

# Syphilitic coronary ostial stenosis

## Case reports

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### Summary

Two young Coloured men with proven syphilitic coronary ostial stenosis had severe angina pectoris unresponsive to conventional medication. One underwent an aortic valve replacement for severe aortic insufficiency associated with subtotal ostial occlusion of the right coronary artery (RCA), which was corrected by an aortocoronary bypass graft; the left coronary artery (LCA) ostium was normal and patent. The other patient had total occlusion of the LCA ostium which resulted in an extensive transmural anteroseptal and anterolateral myocardial infarction; the RCA ostium was unaffected and the aortic valve appeared normal. He was considered unsuitable for cardiac surgery and continued to receive anti-anginal drug therapy with quite satisfactory improvement in symptoms.

Non-atheromatous coronary artery disease must always be sought for and excluded when a non-White patient presents with symptoms of ischaemic heart disease. Although atheromatous coronary artery involvement is becoming increasingly prevalent among 'westernized' Black and Coloured subjects, it is still relatively unusual in comparison with the extremely high incidence in the White population.

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## Case reports

### Case 1

A 37-year-old Coloured fisherman was known to have suffered an acute transmural anteroseptal and anterolateral myocardial infarction some 2 years previously. He had been a moderate cigarette smoker for a long time, but there was no evidence of hyperlipoproteinaemia, hypertension or diabetes mellitus. After his initial discharge from Tygerberg Hospital he was symptom-free and was therefore given no drug therapy. Approximately 6 months before his present admission to the Intensive Coronary Care Unit (ICCU) at Tygerberg Hospital he began experiencing classic effort angina, for which he consulted his local practitioner. At that stage he was given sublingual and oral isosorbide dinitrate (Isordil), as well as 10 mg nifedipine (Adalat) 3 times

daily. Initial relief of his symptoms proved to be somewhat short-lived and he soon began complaining of frequent angina after relatively minor exertion, in addition to angina at rest. These symptoms became far more severe in duration and frequency and were associated with dyspnoea, nausea and sweating. His general practitioner then urgently referred him to Tygerberg Hospital with a diagnosis of unstable angina. The patient had no other symptoms of note.

On admission to the ICCU in July 1982 he was a rather thin young man with no features of hyperlipoproteinaemia and a blood pressure of 130/70 mmHg. The radial pulse was normal and the jugular venous pressure was not elevated. There was no clinical evidence of cardiomegaly and the only abnormal auscultatory feature was a prominent fourth heart sound. Cardiac failure was absent and the rest of the examination revealed no further abnormal physical signs. On radiography the cardiac silhouette was normal and the lung fields were clear. The resting ECG (Fig. 1a) demonstrated sinus bradycardia of 47/min, a P-R interval of 0,15 second and a mean QRS axis of  $-36^\circ$ . There was evidence of an old transmural anteroseptal and anterolateral myocardial infarction as well as borderline left ventricular hypertrophy.

The patient was considered to have crescendo angina and was treated with high doses of nitrates as well as nifedipine. The angina was controlled well with bed rest and this therapy. The biochemical and haematological values were within normal

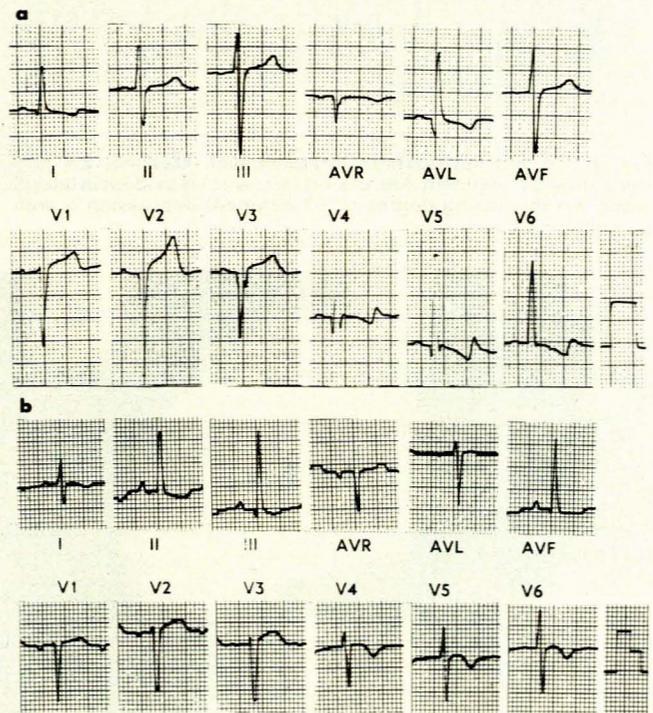


Fig. 1. a — case 1. Resting ECG showing normal P-R interval and left axis deviation of  $-36^\circ$ . Features of an old transmural anteroseptal and anterolateral myocardial infarction and borderline left ventricular hypertrophy are seen. b — case 2. Resting ECG: left atrial and left ventricular hypertrophy is demonstrated, and there is a normal P-R interval and mean QRS axis.

