

Coronary artery ectasia—a variant of occlusive coronary arteriosclerosis

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SUMMARY In a study of 1000 consecutive coronary arteriograms, 12 patients (all men) had coronary artery ectasia.

Ectasia was found most frequently in the circumflex or right coronary artery. Only 1 patient had ectasia in the left anterior descending coronary artery. In 11 patients, ectasia of one artery was associated with severe stenosis or occlusion of other vessels, typical of arteriosclerosis. Histology from an ectatic segment in one of this group showed changes of severe arteriosclerosis with extensive intimal fibrosis and destruction of the media. One patient had a mixed collagen vascular disease. Measurement of coronary sinus flow in 2 patients with coronary artery ectasia showed flows in the range of patients with non-ectatic coronary artery disease. At cardiac surgery flows down the grafts to ectatic arteries were in the same range as in grafts to non-ectatic vessels.

Patients with coronary artery ectasia should be anticoagulated.

Previous reports of pathological dilatation of the coronary arteries have described the abnormal dilatation as an aneurysm, whether saccular or fusiform (Scott, 1948; Daoud *et al.*, 1963; Befeler *et al.*, 1977). Since the dilatation may be diffuse and involve the majority of the artery, it is more appropriate to describe the lesion as ectatic (Markis *et al.*, 1976) rather than aneurysmal.

The incidence of coronary artery ectasia seems remarkably constant. Daoud in a review of 694 necropsies found an incidence of coronary artery aneurysms of 1.4 per cent. Markis *et al.* (1976) in a series of 2457 coronary arteriograms showed 30 patients with ectatic coronary arteries (1.2%). Befeler *et al.* (1977) found 16 patients in a series of 1246 coronary arteriograms (1.3%).

Early necropsy studies suggested that congenital aneurysms were the most common type, with septic emboli also frequent (Scott 1948). More recent work suggests that arteriosclerosis is the commonest aetiological factor, with destruction of the media the underlying cause (Markis *et al.*, 1976).

This paper reports on 12 patients found to have coronary artery ectasia from a series of 1000 consecutive patients undergoing coronary arteriography. Coronary sinus flow and lactate metabolism

were measured in 2 of the 12 patients during atrial pacing. A biopsy of an ectatic coronary artery was obtained at the time of cardiac surgery in 1 patient.

Methods

Coronary arteriography was performed by the Judkins technique using a Siemens X Ray image intensifier system with a caesium iodide tube. The arteriograms were recorded in at least 2 planes on 35 mm film at 50 frames/s. In addition, biplane 35 cm or 100 mm cut film were recorded. The maximum coronary artery diameter assessed from the arteriograms was measured using the methods of Vieweg *et al.* (1976).

Coronary sinus flow was measured in 2 cases using the method of Ganz *et al.* (1971) and was compared with coronary sinus flow in 14 patients with non-ectatic coronary artery disease. A 7FG triple function thermodilution catheter (Wilton Webster Labs) was positioned in the coronary sinus via a left arm vein. Normal saline at room temperature was infused via a Harvard 906 infusion pump. The thermister outputs were fed through a CBA-210 Wheatstone Bridge to a Phillips 7 channel recorder. This technique of measuring coronary sinus flow primarily reflects left coronary artery flow (Rayford *et al.*, 1960) and depends on

Table Clinical data

Case No.	Age (y)	Sex	Ex-smoker	Symptoms	FH	Hypertension	Lipids	Previous infarct	Additional conditions
1	49	M	+	AP	-	-	N	-	Ulcerative colitis
2	50	M	+	AP	-	+	Type IV	-	Cerebral emboli
3	37	M	+	AP,D	+	-	Type IIa	-	-
4	51	M	+	AP	-	+	N	Anterior	-
5	71	M	+	D,AP syncope	-	-	N	-	MR (ant. chordal rupture)
6	56	M	+	AP,D	-	-	Type IV	Anterior	PA myxoedema CVD
7	49	M	+	AP,D	-	-	N	Anterior Inferior	-
8	58	M	+	VT	-	-	Type IV	-	PA
9	63	M	+	AP	-	-	N	Anterior	MR (post. chordal rupture)
10	52	M	+	AP,D	-	-	Type IIa	Anterior	-
11	53	M	-	D	+	-	N	Anterior	-
12	48	M	+	AP	-	-	Unknown	Inferior	Iliofemoral arteriomegaly

