

patients with polyarthritis in Kenya whereas Gelfand⁶ failed to detect SLE in a prospective 5-year study of polyarthritis in Zimbabwe. In a retrospective study during the period 1957-1966, Greenwood⁷ found 2 cases in a group of 104 patients with auto-immune disease, the corresponding number of admissions to hospital over this period being 98 454. Shaper⁸ reported 5 patients with SLE in Uganda, 2 of whom were male. Further evidence of the rarity of SLE in the Black population stems from the study of Seedat and Pudifin² in Durban during 1969-1975. SLE was detected in only 17 out of 62 623 Black patients admitted to general medical wards of King Edward VIII Hospital, the admission rate for SLE being 0,28/1 000. Moreover, at the same hospital during the period 1960-1980, Rovers and Coovadia⁴ found no Black children with SLE among 119 000 admissions. Genetic and environmental factors probably contribute to the difference in prevalence of SLE between the Blacks in Africa and America.

This article illustrates three important aspects. Firstly, SLE does occur in the Black South African male but appears to be rare. Secondly, a wide spectrum of the manifestations of SLE were noted in these 3 cases. Thirdly, it is likely that more cases

may emerge with an increased awareness of SLE and the availability of more sensitive and specific laboratory tests.

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Hypertrophic non-obstructive apical cardiomyopathy

A case presentation and review of the literature

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Summary

A 20-year-old Coloured man gave a history of atypical chest pain, palpitations after strenuous exercise and a single episode of post-exertional presyncope. The diagnosis of hypertrophic non-obstructive apical cardiomyopathy (HNOAC) was established by means of electrocardiography, echocardiography (both M-mode and two-dimensional) and left ventricular cine angiography. This variant of hypertrophic cardiomyopathy is most unusual and has been encountered most frequently in Japan, although a few cases have been diagnosed in the USA. The present case is the second reported from the Republic of South Africa. Important aspects of HNOAC are reviewed.

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Clinical presentation

The patient was a 20-year-old Coloured 1st-year student of physical education at a local university. While at school he was actively involved in rugby, football and athletics. A systolic murmur detected during a routine medical examination led to referral to the Cardiac Clinic of Tygerberg Hospital for further evaluation. Initially he denied having any symptoms, but on interrogation admitted to having experienced a presyncopal episode immediately after severe exertion approximately 2 years previously and palpitations for a few minutes after exercise. A 'heavy' precordial pain lasting some 10-20 minutes and associated with tension was initially noted about 1 year previously and occurred almost weekly. There was no history of effort-induced precordial discomfort or dyspnoea. The only suspicious factor in the family history was the possible presence of a heart condition in a paternal grandmother aged 58 years.

On clinical examination the patient was of athletic build. The only abnormal features were cardiovascular. The blood pressure was 150/70 mmHg. A prominent left ventricular type apex was palpated in the fifth left intercostal space in the midclavicular line and was not 'double' in character. There was no evidence of right ventricular enlargement. Auscultation at the mitral area revealed normal heart sounds without additional sounds. There was an apical grade 2/6 early ejection systolic murmur of fairly high frequency, radiating up the left sternal edge and slightly to

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the axilla where it softened markedly. This murmur diminished marginally in intensity after squatting, but did not alter in quality after performance of the Valsalva manoeuvre. There were no diastolic murmurs and the aortic and pulmonic components of the second heart sound at the base were normal.

A chest radiograph in the postero-anterior and left lateral projection outlined a peculiarly shaped heart with the apex more 'pointed' than usual and tilted upwards somewhat. The resting 12-lead electrocardiogram (ECG) demonstrated sinus rhythm of 75/min, a P-R interval of 0,14 second and a mean frontal QRS axis of +79°. The right atrium also appeared to be enlarged. However, the most striking abnormality of the ECG was the widespread increase in QRS voltage and very abnormal T waves (Fig. 1). The T waves were asymmetrically inverted in all the leads except leads aVL, aVR, V1 and V2. The deepest T wave inversion was in leads V3 - V6 (maximum depth 10,5 mm in lead V5). There were no pathological septal Q waves and the normal small Q waves in leads I and aVL were absent, suggesting a degree of incomplete left bundle-branch block. Submaximal treadmill exercise testing (Bruce protocol) caused a reduction in

the negativity of the T waves and no ST-segment depression was seen. In addition, the T waves 'normalized' in leads V1 - V3.