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limits but all the serological investigations for syphilis returned strongly positive results. Collagen screening tests were all negative. A few days later it was decided to carry out a graded submaximal treadmill stress test (modified Bruce protocol) to detect underlying myocardial ischaemia. Some 3 minutes after the commencement of effort the patient complained of severe angina pectoris. His target heart rate was achieved and an adequate blood pressure response was recorded. Soon after the onset of angina a maximum of 2,4 mm horizontal ST-segment depression was recorded in the lateral leads (Fig. 2). The patient was given sublingual isosorbide dinitrate, which produced rapid relief of his chest pain as well as normalization of the ischaemic electrocardiographic changes. A gated blood pool scintigram revealed the presence of diffusely poor left ventricular contractility with akinesia of the apex, and a reduced ejection fraction of 25%. Contraction of the right ventricle was normal. A diagnosis of left coronary ostial stenosis (probably syphilitic), having given rise to a previous large anterior myocardial infarction, was made.

In view of the clinical presentation, possible involvement of the right coronary artery (RCA) ostium was entertained, and the

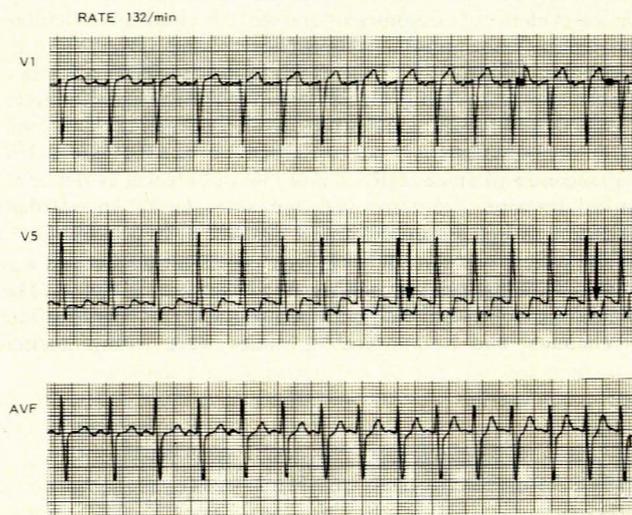


Fig. 2. Graded submaximal treadmill test. Maximal 2,4 mm horizontal ST-segment depression (arrowed) is evident in lateral leads. An increasing degree of ST-segment depression is well demonstrated.

patient was subjected to cardiac catheterization and selective coronary arteriography. The Seldinger technique via the right femoral artery and vein was employed using 7F Goodale-Lubin and pigtail catheters. The cardiac output was normal at 6,31/min with a resultant cardiac index of 3,6 l/min/m<sup>2</sup>. However, a low left ventricular dp/dt of 939 mm/s was recorded. A left ventricular cine angiogram in the right anterior oblique (RAO) projection delineated marked akinesia of the anterolateral and anteroseptal segments secondary to previous myocardial infarction (Fig. 3). The rest of the left ventricle contracted normally and there was no evidence of mitral regurgitation or prolapse. An aortic cine angiogram in the left anterior oblique (LAO) projection demonstrated a competent tricuspid aortic valve. Striking irregularity of the lumen of the ascending aortic wall was noted without calcification (Fig. 4a). Filling of the left coronary artery (LCA) could not be demonstrated, indicating complete occlusion of this vessel at its origin from the aorta (Fig. 4b). In contradistinction, the RCA was seen to fill rapidly and no lesions were seen either at its ostium or along its entire course. Selective coronary arteriography in multiple projections, using 7F Judkins catheters, delineated a normal dominant RCA (Fig. 5). In addition, the RCA provided retrograde filling of the LCA system by means of right-to-left collaterals; a normal-appearing LCA was seen to fill to just distal to its origin from the aorta (Fig. 5). This confirmed the presence of LCA ostial stenosis secondary to syphilitic aortitis. The patient experienced no angina during cardiac catheterization.

Since the RCA was not involved by the chronic syphilitic inflammatory process, and since the patient had already suffered a large myocardial infarction resulting in poor left ventricular function, surgery could not be contemplated. He was therefore given a course of penicillin and discharged on maximal oral doses of long-acting nitrates and nifedipine. He was later seen on several occasions at the Cardiac Clinic at Tygerberg Hospital, complaining of very infrequent angina experienced on effort. Repeated resting ECGs showed no change from the first one (Fig. 1a).