AP, angina pectoris; D, dyspnoea; MR, mitral regurgitation; PA, pernicious anaemia; VT, ventricular tachycardia; CVD, collagen vascular disease; FH, family history of coronary artery disease; N, normal.

good mixing of indicator (Bing *et al.*, 1972) which was infused at a high rate (48 to 50 ml/min) in these studies. Coronary sinus pacing was performed at increments of 20 bpm from 80 or 100 bpm to 160 bpm or to the onset of chest pain. Left ventricular and coronary sinus blood were sampled simultaneously at each pacing rate. The blood was instantly deproteinised in 10 per cent perchloric acid, centrifuged, neutralised, and the lactate measured with a Unicam SP 1800 spectrophotometer at pH 7.2 using a tris-hydrazine buffer, NAD, and lactate dehydrogenase (Hohorst *et al.*, 1959).

Abnormal lactate metabolism was defined as extraction of less than 10 per cent of arterial lactate (Cohen *et al.*, 1966).

Results

The clinical details of the patients are summarised in the Table. All 12 patients were men aged between 37 and 71. Eleven had previously smoked more than 20 cigarettes daily.

Ten presented with angina, 1 with exertional dyspnoea, and 1 with paroxysmal dyspnoea caused by ventricular tachycardia. Two had a family history of coronary artery disease and two were hypertensive. Seven had had a myocardial infarct before study, and 5 had hyperlipoproteinaemia on lipoprotein electrophoresis (2 were Fredrickson Type IIa and 3 were Type IV). Cases 5 and 9 had mitral regurgitation resulting from anterior and posterior chordal rupture, respectively. Pernicious anaemia was present in 2 cases, 1 of whom had a mixed collagen vascular disease. One patient (case 12) also presented with ischaemic symptoms in the right leg and translumbar aortography disclosed iliofemoral arteriomegaly (see Fig. 5).

Fig. 1 shows that ectasia was most commonly seen in the circumflex and right coronary arteries.

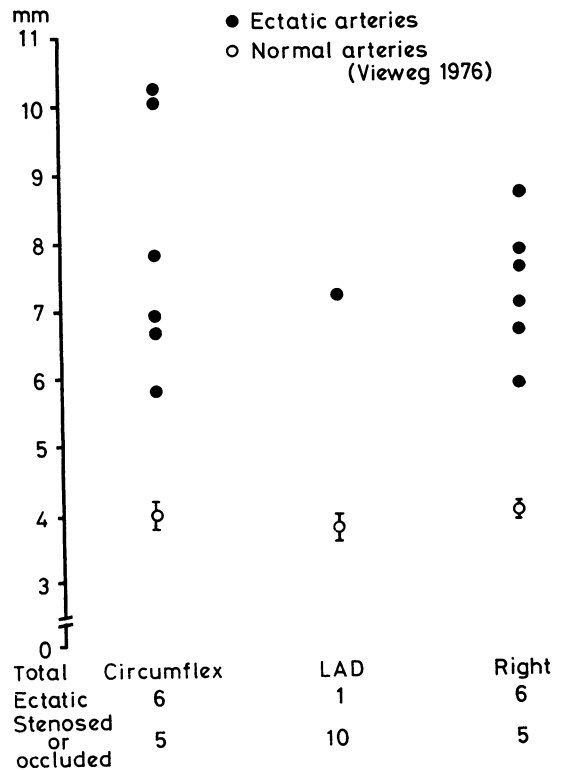


Fig. 1 Maximal coronary artery diameter compared with Vieweg's data for normal coronary arteries. Distribution of ectasia primarily in circumflex and right coronary artery with stenosed or occluded LAD (left anterior descending).

