Radiological Findings in Endomyocardial Fibrosis

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SUMMARY
The radiological findings in 102 cases of endomyocardial fibrosis occurring in Equatorial Africa are presented. Postmortem studies confirmed the diagnosis in 39 cases. Angiocardiography was done in 78. The right side of the heart was primarily involved in 48 patients and the cardiac silhouette was characteristically globular suggesting a pericardial effusion, the large heart having a conspicuous background of oligemic pulmonary fields. Angiocardiography usually revealed a dilated right atrium and ventricle, extremely slow transit of the contrast material, and tricuspid incompetence. Occasionally, a filling defect in the right atrium, interpreted as thrombus, was seen.

The left side of the heart was established as being primarily involved in 13 cases and the picture of rheumatic mitral disease was simulated. Angiocardiography revealed a rather small left ventricle and mitral regurgitation. Biventricular disease was present in 42 patients, the main differentiating point in routine films from right endomyocardial fibrosis being the size of the superior pulmonary veins. The complete diagnosis is difficult to detect without angiographic studies.

Additional Indexing Words:
Cardiomyopathy Mitral regurgitation Angiocardiography
Pericardial effusion Tricuspid regurgitation Calcification of heart

Endomyocardial Fibrosis (EMF) is a disease seen in Equatorial Africa, parts of South America, and rarely elsewhere.¹-³ The pathological features have been well described by Bull and his colleagues of the Kampala School. The clinical aspects and the hemodynamic findings have been recorded from Kampala and from Ibadan by Abrahams.² Though the radiological findings have been briefly summarized and the angiographic features have been described by one of us⁴,⁵ there has been no comprehensive description or analysis of the radiology of EMF.

Cases Studied
This paper describes the radiological findings in 102 patients with EMF. There were 52 males and 50 females. The age range was from 7 to 50 years with 78% in the second or third decade (fig. 1). All patients were Nigerians except for one Caucasian. The diagnosis was established by necropsy in 39 patients and by angiography and catheterization in 78 cases; in some patients both procedures were possible. By insisting on incontrovertible necropsy or angiographic criteria for the confirmation of the disease, we have omitted many cases studied which in all respects were typical clinically, on chest radiography, and hemodynamically. Before describing the radiography of the disease, a brief description of the pathology will make the subsequent discussion clearer.

Pathology
The hallmark of the disorder is the presence of thick white fibrous tissue on the endocardium of the ventricles, which may also penetrate and involve the inner portion of the
myocardium. This fibrous tissue can affect either one or both ventricles and is usually initially deposited at the apex; from there it extends along the inflow tract of the ventricle to implicate the papillary muscles. A consequence of this process is that the leaflets of either the tricuspid or mitral valve become tethered to the ventricular wall and atrioventricular incompetence results. Sometimes the apex of the heart may be spared and the fibrous process is restricted to the inflow part of the ventricle or to a papillary muscle. Ventricular mural thrombi are sometimes seen attached to the area of endomyocardial thickening. The consequences of these pathological processes depend on the situation of the endomyocardial fibrosis as this may affect the right ventricle, the left ventricle, or both ventricles.

With right-sided EMF, there is obliteration of the apex of the right ventricle, dilatation of the outflow tract, tricuspid incompetence, and a greatly enlarged right atrium with thinned-out walls.

In left-sided EMF, there may be partial obliteration of the left ventricle by fibrous tissue and the mitral papillary muscle may be bound down causing mitral incompetence. The left atrium is not usually greatly enlarged.

Cases that are difficult to classify are occasionally seen. In these, histologically associated features such as evidence of rheumatic infection may be seen, or else the fibrous tissue may be slight and atypically placed. No doubt, some of these patients have endomyocardial fibrosis, but for ease of discussion the cases reported herein have been confined to "typical" examples of the disorder and doubtful cases have been excluded. This, of course, may mean we have omitted cases showing the very earliest manifestations of the disorder. The frequency of the different locations of the process are shown in the histogram (fig. 2), but these relative proportions should be considered in the light of the selection factors just mentioned.
ENDOMYOCARDIAL FIBROSIS

Right Ventricular EMF

In the group studied there were 48 patients with right-sided EMF; of these, 29 were males. In chest films, the cardiac contour was invariably found to be greatly enlarged and in 80% the cardiothoracic ratio was greater than 0.7, and in three instances it was greater than 0.9 (fig. 3). The cardiac shape was generally globular and suggestive of a massive pericardial collection. However, even though pericardial fluid was frequently present, probably invariably present in the earlier phase of the disorder, the cardiac shape could be very similar in the absence of pericardial fluid as the large aneurysmal right atrium caused a similar cardiac silhouette (figs. 4 and 5). A convex bulge high on the left ventricular border was sometimes seen even in the presence of a pericardial collection. This prominence, which was pulsatile on fluoroscopy and has been recorded by us by kymographic technique, was due to the diastolic distention of the outflow portion of the right ventricle (fig. 6).