## Case 2

This 36-year-old Coloured farmworker was asymptomatic until 6 months before admission to the ICCU in early 1978. His main complaints were of dyspnoea on minimal effort, orthopnoea, paroxysmal cardiac dyspnoea and decubitus angina pectoris as well as effort-induced angina. There was no previous history of

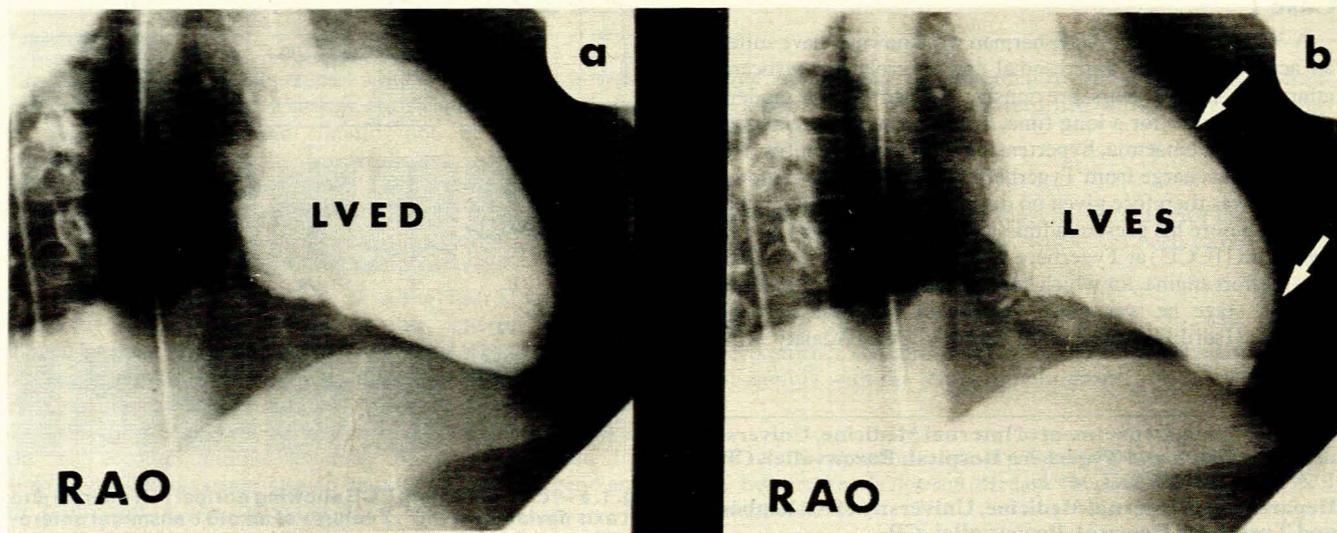


Fig. 3. Left ventricular cine angiograms in the RAO projection. Anteroseptal and anterolateral akinesia is delineated (arrowed), and no mitral insufficiency or prolapse is seen (LVED = left ventricle in end-diastole; LVES = left ventricle in end-systole).

