Radiological Aspects of Heart Muscle Disease in Nigerian Adults

G. J. Thorpe, M.B., M.R.C.P., D.T.M. & H., and

SUMMARY
A congestive cardiomyopathy, known locally as heart muscle disease, accounts for 32% of the cases of cardiovascular disease seen at University College Hospital, Ibadan, Nigeria.

Following a brief outline of the clinical picture and necropsy findings, the radiological features of the disease are described. Thirty-four cases were studied, and angiography was performed in 13. In the acute stage of the disease there was a generalized, globular, cardiac enlargement, and cardiac movements, especially of the left border of the heart, were reduced. Pulmonary venous congestion was invariably present and pleural effusions occurred frequently. Ventricular angiography demonstrated an enlarged chamber with little change in size and shape throughout the cardiac cycle, and persistence of contrast. Mitral incompetence was present in some cases. The value of angiography in differential diagnosis is emphasized.

Additional Indexing Words:
Congestive cardiomyopathy Ventricular angiography Mitral incompetence
Ventricular contractility Cardiac hypertrophy

During the past 25 years heart failure of obscure etiology has been described in many parts of the world. In some areas of the tropics and subtropics, where coronary artery disease is rare, this syndrome occupies a prominent position among cardiovascular diseases. From the African continent most accounts of this entity have come from South Africa, where it has been described in the Bantu and in the white populations. It is also known to occur in West Africa and Southern Rhodesia. In Nigeria the condition has been recognized for a number of years as the commonest cause of heart failure, and at Ibadan it is known as "heart muscle disease." Other designations to be found in the literature for this and similar entities include: (1) myocardial degeneration with hypertrophy and failure of unknown cause; (2) nutritional heart disease; (3) cardiovascular collagenosis; (4) idiopathic hypertrophy of the heart; (5) acute reversible heart failure; (6) cryptogenic heart disease; (7) idiopathic cardiomyopathy; (8) primary myocardial disease; (9) cardiac disorder of unknown etiology; and (10) cardiomegaly of unknown origin.

At University College Hospital, Ibadan, heart muscle disease was the most common cardiac disorder found in a necropsy series, and in an analysis of medical admissions for 1958, the disease accounted for 30% of the cases of cardiovascular disease. More recently at the same hospital, out of 291 consecutive patients registering at the cardiac clinic, 93 (32%) were given a diagnosis of heart muscle disease.

From the Departments of Radiology and Medicine, University College Hospital, Ibadan, Nigeria.

This study was undertaken during Dr. Thorpe’s tenure of a Nuffield Foundation Medical Fellowship.

Address for reprints: Dr. W. P. Cockshott, Professor of Radiology, Faculty of Medicine, McMaster University, Hamilton, Ontario, Canada.

Circulation, Volume XXXVI, September 1967

460
The purpose of this paper is to describe the radiological features of heart muscle disease in the Nigerian adult.

The Clinical Picture

Most patients are over the age of 35 years and present with varying degrees of breathlessness and edema. Cough, ill-defined chest pain, and hepatic pain are often present. The duration of symptoms varies from a few days to many months. Examination reveals biventricular failure. Edema may be slight and restricted to the legs, or gross and generalized with ascites. Crepitations are usually present throughout the lungs. The liver is enlarged and tender.

The pulse is small and extrasystoles are common. The diastolic blood pressure is often raised. The heart is invariably enlarged: the apex beat is often heaving and a left parasternal heave may be present. A third sound is common and a pansystolic murmur may be present at the apex indicating mitral incompetence. The pulmonary closure sound is often accentuated.

Dramatic improvement has been shown to follow rest in bed alone, but as a rule digoxin and a diuretic are given to expedite recovery. In most patients, over a period of a few days, diuresis occurs, the heart becomes smaller, the blood pressure returns to normal, and mitral incompetence disappears.

Clinically, cases may be divided into four groups according to the evolution of the disease: (1) fulminating type, in which despite treatment the heart failure remains uncontrolled and death ensues rapidly; (2) acute type with recovery; (3) relapsing type which is probably the commonest type; after recovery from the first attack, relapses occur at intervals, and ultimately, death may take place during a relapse, recovery may occur, or the patient may pass into group 4; (4) chronic type, in which even with treatment the patient remains either in, or on the verge of, heart failure throughout.