There was only 1 case of left anterior descending ectasia. Eleven patients had severe stenosis or occlusion of non-ectatic vessels. One patient (case 5) had an ectatic coronary artery with other

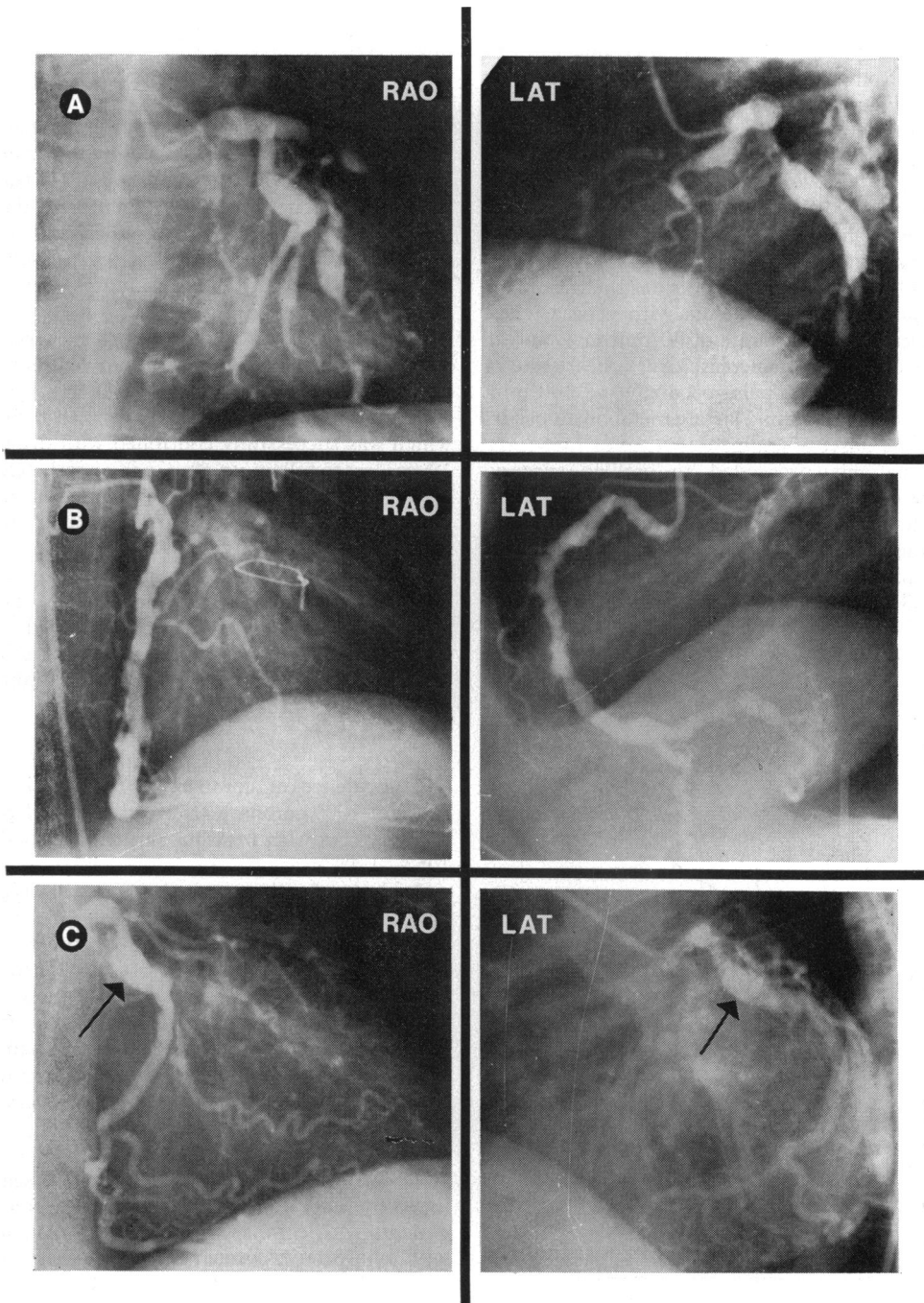


Fig. 2 Typical examples of coronary artery ectasia. RAO, right anterior oblique view; LAT, lateral view. (A) Case 2, grossly ectatic circumflex system; (B) case 4, generalised ectasia in right coronary artery; (C) case 6, fusiform dilatation of proximal circumflex in a man with collagen vascular disease.

vessels normal. Typical coronary arteriograms showing ectasia are shown in Fig. 2.

The measurement of maximum coronary artery diameter from biplane cut-film angiograms is shown in Fig. 1, and is compared with the results of Vieweg's data for normal coronary arteriograms.