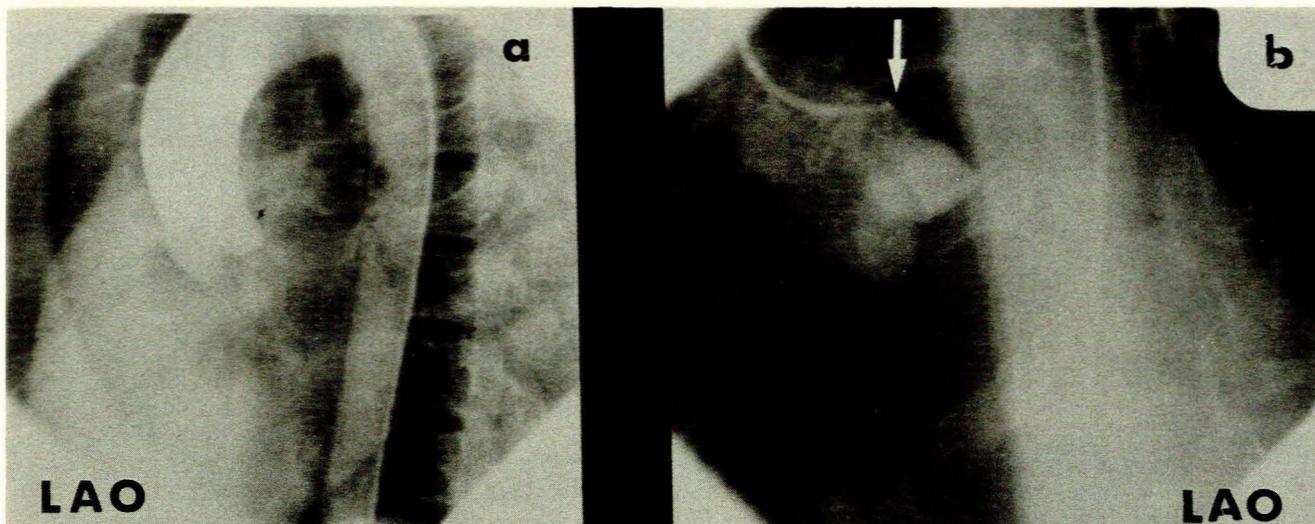


Fig. 4. Aortic cine angiograms in the LAO view. There is no aortic insufficiency and the aortic wall is very irregular but not calcified (a). Occlusion of the LCA ostium (arrowed) is clearly seen (b).

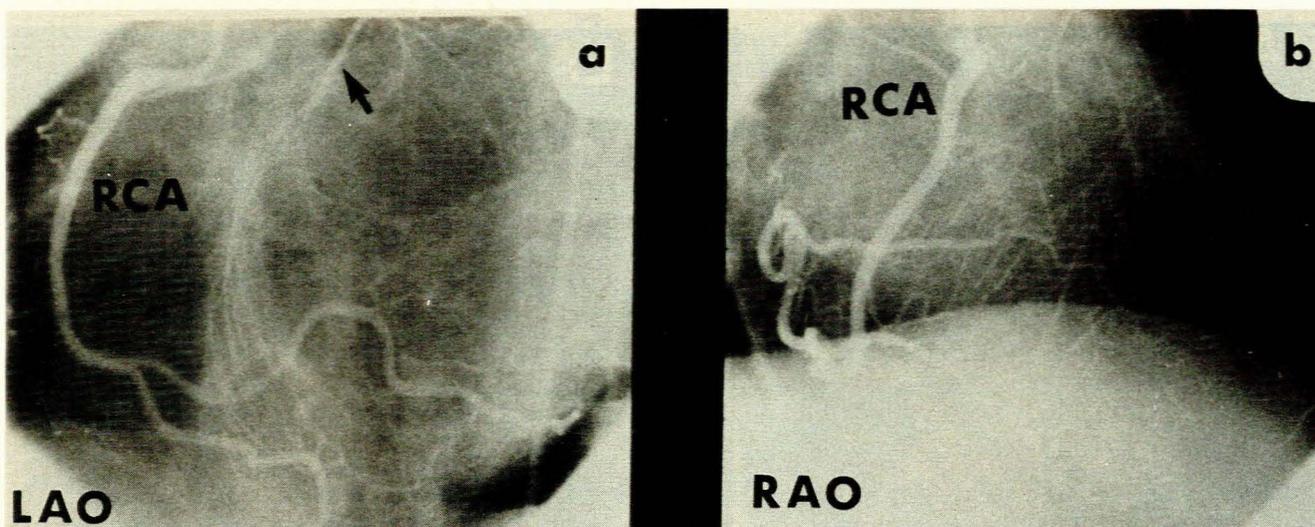


Fig. 5. RCA cine angiograms in (a) the LAO view, and (b) the RAO view. The totally occluded LCA fills retrogradely (arrowed) from the dominant RCA.

acute rheumatic fever and his only risk factor for ischaemic heart disease was moderate cigarette smoking. Clinical examination revealed the features of severe aortic regurgitation complicated by left ventricular cardiac failure. There were no signs of hyperlipoproteinaemia or any other abnormal features. A chest radiograph confirmed the presence of an enlarged left ventricle and left atrium, a dilated and unfolded ascending aorta which was not calcified, and signs of moderate pulmonary oedema. The resting ECG (Fig. 1b) showed sinus tachycardia of 110/min, a P-R interval of 0,14 second, a mean QRS axis of  $+85^\circ$ , and features of left ventricular hypertrophy. The serum biochemical and haematological profiles were within normal limits. Serological tests for syphilis were all significantly positive. A diagnosis of severe syphilitic aortic regurgitation with possible additional coronary ostial narrowing was therefore made. The patient was given the conventional therapy for cardiac failure as well as treatment for syphilis. Response was most satisfactory.