M-mode echocardiography with the patient supine and the transducer over the third intercostal space to the left of the sternum and directed so that the ultrasound beam passed across the left ventricle from apex to base revealed that the inter-ventricular septum (IVS) and left ventricular posterior wall (LVPW) were of almost equal thickness at the base (measured below the tips of the mitral valve leaflets before atrial systole and after the phase of rapid ventricular filling). However, the LVPW thickness increased dramatically towards the apex while the IVS thickness remained virtually unchanged (Fig. 2, Table I). As a result of this the left ventricular cavity was virtually obliterated at the apex, especially during ventricular systole. The LVPW and IVS became more hyperkinetic in the direction of the apex. These echocardiographic features were characteristic of hypertrophic apical cardiomyopathy.

Cross-sectional (two-dimensional) echocardiography with an ATL wide-angle electronic sector scanner with a 3 MHz transducer, beam directed so as to obtain a long-axis (sagittal plane) view (Fig. 3) as well as a short-axis two-chamber view (Fig. 4) confirmed the presence of marked apical hypertrophy which virtually occluded the apical chamber at the conclusion of ventricular systole.

Cardiac catheterization demonstrated normal intracardiac pressures and normal parameters of left ventricular function. No left-sided resting intraventricular gradient could be detected. Provocative manoeuvres were not carried out. A left ventricular cine angiogram in the right anterior oblique projection delineated the characteristic features of hypertrophic apical cardiomyopathy (Fig. 5). Thus, at end-diastole the left ventricular cavity had the configuration of a 'spade' as depicted on playing cards, while the apex was hypercontractile during systole. There was no evidence of mitral valve incompetence.

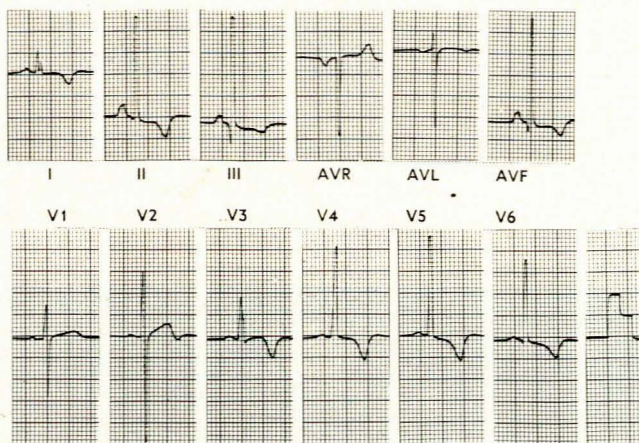


Fig. 1. ECG with half-standardization of the precordial leads. Widespread increased QRS voltage is present and the T waves are deeply and asymmetrically inverted, especially in the left precordial leads. There is possible slight right atrial enlargement as well as incomplete left bundle-branch block.

Discussion

The most commonly appreciated forms of hypertrophic cardiomyopathy are 'obstructive' ('hypertrophic obstructive cardiomyopathy' — HOCM),¹ or 'non-obstructive' ('hypertrophic non-obstructive cardiomyopathy' — HCM).² In both these