With respect to differential diagnosis, heart muscle disease has to be distinguished from hypertensive heart failure, rheumatic mitral incompetence, left ventricular endomyocardial fibrosis, and annular subvalvular left ventricular aneurysm.\textsuperscript{14}

Group Studied

Though the disease is common, in order to be as certain as possible of the diagnosis, only cases in which there was necropsy confirmation, or in which angiography had been performed were included in this series. As a result the series is biased owing to the high proportion of fatal cases.

Of the 34 patients, 22 were male and 12 female; ages varied from 16 to 66 with a mean of 43.8 years. They presented at University College Hospital, Ibadan, between July 1958 and September 1966, and the duration of symptoms was from 8 days to 2 years (mean 5.3 months).

In 20 patients mitral incompetence was considered to be present initially, and in nine patients the diastolic blood pressure was above 90 mm Hg, although in only one patient was the systolic level above 140 mm Hg.

Necropsy was performed in 22 patients, and the findings were in keeping with the description of the pathological features of heart muscle disease recorded by Edington and Jackson.\textsuperscript{8} In all cases the heart weight was increased varying from 360 to 905 g (mean 507 g). In most cases general dilatation of the heart and a flabby myocardium were evident. In nine cases a small pericardial effusion was present, and in one case the amount of fluid was considerable. Ventricular hypertrophy was present in 14 cases, involving both ventricles in nine cases and one ventricle in five cases. In five cases thinning of the left ventricular apex was present, and in two cases this was extreme with reduction of the myocardium to 0.5 cm and to only 3 mm, respectively. Antemortem thrombus was present in one or more chambers of the heart in nine cases. In one case a small submitral left ventricular aneurysm was an incidental finding. Histologically, the ventricular myocardium showed nonspecific changes only.
Radiological Features

For all patients at least one standard posteroanterior chest film was available. In the acute stage of the disease there was invariably a generalized, globular, cardiac enlargement (figs. 1 and 2), and in six cases the contour suggested predominant involvement of the left ventricle. The left cardiac border fre-
HEART MUSCLE DISEASE IN NIGERIAN ADULTS

Figure 3
Kymogram showing normal pulsation of pulmonary artery and right atrium, and greatly reduced left ventricular pulsation.

Figure 4
Angiogram showing enlarged left ventricular chamber and mitral incompetence.

Subsequently had a well-defined, sharp edge. The cause of this stencilled outline is not certain, but it may be attributable to the small amplitude of movement of the ventricles with consequent absence of blurring.

In three instances calculation of the cardiothoracic ratio was impossible owing to pleural effusions, but in the remaining 31 cases it was always above normal, ranging from 0.61 to 0.78 (mean 0.67).

Even with fluoroscopic and barium studies, it proved difficult to determine the relative...
enlargement of the cardiac chambers. The esophagus was usually displaced backward but a frank atrial impression was infrequent. Cardiac movements were reduced, and the left cardiac border was especially affected. The pulsations on the right side, and of the

Figure 5

Angiogram showing enlarged left ventricular chamber, and in the lateral view, little change in size and shape between systole and diastole.
pulmonary artery, could be more vigorous than those of the ventricles. This was readily demonstrable by kymography (fig. 3).

Cardiac calcification was not detected in any patient. Pulmonary venous congestion was present at some stage in all patients, but pulmonary edema was seen in only four, and septal lines were evident in only three.

Pleural effusions were present during episodes of failure in 17 cases (figs. 1B and 2A), being bilateral in 14, right-sided in two, and left-sided in one.

In one patient the aorta was a little dilated, in another it was slightly unfolded, but in all the remaining cases it appeared to be of normal caliber or small. Atheroma at the level of the arch was not encountered.

Where the disease was of the relapsing type, the radiological features followed the clinical course, the heart enlarging and the pulmonary venous congestion appearing during phases of decompensation (fig. 2).

Whether pericardial collections contributed to the enlargement of the cardiac contour that occurred with failure was not determined. Special forms of examination are necessary to ascertain the presence of a small pericardial effusion in these large quiet hearts, and they were not carried out.

Angiography was performed on 13 patients. Some studies were made soon after clinical improvement following the first acute attack; others were performed after an interval of up to 2 years from the initial episode of heart failure.

In 12 patients an angiographic catheter was inserted percutaneously into the femoral artery and passed in a retrograde manner into the left ventricle where contrast medium was injected. In two patients the injection was made selectively into the right ventricle, the catheter having been passed from an antecubital vein. Biplane rapid serial films were obtained with Schonander film changers. The electrocardiogram and the time and duration of the radiographic exposures were recorded simultaneously on a two-channel writer. The injection of contrast often induced a run of extrasystoles, but as a rule, it was possible to obtain views of the opacified ventricle in normal systole and diastole in the later films of the series.