A few weeks after admission the patient was subjected to cardiac catheterization and coronary angiography using the Seldinger technique from the groin. Haemodynamic evaluation

showed that left ventricular cardiac failure was no longer evident. Left ventricular cine angiography in the RAO projection (Fig. 6) revealed a markedly dilated ventricle with adequate contractility and no mitral regurgitation. An aortic cine angiogram in the LAO projection (Fig. 7b) demonstrated severe, non-calcific, aortic regurgitation. The ascending aorta was also grossly distorted with some constriction but no calcification in the wall. The left coronary ostium was also normal (Fig. 7a). Selective injection of dye into the LCA (Fig. 8) showed this vessel to be completely normal. However, the ostium of the RCA could not be entered by the catheter owing to subtotal occlusion. Nonselective coronary angiography confirmed the presence of severe ostial stenosis of the dominant RCA with a normal-appearing vessel distal to the stenosis (Fig. 9).

In view of the cardiac catheterization findings the patient was operated upon in May 1978. The ascending aortic wall was exceptionally thick and fibrotic with a resulting supra-aortic stenotic ring. The LCA ostium was inferior to this ring and was normal and patent, but the RCA ostium was almost entirely occluded by the chronic syphilitic aortitis. The aortic valve was

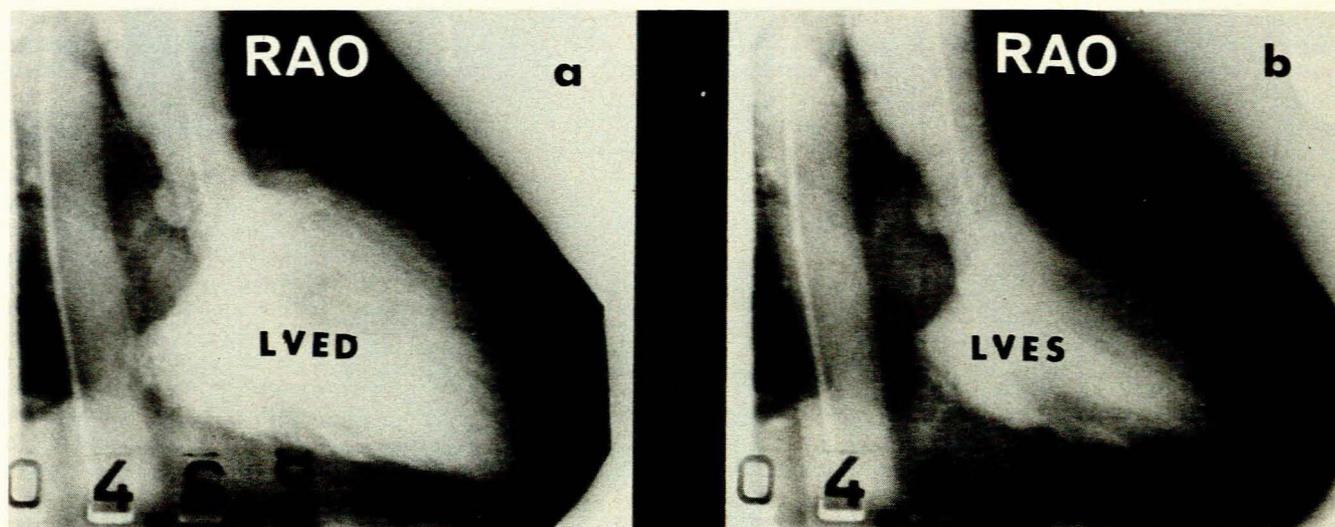


Fig. 6. Left ventricular cine angiograms in the RAO projection. There is marked chamber dilatation with good contractility and no mitral insufficiency.