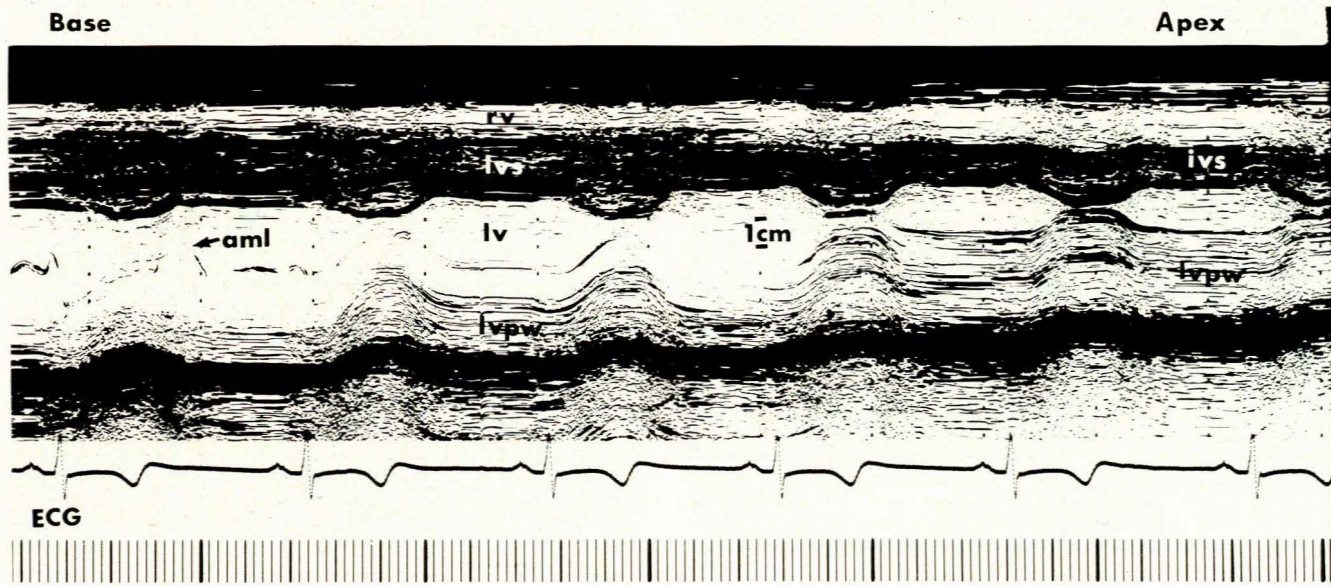


Fig. 2. M-mode echocardiogram demonstrating reduction in the left ventricular cavity towards the apex (especially during systole) mainly due to gross hypertrophy of the posterior wall (rv = right ventricle; lv = left ventricle; ivs = interventricular septum; lvpw = left ventricular posterior wall; aml = anterior mitral leaflet).

TABLE I. ECHOCARDIOGRAPHIC MEASUREMENTS

Parameter	Result	Normal value (mean)
LV end-diastole (base)	53 mm	35 - 56 (46 mm)
LV end-systole (base)	35 mm	
IVS thickness (base)	13 mm	7 - 11 (9 mm)
LVPW thickness (base)	15 mm	7 - 11 (9 mm)
IVS/LVPW thickness ratio (base)	0,9	<1,3
LV end-diastole (apex)	27 mm	
LV end-systole (apex)	0 - 5 mm	
IVS thickness (apex)	15 mm	
LVPW thickness (apex)	33 mm	
IVS/LVPW thickness ratio (apex)	0,45	
Left atrium/aortic ratio	1,2	0,9 - 1,2
Left atrium dimension	41 mm	19 - 40 (29 mm)
LV end-diastolic volume (base)	135 ml	130 ml/m ²
LV end-systolic volume (base)	51 ml	25 ml/m ²
LV ejection fraction (base)	62%	>60%
LV end-diastolic volume (apex)	27 ml	
LV end-systolic volume (apex)	0,3 ml	
LV ejection fraction (apex)	98%	
LV shortening fraction	33%	28 - 38%
LV velocity of circumferential shortening	1,26	1,15 - 1,35
Systemic pre-ejection period (PEP)	90 msec	
LV ejection time (LVET)	270 msec	
PEP/LVET ratio	0,33	<0,28 - 0,38

LV = left ventricular.