When air was introduced into the pericardial cavity following removal of fluid, the pericardium was invariably seen to be thin (fig. 5). Cardiac calcification was observed

Figure 3
Cardiac size in cases of endomyocardial fibrosis.

Figure 4
Right-sided EMF without effusion. (A) The plain film shows an enormously enlarged cardiac contour, no visible aortic knuckle, but a slight bulge from the dilated outflow portion of the right ventricle. No oligemia of lung fields. (B) Opacified right atrium showing its vast size and the absence of pericardial fluid. The dilated outflow tract of the right ventricle is also visible.
extending from the apex obliquely to just below the outflow part of the right ventricle in three cases (fig. 7).

The pulmonary vasculature showed characteristic changes in right-sided EMF. The enormous cardiac contour contrasted with the oligemia of the lung fields. The cardiac contour opacity hid the hilar vessels, but angiography confirmed that these were smaller than normal. Doubtless the clear lung fields were a consequence of the diminished output of the damaged and deformed right ventricle. The superior mediastinum was widened in some instances because of an engorged superior vena cava. Pleural collections were detectable during the course of the disease in nearly half the cases with an almost equal frequency in right, left, and bilateral effusions.

The ribs and skeletal elements visible in the chest roentgenogram of the younger patients with right EMF were sometimes somewhat osteoporotic with a structure suggestive of a hypertrophic marrow cavity. The common reason for this change in West Africa was that sickle cell anemia was not present, and this feature may be related to the central cyanosis that these patients exhibited. It is
well recognized that such bone change may develop in cyanotic congenital heart disease.\textsuperscript{6}

The angiographic features of right-sided EMF have been described elsewhere,\textsuperscript{5} and in the 35 procedures reported herein the findings were similar. Because of the large right atrium, a venous or atrial injection had to be made with a large volume of contrast medium owing to the inevitable dilution by the considerable volume of blood in these cavities. The catheter tip was kept away from the right atrial appendage for fear of dislodging thrombi which frequently form in this area. With selective right ventricular injections 40 ml of contrast medium is adequate in an adult.

The right atrium was always enlarged, often greatly enough to achieve aneurysmal proportions (fig. 4). Filling defects were occasionally seen in the atrial appendage or extending into the atrial cavity, indicating the presence of thrombi. When injection was made into the right atrium, dilated hepatic veins and a distended superior vena cava were often demonstrated. Sometimes reflux occurred down the azygos vein as the venous valves had become incompetent due to the high venous pressure.

When injection was made selectively into the outflow portion of the right ventricle, contrast material refluxed into the right atrium owing to the tricuspid incompetence (fig. 6).

In many cases the ventricular chamber from the tricuspid valve to the outflow portion had an oblique linear lower border which corresponded to the area of calcification that was sometimes seen. This immobile part of the ventricle contrasted with the vigorous contractions of the distended outflow portion, the shape of which differed markedly between systole and diastole.

The pulmonary vessels were attenuated and this observation confirmed the pulmonary oligemia that was noted in the plain chest films.

The later phases of an examination of the right side of the heart revealed an extraordinarily slow transit of the contrast material.
In cases of large right atrium, contrast persisted in this chamber for 15 seconds after an injection. If the heart was not too large, there was often sufficient contrast to give a faint outline of the left ventricle at 10 to 11 seconds in the anteroposterior view but not in the lateral projection since the continuously opacified right atrium inevitably obscured all detail.

**Left Ventricular EMF**

The number of patients available in this group for analysis was small. Further cases seen were incompletely investigated and, therefore, did not satisfy all the diagnostic criteria necessary for inclusion in this series. In 10 of the 11 cases there was necropsy proof of the diagnosis and in the eleventh the catheterization data and the angiogram were typical. Angiograms in two patients who later came to postmortem examination were also available.