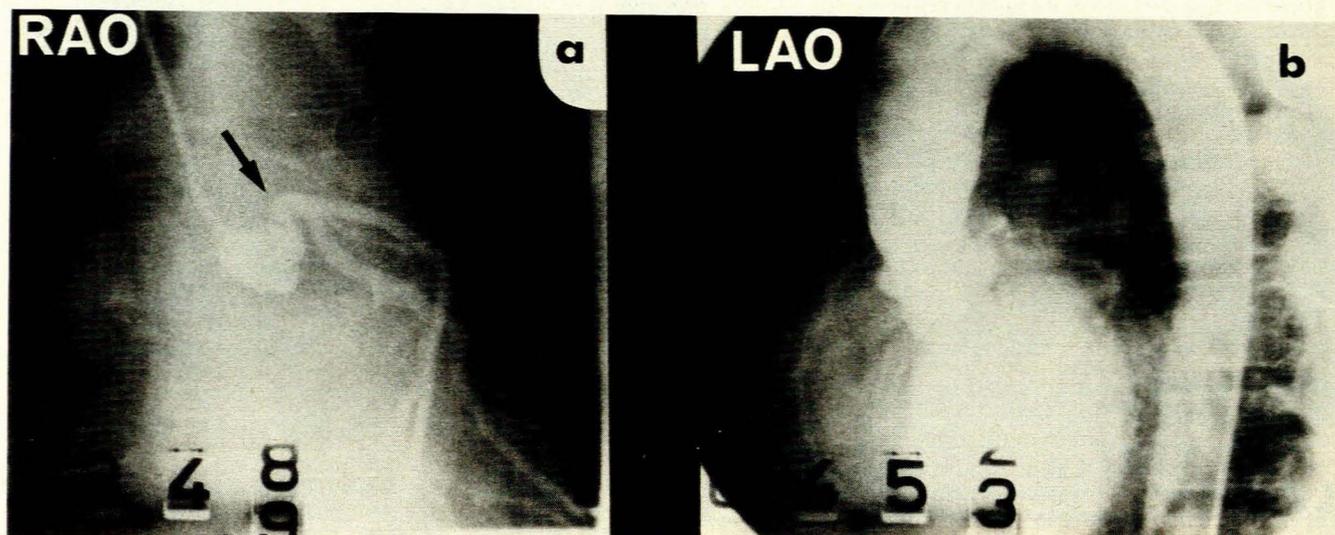


Fig. 7. (a) Subselective LCA injection in the RAO projection; the LCA ostium (arrowed) is normal. (b) Aortic cine angiogram in the LAO view showing gross, non-calcific aortic regurgitation; irregularity and dilatation of the ascending aortic wall is visualized.

freely incompetent and was not calcified. This valve was therefore excised and replaced by a No. 21 Carpentier bioprosthesis. An aortoplasty, using woven Dacron, was also performed. Finally, an aortocoronary saphenous vein bypass graft was inserted into the proximal RCA.

The patient made a most satisfactory recovery postoperatively and was discharged on low-dose diuretic and vasodilator therapy. He was able to return to his fairly strenuous employment without experiencing undue effort intolerance. Long-term follow-up at the Cardiac Clinic found him to be free from signs of left ventricular failure; his bioprosthesis was normal on clinical examination. In addition, he no longer complained of angina pectoris and repeated ECGs were essentially the same as the pre-operative tracing.

## Discussion

Syphilis can affect the cardiovascular system in several ways,<sup>1</sup> aneurysm of the aortic arch with its resulting complications

probably being the best known manifestation. This is accepted as following an aortitis, and the patient presents several decades after the initial infection.<sup>2</sup> Involvement of the aortic valve results in aortic insufficiency which, with increasing haemodynamic severity, causes decreasing effort tolerance as well as angina pectoris. Acute luetic myocarditis (either focal or diffuse) with the histological features of gummatous lesions may result in complete heart block and is exceptionally uncommon. Classically, the coronary arteries are involved in the disease process because the aortitis has given rise to varying degrees of coronary ostial stenosis.<sup>3-5</sup> This rare lesion may or may not be accompanied by aortic insufficiency, a combination which can make the assessment of angina pectoris difficult. It is usually stated that angina disproportionate to the degree of aortic insufficiency in a young person should make syphilitic coronary ostial stenosis a strong likelihood. The existence of an actual diffuse coronary arteritis secondary to syphilis has few supporters.

Scharfman *et al.*<sup>6</sup> reviewed several autopsy series and reported on a total of 935 cases of luetic aortitis, of which only 24% showed evidence of varying degrees of coronary ostial involvement. They