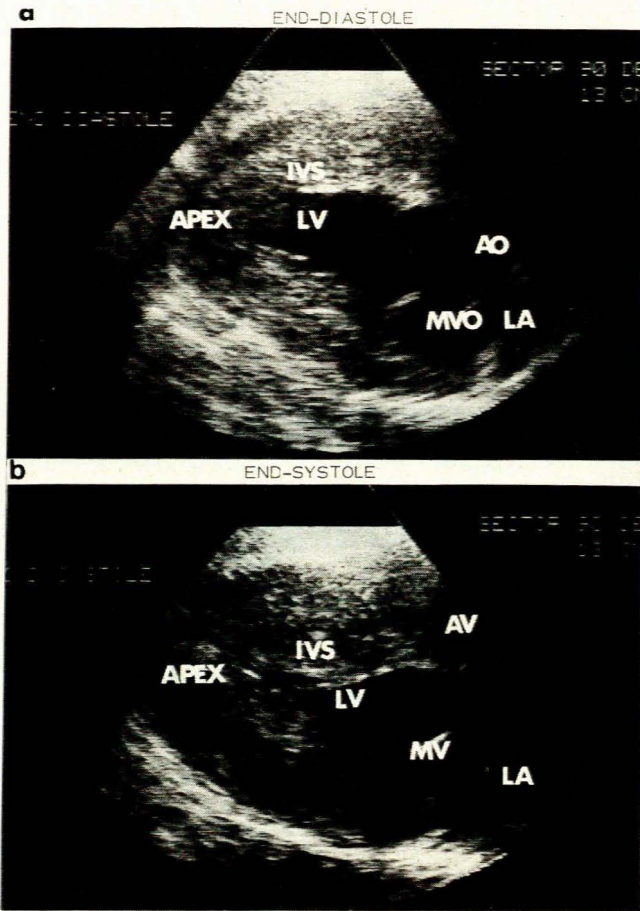


Fig. 3. Long-axis two-dimensional (cross-sectional) echocardiogram in (a) end-diastole and (b) end-systole. There is gross thickening of the LVPW giving rise to virtual obliteration of the apex at the end of systole (LV = left ventricle; AO = aorta; LA = left atrium; MVO = mitral valve orifice; AV = aortic valve).

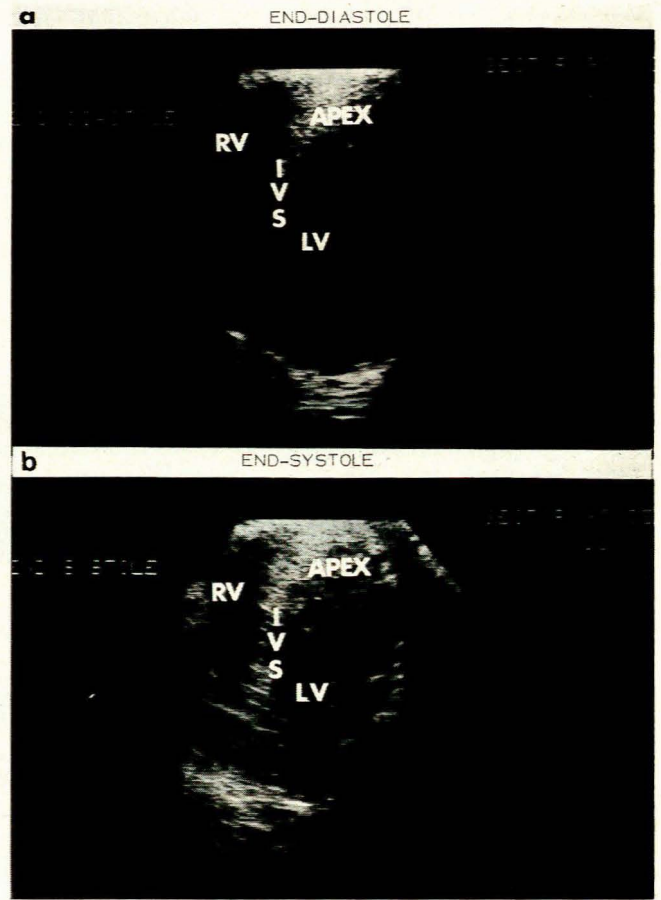


Fig. 4. Short-axis two-chamber view two-dimensional (cross-sectional) echocardiogram in (a) end-diastole and (b) end-systole. Marked reduction of the left ventricular apical cavity is visualized at the end of systole.



Fig. 5. Left ventricular cine angiograms in the right anterior oblique (RAO) projection. The classic 'spade' appearance of the left ventricle due to localized hypertrophy of the apex (arrowed) is visualized. There is no mitral valve incompetence. a — ED = end-diastole; b — ES = end-systole.

forms the IVS is hypertrophied most significantly in the left ventricular outflow tract, hence the commonly used synonym 'idiopathic hypertrophic subaortic stenosis'. The exact mechanism of obstruction is the subject of intense debate. A far less common form of HOCM is hypertrophy of the IVS and, less significantly, of the LVPW in the mid-portion of the left ventricle.³

In 1976 Yamaguchi *et al.*⁴⁻⁶ described an apical form of hypertrophic cardiomyopathy which they termed 'hypertrophic non-obstructive apical cardiomyopathy' (HNOAC), characterized by giant T waves in the precordial leads, and later reported further patients with this variety of cardiomyopathy.⁷⁻⁹ The first patients with apical hypertrophic cardiomyopathy reported outside Japan were reported by Maron *et al.*¹⁰ in the USA in 1982. Soon afterwards the first case in the Republic of South Africa was documented by Steingo *et al.*¹¹ A further case report from the USA was that of Kereiakes *et al.*¹² in 1983. Our article describes the second case of HNOAC in South Africa. A review of the most important features of this uncommon and interesting type of hypertrophic cardiomyopathy follows.