Left-sided EMF was difficult to diagnose from plain films as the heart did not always look abnormal and when abnormal the appearance often simulated rheumatic mitral disease (fig. 8). As can be seen from the scattergram (fig. 3), in two individuals the cardiothoracic ratio was less than 0.5 and in both instances the presence of EMF was first discovered at necropsy. The typical patient with left-sided EMF usually showed a slightly enlarged or a normal-sized heart. There may be a combination of left and right ventricular enlargement following mitral incompetence and pulmonary hypertension but usually the left ventricle was small. The aorta was small. The left atrium was enlarged, but as a rule not greatly so, and certainly never to the extent that occurs in rheumatic mitral incompetence. Curvilinear calcification lying subendocardially in the region of the apex of the left ventricle was seen in two cases (fig. 7). The incidence of this form of calcification may be high, as we have noted a similar appearance in three other patients but they have not yet been adequately investigated.

The pulmonary vasculature showed evidence of pulmonary hypertension with prominent hilar vessels and attenuated peripheral vessels. The pulmonary hypertension can be of a high order. Though perihilar clouding was frequent, pulmonary edema was only noted transiently once and costophrenic septal lines were also seen only twice. The pulmonary veins were not engorged except at a terminal phase.

Angiography of left-sided EMF is best carried out by percutaneous retrograde left ventricular catheterization. Transseptal punctures or direct cardiac punctures are contraindicated because of the greatly distorted anatomic relationships that may be present.

The left ventricular chamber was usually, but not invariably, reduced in size (fig. 9). The shape of the chamber was altered, particularly at the apex and near the posterior leaflet of the mitral valve (fig. 10). When no obvious filling defects or alterations of contour were present, abnormal areas could be noted by a comparison of films taken during systole and diastole when it was noted that some parts of the ventricle were rigid. Contrast medium refluxed into the left atrium which in pure left-sided EMF was enlarged. Slight systolic expansion of the left atrium was sometimes noted. The cusps of the mitral valve, which are usually

**Figure 8**

Plain film of left-sided EMF showing enlarged heart and pulmonary hypertension.
Figure 9

Left-sided endomyocardial fibrosis. Left ventricular angiogram. Note apical filling defect and large left ventricle and gross atrioventricular incompetence. (A) Frontal projection. (B) Lateral projection.

Figure 10

(A) Left-sided EMF in biventricular disease showing small size of ventricle with filling defects and displaced ascending arch due to large right atrium. (B) Left ventricular EMF with the largest left atrium seen in this series. (Systolic film). Note step formation at site of attachment of posterior leaflet of mitral valve.
evident in diastole as partial filling defects could not be seen in left-sided EMF, presumably as they were bound down to the ventricular walls by the fibrotic process. The coronary vessels were normal. Left ventricular myocardial hypertrophy was not a feature, and pericardial collections were not encountered.

**Biventricular EMF**

Biventricular disease was present in 42 patients of whom 22 were males. In all but two the hearts showed a cardiothoracic ratio greater than 0.5 and 77% showed a cardiothoracic ratio of 0.6 or greater (fig. 3). The plain film features were very similar to those of pure right-sided EMF, and the only useful differential point was the size of the upper lobe pulmonary veins. In pure right-sided EMF the pulmonary oligemia was striking and the pulmonary veins were inconspicuous. In 14 patients (33%) with biventricular EMF, the upper lobe veins were obvious or engorged (fig. 11). Pleural collection occurred at some time in 12 patients.

From these observations it is clear that radiographic evidence of left-sided EMF in the presence of established disease of the right ventricle may be very difficult to detect unless angiography is performed. The left ventricle when affected by EMF in biventricular disease is not under as great hemodynamic stress as lone left-sided EMF since it is protected by the low cardiac output of the right side. The functional disturbance on the left side must therefore be relatively severe before decompensation occurs as reflected by pulmonary vein changes. A similar mechanism probably accounts for the left atrium not becoming significantly enlarged. Therefore, changes in the left atrium cannot readily be detected by fluoroscopy, particularly as pericardial fluid may also be present. In consequence, unless left ventricular angiography is carried out, it is impossible to exclude left-sided disease with certainty since murmurs of mitral incompetence can be absent presumably due, once again, to low cardiac output and reduced blood flow.