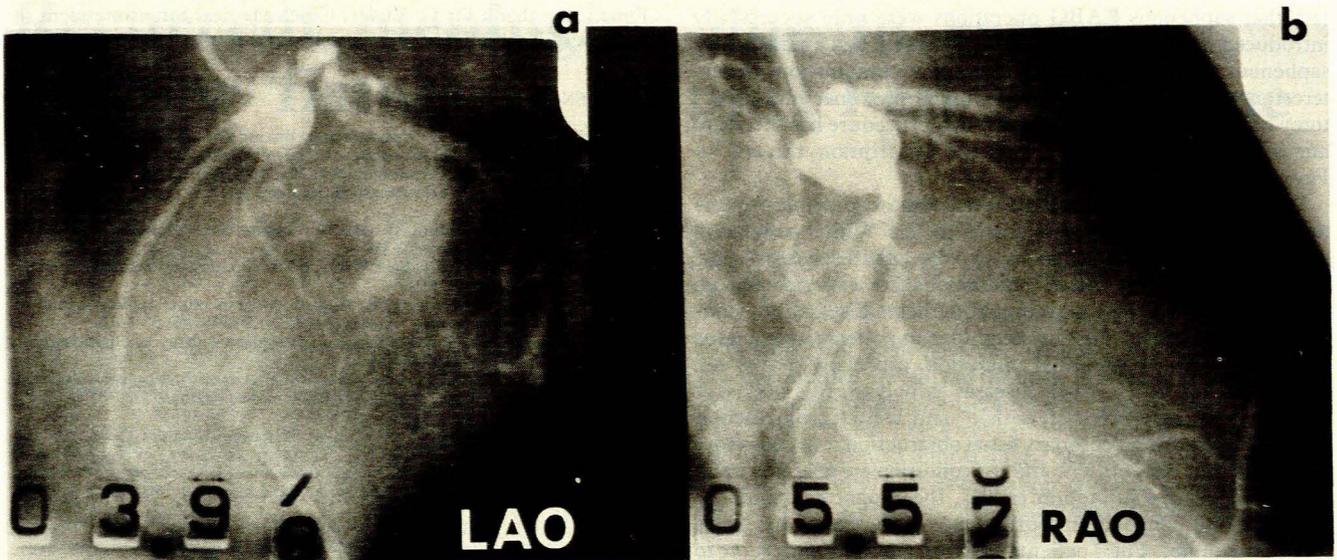


Fig. 8. Selective cine angiograms of the LCA in (a) the LAO, and (b) the RAO projections. No lesions are visualized.

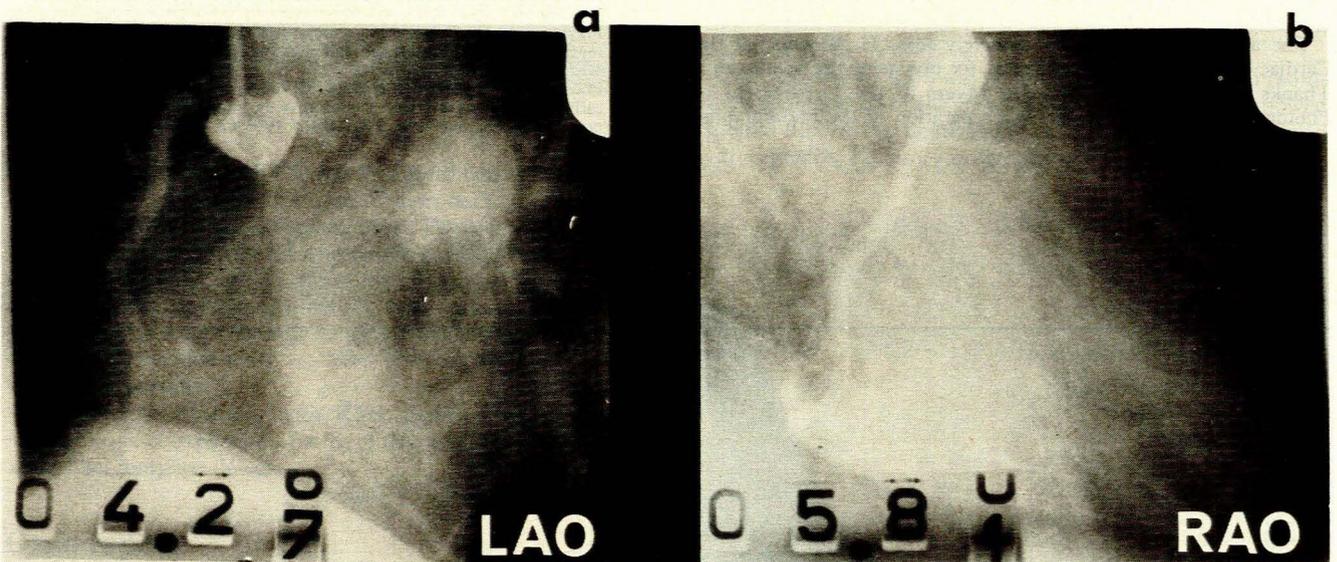


Fig. 9. Subselective cine angiograms of the RCA in (a) the LAO, and (b) the RAO views. Subtotal ostial obstruction is present; the remainder of the vessel appears normal.

also noted the exceptional rarity of acute myocardial infarction in cases of syphilitic coronary ostial narrowing, attributing this to the lapse of sufficient time for development of an adequate collateral circulation. Burch and Winsor<sup>7</sup> found myocardial infarction in only 3 (0,9%) of a total of 326 cases of myocardial infarction due to a variety of causes.