Genetic aspects

Not much is known about the mode of inheritance of HNOAC. What is rather striking is the preponderance of male patients, since 42 of 49 cases recorded to date were in males (Table II). This fact prompted Yamaguchi *et al.*⁹ to suggest a sex-linked recessive mode of inheritance. However, Maron *et al.*¹⁰ thought that the disease was transmitted by an autosomal dominant gene, an inheritance pattern classic in the better-known HOCM.¹³ Maron *et al.*¹⁰ performed echocardiography on 20 relatives of 4 of the 5 families, some of whom also underwent cardiac catheterization, and documented features of the usual form of hypertrophic cardiomyopathy in 11 of the 20. Seven of the 8 actual family members had both basal and apical ventricular septum hypertrophy, 4 also having evidence of asymmetrical septal hypertrophy. Maron *et al.*¹⁰ concluded that HNOAC was merely part of the spectrum of the more classic HOCM, especially since the 2 patients who came to autopsy had histological changes and areas of abnormal myocardium distribution similar to those encountered in HOCM. Although our patient has young parents and several sibs, it was not possible to

examine them. Therefore, at this stage we cannot comment on the possible mode of inheritance.

Symptoms

The majority of patients with HNOAC are asymptomatic, and the diagnosis is often made on the basis of routine physical examination supported by investigations such as ECGs and echocardiography. Chest pain, usually atypical, and dyspnoea on exertion are the most frequent complaints (15 cases out of 49 reported in the literature) (Table II). Palpitations, usually related to effort, were the next most common symptom (9 patients). In these cases the history may suggest the possibility of underlying ischaemic or even valvular heart disease.

Clinical features

The bedside diagnosis of HNOAC is subtle and is probably impossible without an ECG (and even possibly additional M-mode and two-dimensional echocardiography). The most frequent physical finding is that of an ejection systolic murmur heard best at the third or fourth left intercostal space and sometimes over the apex. This murmur was heard in 47 of the 49 cases in the literature, and does not alter with performance of the Valsalva manoeuvre or squatting, as does the murmur of the more common HOCM. Therefore, it may well be mistaken for the murmur of mitral valve prolapse or aortic sclerosis, or even for an innocent systolic murmur. A fourth heart sound was heard in at least 30 of the 49 cases in the literature but there is no evidence of the presence of a third heart sound. Clinical left ventricular cardiomegaly is not a feature, and neither is the classic 'double apical impulse' or 'brisk to upstroke' peripheral pulse. This condition is therefore usually not mistaken for HOCM on clinical examination.

ECG characteristics

The ECG is often the first clue to the diagnosis. The almost pathognomonic findings are of giant T-wave inversion in the left precordial leads accompanied by increased QRS voltage, particularly in leads V4 and V5. These features were initially noted by

TABLE II. CLINICAL AND HAEMODYNAMIC FEATURES OF PATIENTS WITH HYPERTROPHIC APICAL CARDIOMYOPATHY

Study	Year	No. of cases	Sex		Mean age (yrs)	Symptoms			Auscultation			ECG			M-mode echo.		Cardiac catheterization	
			M	F		Paalp.	DOE	Chest pain	Gallop rhythm	ESM	Q waves	SV1 + RV5 (mm)*	T wave (mV)	Max. neg. (V5)	IVS (apex/base)	LVPW (apex/base)	Done (No.)	IV gradient
			7	2		4	No	No	?	9	No	21 + 43 (64)	2,1 (V5)	2,2 (V5)	2,2	2,2	No	Unknown
Sakamoto <i>et al.</i> ⁷ (Japan)	1976	9	7	2	43	4	No	No	?	9	No	21 + 43 (64)	2,1 (V5)	2,2 (V5)	2,2	No <td>Unknown</td> <td>No</td>	Unknown	No
Yamaguchi <i>et al.</i> ⁹ (Japan)	1979	30	29	1	48	4	10	15	30	30	4	>35 (V5)	>1,5 (V5)	1,9	Yes (30)	No	Yes (30)	No
Maron <i>et al.</i> ¹⁰ (USA)	1982	7	4	3	34	No	6	No	?	7	1	Unknown (V5)	0,6 (V5)	Unknown	Yes (5)	Yes (1 case)	Yes (5)	Yes (1 case)
Steingo <i>et al.</i> ¹¹ (South Africa)	1982	1	1	—	32	No	No	No	No	No	No	22 + 40 (64)	1,0 (V4)	Unknown	Yes	No	Yes	No
Kereiakes <i>et al.</i> ¹² (USA)	1983	1	—	1	55	No	No	No	No	No	No	9 + 15 (24)	1,5 (V4)	Unknown	Yes	Yes	Yes	No
Przybojewski and Blake (South Africa) — present study	1983	1	1	—	20	Yes	No	Yes	No	Yes	No	26 + 46 (72)	1,05 (V5)	1,1	2,2	Yes	Yes	No

