronary. Successful surgical correction of other coronary anomalies has been reported\(^22\) either by saphenous vein grafting or enlargement of the origin of the anomalous vessel.

It is not known at present whether surgical correction of the type of anomaly described here will be effective in preventing infarction or sudden death in the long term. The reversal of impaired perfusion and abnormal ventricular function under stress is objective evidence that surgical correction is beneficial, and may indicate that direct myocardial revascularisation is the operation of choice in this condition. Stress perfusion scintigraphy and radionuclide ventriculography appear well suited to the non-invasive evaluation of these anomalies.

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Fig. 6 (a) Right coronary arteriogram after vein grafting, showing no retrograde filling of the left coronary tree, and absence of the apical connecting vessel. (b) Selective injection into the left coronary artery graft. The graft is patent and fills the left anterior descending and circumflex arteries anterogradely.
References


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