In one case the left ventricular chamber appeared to be of normal size, but in the remaining 11 cases this chamber was enlarged, often greatly so (fig. 4). In all but one case, in which left ventricular contractions appeared normal, the striking feature was the great reduction in the normal variation in size and shape of the left ventricular chamber throughout the cardiac cycle (fig. 5). In three patients the impairment of myocardial contraction was so great that there was almost no difference between films obtained in systole and those taken in diastole. Because of the low ventricular ejection fraction, persistence of contrast occurred within the left ventricle so that it remained opacified for a much longer time than normal.

Mitral incompetence was demonstrated by reflux of contrast material into the left atrium in the eight cases in which it had been suspected clinically (fig. 4). The amount of contrast entering the left atrium could be slight or considerable, depending on the degree of atrioventricular incompetence. In the seven cases in which the left atrium was adequately opacified, it appeared to be of normal size in three, slightly enlarged in one, moderately enlarged in two, and greatly enlarged in one. There was no significant systolic expansion of the left atrium. In five cases thinning of the left ventricular apex was considered to be present. The trabeculae carneae were never very obvious, and no patient showed evidence of ventricular outflow tract obstruction. The coronary arteries appeared to be normal in all cases, and there were no irregularities suggestive of atheroma.

In the two cases in which right ventricular angiography was performed, this chamber was enlarged, and in one case it was also compressed in the anteroposterior plane by the large left ventricle. Throughout the cardiac cycle the changes in the size and shape of the right ventricle were reduced compared with those of the normal heart.
Discussion

Having defined the term "cardiomyopathy," Goodwin\textsuperscript{15} suggested four groups according to clinical and angiographic characteristics: (1) congestive cardiomyopathy; (2) constrictive cardiomyopathy; (3) hypertrophic obstructive cardiomyopathy; and (4) unclassified group (showing features of several of the other groups). Heart muscle disease in Nigeria may be regarded as one of the many conditions comprising the group of congestive cardiomyopathies.

The radiological features conform to the general pattern of this group.\textsuperscript{18} The plain film reveals a large heart in failure, and angiography confirms the poor myocardial function, demonstrates atrioventricular incompetence, and excludes obstruction, or considerable hypertrophy, of the ventricular muscle.

From the point of view of diagnosis, left ventricular angiography may be the only way to differentiate heart muscle disease from rheumatic mitral incompetence, left ventricular endomyocardial fibrosis,\textsuperscript{16} and annular subvalvular left ventricular aneurysm.\textsuperscript{14,17} Hypertensive heart failure may pose some problems in diagnosis, but the presence of considerable left ventricular hypertrophy is a feature in favor of this condition rather than heart muscle disease.

In the Nigerian, coronary artery disease resulting in myocardial fibrosis or infarction is so uncommon that for practical purposes it may be excluded as a cause of heart failure.

The thinning of the apex of the left ventricle, which is occasionally seen on angiography and found at necropsy in a proportion of cases, is of interest, with the exception of Chagas' heart disease,\textsuperscript{12,18} this does not appear to be a feature of other cardiomyopathies.

Recently, in England, it has been suggested that an infective agent may be concerned in the etiology of congestive cardiomyopathy,\textsuperscript{19} but as yet, the cause of heart muscle disease in Nigeria is unknown. It is probable that this term includes several disorders of diverse etiology but similar clinical features.

The relationship of heart muscle disease to the other congestive cardiomyopathies seen in the African continent and elsewhere awaits elucidation.

Acknowledgment

We wish to thank Professor G. M. Edington for permission to study the necropsy records and also Mr. F. Speed for the illustrations.

References

HEART MUSCLE DISEASE IN NIGERIAN ADULTS


---

Old Age Is a Race Between Death and Psychosis

The body, as we know it, was not built
To last forever (forever is a long time); It is usually good for possibly 70 years, More or less; of course, there are exceptions, But Nature, in her manner, has her rules Not always taught (or known) in certain schools.

Perhaps it is well that Death may intervene Before psychosis dominates the scene; It is good to die with an unclouded mind; Senility just several laps behind In that gruelling grind, that marathon Of decade after decade towards the setting sun, The channel of the river clogged with silt, Foundations crumbling where the tower was built, Juices drying, leaves and vines that wilt.