*QRS voltages on the ECG are given indicating the depth of the S wave in lead VI and the height of the R wave in lead V5 with the sum in millimetres in parentheses. Paalp. = palpitations; DOE = dyspnoea on effort; ESM = ejection systolic murmur; Max. neg. = maximum negativity; IV = intraventricular.

Yamaguchi *et al.*⁴⁻⁶ who later correlated these ECG findings with those found on echocardiography and cine angiography.^{8,9} In their series of 30 successive patients Yamaguchi *et al.*⁹ reported abnormal QRS-voltage criteria in all and a maximum negative T-wave value in lead V5 in excess of 1,5 mV (Table II). Associated with these changes was the absence of septal Q waves, contrary to the finding in HOCM. Sakamoto *et al.*⁷ reported on an even greater mean QRS voltage of 64 mm and a maximum negative T-wave value of 2,1 mV in lead V5 in their 9 cases. Maron *et al.*¹⁰ on the other hand, documented only slight degrees of T-wave inversion reaching a maximum negativity of 0,6 mV in lead V5; they did not report on actual QRS voltage in their 7 patients. Steingo *et al.*¹¹ obtained a QRS-voltage sum of 64 mm in leads V1 and V5, a figure identical to that reported by Sakamoto *et al.*⁷ (Table II). The maximum T-wave negativity in lead V4 was 1,0 mV in Steingo *et al.*'s¹¹ patient, a figure below that usually encountered. Kereiakes *et al.*'s¹² patient had a low QRS-voltage sum of 24 mm with a maximum negative T-wave value of 1,5 mV in lead V4. Our patient had a remarkably high QRS-voltage sum of 72 mm, one of the largest reported (Fig. 1, Table II). The maximum T-wave negativity encountered was 1,05 mV in lead V5. Interestingly, some workers have documented increasing QRS voltage and T-wave negativity with a progressive increase in the degree of apical hypertrophy.^{7,8} This observation has led Sakamoto *et al.*⁷ to postulate that the T-wave abnormalities are due to abnormal repolarization within the hypertrophied apical myocardium. Sakamoto *et al.*⁷ also noted that 3 of their 'control' patients with HOCM had T-wave changes identical to those found in HNOAC, and suggested that these patients fell into the spectrum of hypertrophic cardiomyopathy. Giant negative T waves have also been noted by Savage *et al.*¹⁴ in both the obstructive and non-obstructive forms of hypertrophic cardiomyopathy, a finding thought to be due to the disproportional hypertrophy of the cardiac apex. However, Maron *et al.*¹⁰ claimed that these giant negative T waves were rarely encountered in HOCM and that their presence in that disease did not necessarily imply hypertrophy of the left ventricular apical myocardium.

Apart from HNOAC, giant T-wave inversion has been documented¹⁵ in ischaemic heart disease, right bundle-branch block and right ventricular hypertrophy, various metabolic disorders, coronary arteriography, complete heart block presenting as Stokes-Adams attacks, and cerebrovascular accidents.¹⁶ Marked increase in QRS voltage as well as deeply inverted T waves have been encountered as a 'normal variant pattern' in South African Blacks.¹⁷⁻¹⁹ Similar QRS-voltage changes but with less severe T-wave inversion and localization to the right ventricular precordial leads have been seen in the 'athletic heart syndrome'.^{20,21}

Another interesting ECG finding in patients with HNOAC is a prolonged QTc interval (37 out of 49 cases in the literature). Some patients with classic HOCM have this abnormality as well, but the incidence is far lower. This prolongation of the Q-T interval may play an important role in prognosis through the genesis of potentially fatal ventricular arrhythmias.