A comparison of coronary sinus flow in 2 patients with ectatic and 14 patients with non-ectatic coronary artery disease is shown in Fig. 3. In the patients with non-ectatic coronary artery disease mean coronary sinus flow increased up to a heart rate of 140/min. At higher heart rates the mean flow did not increase. In the patients with ectasia, chest pain occurred at a heart rate of 80/min in 1, and in the other flow did not increase over a heart rate of 120/min, with chest pain occurring at 140/min. In both patients abnormal lactate metabolism occurred with the onset of angina.

Eight patients proceeded to cardiac surgery. Coronary artery bypass grafting was performed in 7, 1 of whom in addition had a mitral valve replacement. One patient had a mitral valve replacement without additional coronary artery bypass grafting (case 5). Graft flows were measured using a Carolina

Medical Electronics 601D Cliniflow electromagnetic flowmeter. Basal flows ranged from 30 to 150 ml/min down grafts to arteries with areas of ectasia, and 22 to 115 ml/min down grafts to non-ectatic vessels.

The histology from an ectatic segment obtained from case 2 at saphenous vein bypass grafting is shown in Fig. 4. The intima was grossly thickened and fibrotic. The media was extensively damaged, with destruction of both internal and external elastic laminae. Cholesterol clefts occurred in giant cells in the intima. The appearances were typical of severe arteriosclerotic disease.

Four patients did not proceed to surgery. Case 6 was diagnosed as having a collagen vascular disease and treated with steroids. Case 8 had symptoms resulting from paroxysmal ventricular tachycardia which was successfully controlled with drugs. Case 10 was lost to follow-up after cardiac catheterisation, and case 11 was considered unsuitable for operation because of very poor left ventricular function (ejection fraction 0.2).

The 1 patient who was not improved by coronary artery bypass grafting was restudied. The two grafts he had received were both occluded.

All patients with coronary artery ectasia were anticoagulated in the absence of contraindications.

Discussion

The incidence of coronary artery ectasia in this series of 1000 coronary arteriograms is 1.1 per cent which agrees with previous findings (Daoud *et al.*, 1963—1.4%).

Analysis of risk factors shows results typical of patients with occlusive arteriosclerotic coronary artery disease. All the patients were men, 11 were heavy smokers or ex-smokers. Only 2 had hypertension at the time of study and 1 of these (case 2) was the case from whom histology was available. Five of the patients (45% of the group) had hyperlipoproteinaemia which is no higher an incidence than in patients with non-ectatic coronary artery disease (Barboriak *et al.*, 1974—49%; Murray *et al.*, 1975—55%; Salel *et al.*, 1974—64%). A family history of coronary artery disease was found by Markis *et al.* (1976) to be more common in patients with coronary artery ectasia than in those with non-ectatic coronary artery disease. Only 2 of our 12 patients had positive family histories.

Aetiological factors in coronary artery ectasia are numerous. Scott (1948) considered congenital aneurysms the most common type. Seabra-Gomes *et al.* (1974) noted the association of congenital aneurysms with coronary arteriovenous fistulae and suggested that the fistula resulted from the

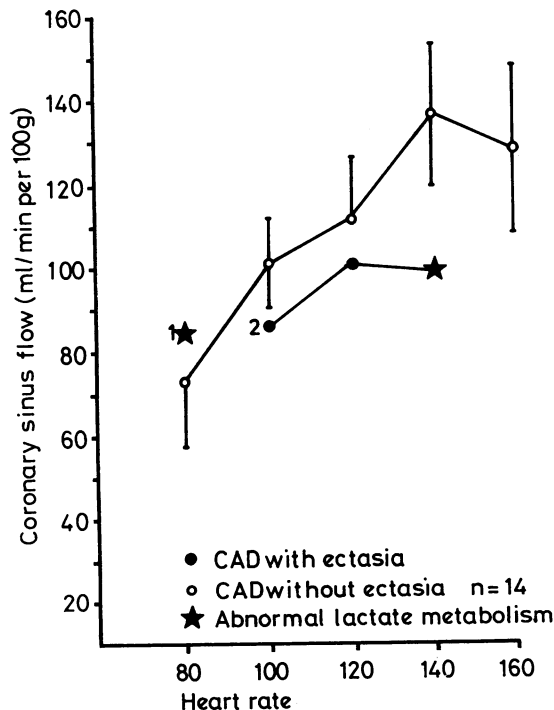


Fig. 3 Compares mean \pm ISD flow in patients with non-ectatic coronary artery disease with 2 patients (cases 1 and 2) with ectatic coronary arteries. CAD, coronary artery disease.