Heggtveit<sup>2</sup> documented a 26% incidence of coronary ostial stenosis among 100 cases of syphilitic aortitis seen at autopsy. In the majority involvement of both ostia and atherosclerosis was an associated factor. Myocardial infarction was seen in 24 cases, 4 due to syphilitic coronary ostial occlusion and a further 3 secondary to a combination of atherosclerosis and lues. In 1 case there was evidence of syphilitic coronary arteritis, whereas 5 of the patients had died suddenly and failed to show any sign of acute myocardial infarction at autopsy.

Schrire *et al.*<sup>5</sup> noted that syphilis was responsible for 0,6% of all cases of cardiovascular disease seen at Groote Schuur Hospital, Cape Town. Syphilis accounted for 1 - 2% of cases of heart disease among non-White patients, a figure stated to be 10

times greater than that for White patients. Furthermore, these authors stated that 5 out of 150 patients with syphilitic heart disease had undoubted coronary ostial stenosis.

More recently Rissanen<sup>8</sup> has described an autopsy series of 255 cases, comprising deaths from acute myocardial infarction, sudden deaths without evidence of recent myocardial infarction, and violent deaths. Ostial stenosis was noted in 50% but only 1 death was definitely due to syphilis. Shichida *et al.*<sup>9</sup> studied the cardiovascular complications of syphilis and noted an almost equal incidence of myocardial infarction in elderly patients with negative (1,65%) and positive (1,67%) serological tests.

The first attempt at endarterectomy for syphilitic coronary ostial stenosis was made in 1960 by Dubost *et al.*<sup>10</sup> after the introduction of cardiopulmonary bypass and hypotensive techniques. Further endarterectomy procedures used for this condition were subsequently reported by Connolly *et al.*<sup>11</sup> and Beck *et al.*<sup>12</sup> As far as the present authors are aware, there is no report of an aortocoronary bypass graft (CABG) utilizing the saphenous vein in syphilitic coronary ostial stenosis. This may well be due to the

fact that saphenous CABG operations were only successfully introduced in 1968 by Favaloro.<sup>13</sup> Our case 2 had a satisfactory saphenous CABG without any attempt at simultaneous endarterectomy. Many surgeons have avoided proximal endarterectomy because of the complications; this procedure has therefore not often been employed since the description thereof by Favaloro.

Ischaemic heart disease has assumed almost epidemic proportions in South Africa, particularly in the White population group, in which the cause is usually atherosclerosis. The Coloured and urban Black populations are experiencing an increasing incidence of atherosclerotic heart disease, but non-atheromatous causes must be sought for in patients with angina pectoris or acute myocardial infarction, especially if they are young and not White. Such diseases as polyarteritis nodosa,<sup>14</sup> 'coronary intimal fibrous stenosis',<sup>15</sup> and luetic cardiovascular disease with resultant coronary ostial obstruction must therefore be excluded. If patients with the last condition are operated upon before myocardial infarction ensues, the prognosis should be good whether concomitant aortic valve replacement has to be carried out or not. The 2 patients described in this report serve to illustrate these important points.

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## News and Comment/Nuus en Kommentaar

### JAMA Centennial

While the American Medical Association is very much our senior, their journal *JAMA*, like the Medical Association of South Africa, first saw the light precisely a century ago. With considerable difficulty the present editorial staff have succeeded in producing a replica of the first number, that of 14 July 1883, in the issue of 8 July 1983, together with other items from *JAMA* in the past.

It had been decided in 1883 that a weekly journal should take the place of the annual volume of AMA transactions, and the new journal sold at 10 cents a copy (free to members) and was edited by a formidable character, Dr Nathan S. Davis, who had been a good AMA man even before the Association's foundation 36 years previously. Davis served for only 5 years but lived on until the age of 87. Of the 13 editors to date none has exceeded in colour and personality the famous Morris Fishbein, who became the unappointed spokesman for the entire Association and the best-known doctor in the USA.

*JAMA* will publish in forthcoming issues what are considered its 50 best articles, beginning with Duhring's classic description in 1884 of dermatitis herpetiformis. The Centennial edition of *JAMA* also includes one reject, 'The gynecologic consideration of the sexual act', by Denslow Lewis, in which he dared to suggest (in 1899) that women were intended to enjoy intercourse. The editor turned it down as obscene and all appeals to higher authority failed to reverse this verdict. When the paper was first presented, one eminent gynaecologist hoped that it was not going to be discussed in detail for 'its discussion is attended with more or less filth and we besmirch ourselves by discussing it in public'. This at a time when according to Lewis most men received their sexual initiation from prostitutes.

*JAMA* now has a circulation of 322 000 copies in English together with 121 000 in five other languages in six countries. Congratulations to the *Journal* and its present editor, George Lundberg, are perhaps superfluous but nevertheless tendered by its smaller South African counterpart.