Exercise ECG

Sakamoto *et al.*⁷ noted that in all their 9 cases of HNOAC stress testing reduced the degree of deep T-wave inversion during the exercise phase and that ST-segment depression did not occur during or afterwards. Their patients also did not complain of angina on effort and arrhythmias were not precipitated. The 30 patients of Yamaguchi *et al.*⁹ all had negative stress tests, but these authors did not comment on the T-wave response. It is not clear whether Maron *et al.*'s¹⁰ 7 patients with HNOAC underwent exercise ECGs. The patient documented by Steingo *et al.*¹¹ had worsening of T-wave inversion immediately after a submaximal stress test and significant ST-segment

depression 2 minutes after exercise; ischaemic heart disease was excluded on the basis of normal coronary cine angiograms. The patient of Kereiakes *et al.*¹² was not subjected to an exercise ECG. Our patient had less T-wave inversion with submaximal stress, and normalization (upright T waves) in the right precordial leads was seen with no ST-segment shift or precipitation of arrhythmia or angina. Thus, most patients with HNOAC show improvement of ECG appearances after stress testing.

It may be difficult to differentiate the 'athletic heart' from HNOAC purely by stress ECG. Hanne-Paparo *et al.*²² and Zeppilli *et al.*²³ both noted a decrease in T-wave inversion or even normalization after effort; similarly, Oakley and Oakley²⁰ documented 'normalization' of T waves in 5 of 10 athletes. Differentiation of HNOAC from HOCM by effort ECG is controversial. McKenna *et al.*²⁴ noted that submaximal stress did not normalize the T waves in 30 patients with HOCM, while Rubin *et al.*²⁵ found little change in ST segments or T-wave inversion in 10 patients with HOCM, and in fact noted a deterioration in the ECG appearances in some. In our experience submaximal stress testing increases T-wave inversion without causing ST-segment depression in patients with HOCM and normal coronary arteries on angiography.

M-mode echocardiography in diagnosis

HNOAC can be missed in echocardiography if the short-axis view is employed and the echo beam is not directed from the mitral valve in a continuous sweep (long-axis view) towards the left ventricular apex. Asymmetrical septal hypertrophy and systolic anterior motion of the anterior mitral valve leaflet are not usually present to support a possible erroneous clinical diagnosis of HOCM, thus excluding further searching for a possible cause. This is further complicated by the fact that the movement of the IVS and LVPW in the region of the tips of the mitral valve leaflets is usually normal. With scanning from the left ventricular base to the apical region a progressive increase in the thickness of the LVPW (less so as regards the IVS) is demonstrated (Fig. 2). The apex/base ratio of the IVS and apex/base ratio of the LVPW is therefore always greater than unity and is often double this (Table II). This progressive thickening of the LVPW and IVS towards the apex causes a marked reduction in the left ventricular cavity, particularly at end-systole.^{9,26} Maron *et al.*¹⁰ found that in 6 of their 7 patients with HNOAC the mitral valve was situated abnormally anteriorly, within the cavity of the left ventricle, a finding previously encountered by others.²⁷ Additional features reported are an increased ejection fraction as well as increased velocity of circumferential shortening of the left ventricle.⁷

Two-dimensional (cross-sectional) echocardiography

This form of echocardiography can be diagnostic of HNOAC when the short-axis view is normal. A strong clinical suspicion of this type of hypertrophic cardiomyopathy must therefore exist before the classic features are detected on cross-sectional echocardiography. Sakamoto *et al.*⁷ clearly demonstrated the advantages of the two-dimensional approach. A wide angle of view or sector of at least 70° is usually advocated. Both long-axis (sagittal plane) images, obtained by directing the echo beam in a sweep from the apex to the base of the heart, as well as short-axis (transverse plane) views, derived by sweeping from the right hip to the left shoulder perpendicular to the long axis of the left ventricle, at end-diastole are usually used in diagnosis. This technique allows for measurement leading to delineation of severe narrowing of the left ventricular apical cavity as demonstrated in our patient (Figs 3 and 4). In their series of 7 patients with HNOAC Maron *et al.*¹⁰ used a real-time phased-array sector scanner with an 80° field of view, and four additional

apical chamber views to demonstrate the localized apical hypertrophy. Subsequently others^{11,12} successfully used two-dimensional echocardiography, although most of their cases were also defined by the M-mode technique. It appears that two-dimensional echocardiography is the most accurate non-invasive means of diagnosing this form of cardiomyopathy, and this investigation should be requested if there is evidence of giant T waves on the ECG.