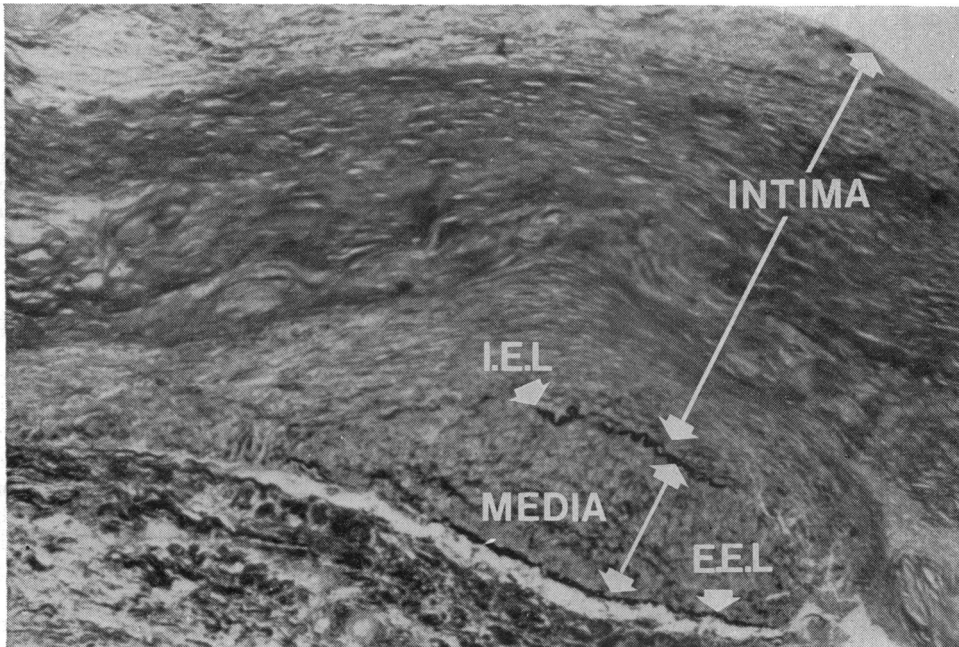


Fig. 4 Histology from ectatic coronary artery (case 2 stained with elastic Van-Gieson). Grossly thickened and fibrotic intima. Elastin of both internal and external elastic laminae degenerate. IEL, internal elastic lamina; EEL, external elastic lamina.

retention of primitive sinusoids in the myocardium. Ebert *et al.* (1971) considered that the coronary artery dilatation caused by a coronary arteriovenous fistula did not represent a true aneurysm. Increased flow caused by left-to-right shunting in anomalous origin of the left coronary artery from the pulmonary artery results in a very large right coronary artery (Hudson, 1965; Barrand *et al.*, 1975). However, this enlargement resulted from an increased flow rather than a diseased vessel wall and does not represent true aneurysmal dilatation or ectasia.

Coronary artery ectasia may occur in association with other congenital heart lesions (Björk, 1966). It may cause myocardial infarction in childhood (Sherkat *et al.*, 1967), or in the young adult (Ashton and Munro, 1948; Gore *et al.*, 1959) from thrombosis or rupture of an aneurysm. Congenital weakness of the elastin in the media may result in dissection, and may be associated with berry aneurysms intracranially (Iglauer *et al.*, 1959). Coronary artery ectasia or aneurysm in childhood may also result from an infantile polyarteritis (McMartin *et al.*, 1974; Chamberlain and Perry, 1971). More recently a mucocutaneous lymph node syndrome in Japanese children has been noted to cause coronary artery aneurysms resem-

bling infantile polyarteritis (Kato *et al.*, 1975).