In the left angiogram in biventricular disease the chamber is seen to be small, considerably smaller than in pure left-sided EMF. There is evidence of atrioventricular incompetence with visualization of a small or slightly enlarged left atrium. Apical filling defects and failure of visualization of the mitral valve cusps are seen as in left-sided EMF (fig. 12). The arterial phase of the examination showed two characteristic features: the large right atrium frequently displaced the ascending arch of the aorta across the midline; and the enlarged right atrium caused displacement of the right coronary artery as it lay in the atrioventricular groove in a most characteristic arc like fashion (fig. 12).

**Discussion**

The assembly of clinical data in EMF has been hampered as hitherto the definitive diagnosis could only be made at autopsy. Angiocardiology has now greatly facilitated the diagnosis of the disease in life, however, and

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**Figure 11**

*Biventricular EMF. Note distended pulmonary veins and hypertrophic marrow changes in this patient who showed considerable central cyanosis.*
has made it possible to recognize those features from which a clinical diagnosis could be made. The difficulties in getting an adequate number of autopsies is exemplified in this series in which the diagnosis was made by autopsy in only 39 cases and by angiocardiography, which has become a routine in only recent years, in 78 patients. At present, diagnosis in life has limited therapeutic value, since therapy is symptomatic, but collection of data from proved cases will undoubtedly aid in the elucidation of the etiology of this idiopathic disorder.

Endomyocardial fibrosis is a disease that can be difficult to diagnose radiologically and clinically. Thus, with a plain chest film which shows a large cardiac contour suggestive of a pericardial collection, one cannot state with certainty whether the patient has merely a pericardial collection, right-sided EMF with a pericardial collection, a grossly enlarged right atrium from EMF, or a combination of pericardial fluid and a large right atrium due to EMF. If there is clear clinical evidence of tricuspid incompetence, the diagnosis of right-sided EMF is easy, and if there is a marked bulge in the pulmonary artery segment, a large pericardial collection is unlikely. However, frequently angiocardiography only can resolve the issue. Similarly, angiography is of value in differentiating constrictive pericarditis which can be difficult on clinical grounds.

We have sometimes found difficulty in differentiating left-sided EMF from mitral subvalvular annular left ventricular aneurysm when the cardiac contour was not altered by an external aneurysmal bulge. The electrocardiogram may be helpful, but only a left ventricular angiogram objectively differentiates between the two conditions. The diagnosis of left-sided EMF in the presence of a small or normal-sized heart is difficult. In neither of the two cases in this series was angiography or cardiac catheterization carried out as both patients died shortly after admission. In neither case was a murmur heard. We do not know whether it is possible for left-sided EMF to occur without significant hemodynamic consequences when the apex is predominantly affected.

Left ventricular angiography is capable of differentiating rheumatic mitral incompetence from left ventricular EMF and this is of importance since the clinical findings in the two disorders are very similar.

We have little information on the angiographic appearance of the earliest phases of the disorder for two reasons: (1) We have only had the opportunity in a few cases of

Figure 12
Biventricular EMF. (A) Gross filling defect at apex and inflow portion of the right ventricle. There is reflux into the left atrium. The right coronary artery is elevated. (Right coronary has been touched up.) (B) Frontal projection showing changes in shape of the ventricular chamber. (C) Lateral view in another case showing very small left ventricle with opacified left atrium. The right coronary artery is elevated.
determining the evolution of the disease by repeated angiography, and thus our experience is too limited to permit firm conclusions. (2) The earliest lesions of the left ventricle will, we believe, require careful analytical study by cineangiography, but our present facilities do not permit us to carry out this technique.

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Osler's account conveys well the great variability in the manifestations and course of rheumatic fever. It is difficult to agree today with his statement that "the recognition of acute rheumatism is very easy." The manifestations vary in intensity as well as in kind, and there is still no specific diagnostic test to assist the clinician in doubtful cases. The only significant advance in laboratory diagnosis of the disease depends on the demonstration of an increase in titer of antibodies to certain streptococcal antigens as evidence for a recent infection with group A streptococci. While these tests are useful, they fall short of conclusively establishing the diagnosis.—Maclyn McCarty. In Osler's Textbook Revisited, edited by A. McGehee Harvey and Victor A. McKusick, New York, Meredith Publishing Co., 1967, p. 141.
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