Haemodynamic findings and left ventricular cine angiography

This invasive investigation was carried out in 38 of the 49 cases documented in the literature (Table II); Sakamoto *et al.*⁷ did not perform this procedure on their 9 patients, while 2 of the 7 patients reported on by Maron *et al.*¹⁰ did not undergo this investigation. Only 1¹⁰ out of 38 patients who underwent cardiac catheterization had a resting left ventricular outflow-tract gradient of 63 mmHg. In many other patients techniques such as sublingual nitroglycerin administration, intravenous infusion of isoproterenol and post-extrasystolic potentiation failed to provoke a left ventricular outflow-tract gradient, in marked contrast to patients with HOCM. Yamaguchi *et al.*⁹ using an upper limit of normal of 10 mmHg for left ventricular end-diastolic pressure (LVEDP) found this to be elevated in 29 out of 30 patients, the highest value being 31 mmHg. Ejection fractions of the left ventricle in Yamaguchi *et al.*'s⁹ patients were well above 70% in 29 of the 30. The LVEDP was raised in 4 of 5 patients of Maron *et al.*¹⁰ the highest pressure recorded being 40 mmHg. Steingo *et al.*'s¹¹ patient had a LVEDP of 10 mmHg with a very high calculated ejection fraction of 84%. Kereiakes *et al.*'s¹² patient had a 6 mmHg LVEDP which rose to 23 mmHg after left ventricular cine angiography, and the calculated ejection fraction was 76%. Our patient had a borderline LVEDP of 12 mmHg with an ejection fraction of 62% at the base and 98% at the left ventricular apex (Table I).

Left ventricular cine angiography in the right anterior oblique projection delineates the pathognomonic appearance at end-diastole, described⁹ as a 'spade on a playing card', due to concentric apical hypertrophy. This was well illustrated by our patient (Fig. 5). Furthermore, very energetic systolic symmetrical contraction is noted. These features are in contrast to those in HOCM, in which the cardiac outline resembles a banana. Yamaguchi *et al.*⁹ found the thickness of the left ventricular anterior wall to be markedly increased towards the apex with a resultant decrease in intracavity dimension, and also noted an abnormally thickened left ventricular posterior free wall. Maron *et al.*¹⁰ noted that the left ventricular cine angiographic configuration differed from those reported by Yamaguchi *et al.*⁹, since the small apical segment displayed poor contractility and communicated with a normal-sized, more proximal chamber via a narrow intermediate segment, whereas Yamaguchi *et al.*'s⁹ patients had a small, hypercontractile apical segment. Cases described by Steingo *et al.*¹¹ and by Kereiakes *et al.*¹² had the classic 'spade on a playing card' configuration. None of the patients had reported mitral insufficiency, a fairly frequent finding in HOCM, but only a minority had undergone selective coronary angiography to exclude ischaemic heart disease since they did not complain of classic angina pectoris.

Management and prognosis

There has been very little discussion of prognosis in patients with HNOAC. Most are entirely asymptomatic or minimally symptomatic, which is probably why little attention has been given to prophylactic or symptomatic drug therapy. It would seem logical to postulate that cessation of strenuous physical activity might diminish the ECG or even echocardiographic

signs of left ventricular hypertrophy as in the 'athletic heart' syndrome. However, since there are no signs of obstruction to left ventricular outflow it is probably unlikely that physical exertion would be deleterious.

Since there is no resting or provokable gradient present it would seem somewhat unscientific to administer long-term β -blockers,²⁴ calcium blockers,²⁸ a combination of these substances or disopyramide²⁹ as previously advocated for HOCM patients. By the same token, if no ventricular arrhythmias can be detected amiodarone³⁰ should not be prescribed. Nevertheless, the frequent occurrence of a prolonged QTc interval may indicate a risk of fatal ventricular arrhythmias — careful follow-up by repeated Holter monitoring would therefore be necessary.

Only 2 patients¹⁰ out of 49 in the literature have died; both were young (26 and 38 years of age) and both died suddenly. The younger had had minimal symptoms before death, and the older was in severe congestive cardiac failure. Autopsy identified marked cellular disorganization, especially in the apical part of the IVS, as well as the LVPW. Maron *et al.*¹⁰ believe that HNOAC is part of the spectrum of hypertrophic cardiomyopathy in view of the inheritance pattern in their patients, as well as their previous documentation of a similar histological distribution of myocardial disorganization in the more classic variety of hypertrophic cardiomyopathy. This raises the question whether HNOAC can eventually evolve into classic HOCM. Only longitudinal ECG and echocardiographic studies will answer this interesting question.

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