Other conditions in which coronary artery ectasia or aneurysm has been noted include Ehlers-Danlos syndrome (Imahori *et al.*, 1969), scleroderma (Chaithiraphan *et al.*, 1973), cystic medionecrosis (Boschetti and Levine, 1958; McKeown, 1960), trauma (Konecke *et al.*, 1971), mycotic embolus (Crook *et al.*, 1973), and syphilitic aortitis (Morgagni, 1940). All these causes, however, are rare compared with arteriosclerosis in the series of Daoud *et al.* (1963) and Markis *et al.* (1976). Hypertension has been incriminated as a cause of medial destruction leading to ectasia (Markis *et al.*, 1976). The results of this series confirm that coronary artery ectasia is most frequently a variant of occlusive coronary arteriosclerosis. Ectasia and major stenoses may coexist in the same artery. The histology in the one case in this series illustrates the severe damage to the media that occurs in the ectatic segment. In only one case in this series was arteriosclerosis not the primary aetiological factor. This man (case 6) had pernicious anaemia, autoimmune thyroiditis, and polyarthritis. On investigation he had an eosinophilia and an antinuclear factor of 1:1250. His coronary arteriogram (Fig. 2C) showed a fusiform dilatation of the proximal circumflex, but a totally occluded anterior descending and right

coronary artery. Though biopsy evidence was not available, a diagnosis of an arteritis caused by a collagen vascular disease was made and he was treated with steroids.

A weakened media with degenerating elastin is a major factor in all causes of ectasia. Deposition of acid mucopolysaccharide in Ehlers-Danlos syndrome occurs primarily in the media (Imahori *et al.*, 1969). Cystic medial necrosis results in dissection of the media in Marfan's syndrome. Fibrinoid necrosis in polyarteritis involves inner media and intima of small arteries (Hudson, 1965), and the internal elastic lamina rapidly fragments. The weakened elastin then allows dilatation or dissection of the arterial wall especially in the presence of hypertension. The mechanism is not necessarily post-stenotic turbulence producing dilatation, as ectasia may occur in the absence of stenoses.

The distribution of ectasia in this series was principally in the circumflex or right coronary arteries. In 10 patients the left anterior descending was either severely stenosed or occluded and 6 had had previous anterior infarcts (Table). The necropsy series of Daoud *et al.* (1963) has a higher incidence of aneurysm in the anterior descending but states

that they were equally distributed between left and right systems. This series agrees with the equal distribution, but ectasia in the anterior descending occurred in only one patient (case 2). Only 1 patient (case 5) had an ectatic coronary artery with other vessels normal. He presented with effort dyspnoea and angina and at cardiac catheterisation was found to have mitral regurgitation caused by anterior chordal rupture. His chest pain may have been caused either by abnormal tensing of the remaining chordae (Gooch *et al.*, 1972) or by mural thrombus in the ectatic artery embolising distally.

Coronary artery ectasia may be associated with arteriomegaly elsewhere. The combination of coronary artery aneurysms and aortic aneurysms was reported by Daoud *et al.* (1963). In this series case 12 had coronary artery ectasia and arteriomegaly of the iliofemoral segments as described by Lea Thomas (1971) (Fig. 5). Histological sections of arteriomegalic leg vessels have shown changes indistinguishable from arteriosclerosis. A second patient (case 2) had a history of cerebral emboli, but cerebral angiography was not performed. Radiological changes very similar to coronary artery ectasia have been shown in cerebral arteries



Fig. 5 Iliofemoral arteriomegaly in a case of coronary artery ectasia (case 12).

(Ross Russell, 1972) and the dilated segments may contain mural thrombus.

The limited data on coronary sinus flow in patients with ectatic arteries suggest that the presence of ectasia does not necessarily result in an increased flow (compared with non-ectatic coronary artery disease). In case 2 (the patient with ectatic circumflex and anterior descending arteries) the coronary sinus flow was lower than in the group of patients with non-ectatic coronary artery disease. This method of measurement primarily reflects left coronary artery flow. At cardiac surgery, measurement of flows down the grafts into arteries with areas of ectasia were in the same range as in grafts to non-ectatic arteries. With a vessel of greater diameter but an equal or reduced flow the velocity of blood must be reduced. It is of interest to compare these results with studies of blood flow in arteriomegalic pelvic and leg vessels (Callum *et al.*, 1974) which showed no difference between normal and arteriomegalic patients. The velocity of blood in the arteriomegalic group was reduced.

The treatment of the arteriosclerotic group was mitral valve replacement in the 2 cases of chordal rupture and saphenous vein bypass grafting in 7 patients. In view of the tendency for mural thrombosis to occur in arteriomegalic leg vessels (Lea Thomas, 1971), cerebral vessels (Ross Russell, 1972), and probably coronary arteries (Ebert *et al.*, 1971) patients with coronary artery ectasia should be anticoagulated.

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