Pathology of idiopathic cardiomegaly in Jamaica

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Twenty-one cases of idiopathic cardiomegaly which came to necropsy have been reviewed. Investigations included injection and x-ray studies, careful microscopical examination and skip serial sectioning of the conduction systems. The obvious feature was gross cardiomegaly in the absence of atheroma of the large coronary vessels and an associated fibrosis of the right and left bundle-branches. Three hearts showed thickening of the intramural vessels of 80μ to 400μ diameter. We suggest that such changes may be one of the causes of the ischaemic electrocardiographic findings reported in previous epidemiological surveys.

In 1963 Stuart and Hayes described patients with cardiomegaly of unexplained aetiology seen clinically and at necropsy at the University Hospital of the West Indies (U.H.W.I.) in Jamaica. This paper suggested that a form of idiopathic cardiomegaly occurred with some frequency in the Jamaican community. Their patients were mainly from the lower income group, the average length of illness being 18 months. In the majority of patients there was a gradual downhill course though in the early stages congestive cardiac failure was easily controlled. Pulmonary and systemic emboli from cardiac mural thrombi were not uncommon. Hypertrophy of the heart was an incidental finding in those patients not presenting in congestive cardiac failure. Three clinical types were described: (1) simulating ischaemic heart disease; (2) simulating valvular disease; and (3) a constrictive type simulating constrictive pericarditis. At necropsy the hearts showed conspicuous uniform enlargement and dilatation of all four chambers. Myocardial fibrosis was nonspecific and cellular infiltrate was mild or absent. In 9 of the 29 cases there was associated hepatic cirrhosis or fibrosis.

More recent epidemiological studies in Jamaica showed an unexpectedly large number of subjects with electrocardiographic signs of ischaemic heart disease (Fodor et al., 1964). Their survey of two communities in a rural and suburban area showed a higher prevalence of such changes than were found in similar surveys in the U.S.A. or Britain. The cardiothoracic ratio calculated from the antero-posterior chest x-ray was also higher than in a similar study from Wales (Stuart et al., 1962; Ashcroft and Miall, 1969). These findings are difficult to explain in a community in which coronary atheroma and myocardial infarction are not frequent (Robertson, 1959).

Summerell, Hayes, and Bras (1968) analysed the heart lesions in 1500 consecutive adult necropsies performed at U.H.W.I. Whereas 50 per cent of all patients had some heart lesion, with half of these due to hypertension, there were only 30 cases of idiopathic cardiomegaly, i.e. 2 per cent of the adult necropsy population studied and 4 per cent of those showing pathological changes of the heart.

The present study was undertaken to examine in more detail any large heart seen at necropsy which was unexplained by the usually accepted causes of cardiomegaly (see below). The investigation included post-mortem coronary angiography and histological examination of the conduction system of the heart.

Materials and methods
Cardiomegaly was defined as a heart weighing more than 350 g. in women and 400 g. in men.
Cases were accepted as idiopathic cardiomegaly if they fulfilled the criteria proposed by the WHO group of Investigators (World Health Organization, 1968), i.e. excluding the known causes of heart enlargement.
The hearts were photographed fresh and coronary vessels injected with a barium sulphate-gelatine mixture as described by Rodriques (1966). X-rays were taken with the heart intact and unrolled, then the large coronary vessels were dissected and examined directly after being opened longitudinally.

Ten blocks from each heart, to include representative portions of each cavity and valve, were examined histologically. The conduction system was investigated by the technique of Hudson (1963), tissue blocks being processed by a modification of the double embedding method of Péterfi (1921). Skip serial sections were stained routinely with haematoxylin and eosin; additional sections were examined with other stains when indicated. These included elastic van Gieson, Giemsa, and Mallory's trichrome stains.

**Results**

A total of 21 cases was accepted as having idiopathic cardiomegaly. Two of these patients were diabetics for five and two years, respectively. Three others gave a history of heavy alcoholic intake for many years and have been considered separately.

The ages ranged from 19 years to 79 years, with a mean age of 56 years at death. There is no significant difference in the incidence of men and women, though the average age was 54 years for men and 58 years for women.

**Clinical findings**

These are listed in Table 1. The commonest presentation was congestive cardiac failure for which no cause could be found. The clinical course of the illness was typified by persistent or recurrent cardiac failure though death was often sudden at a time when the patient was apparently well compensated. The duration of the illness from first symptoms to death averaged 29 months, with the longest survival being 10 years and the shortest 10 weeks. The 2 patients with evidence of complete heart block suffered from Adams-Stokes attacks, and 3 patients complained of angina.

Auscultatory findings indicated tricuspid incompetence in 1, mitral incompetence in 3, apical systolic murmur in 6, and gallop rhythm in 7 cases.

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<tr>
<th>P.M. No.</th>
<th>Duration of symptoms (yr.)</th>
<th>Age (yr.)</th>
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* Heart injected with micropaque.
TI, tricuspid incompetence.
MI, mitral incompetence.
RPCFT, Reiter protein complement fixation test.
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<td>-</td>
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On electrocardiographic examination, 11 patients showed some form of conduction defect, 2 had nonspecific myocardal damage, 1 had left atrial hypertrophy, and 4 had left ventricular hypertrophy.

**Histopathology** (see Table 2) Gross cardiomegaly was present in all cases with an average heart weight of 675 g., two hearts weighing more than a kg. each. The typical globular-shaped appearance is seen in Fig. 1.

Biventricular dilatation and hypertrophy was the rule, usually with dilatation of the atria also. In some cases this was so conspicuous that the atrial wall was reduced to paper-thin dimensions. Pericardial abnormalities were limited to an occasional ‘milk spot’ on the anterior aspect of the heart. The myocardium was usually of normal consistency though in 3 instances it could be described as flabby. In only 2 hearts was there easily discernible fibrosis in the gross specimen. Endocardial thickening was not a constant feature and was usually limited to irregular mild fibrosis in the outflow tract of either or both ventricles and in the right atrium in association with mural thrombi. In 4 hearts small mural thrombi were found in the apices of both ventricles.

Valves were in all cases normal apart from a minimum degree of thickening of the mitral valve leaflet in the elderly patients. The mitral and tricuspid valve rings were usually dilated, however. The large coronary vessels were free from atheromatous narrowing and in the majority of cases presented a normal picture when opened longitudinally.

Coronary angiography was performed on 7 hearts. Fig. 2 is a typical result. The vessels had a smooth outline and were straight with no evidence of occlusion or narrowing.

Histological examination showed a variety of mainly nonspecific features. The epicardium, particularly of the atria, contained small foci of round cells and sometimes a little fibrous thickening but no active pericarditis. Myocardial fibres generally showed hypertrophy with prominent nuclei of bizarre shape (Fig. 3). Some nuclei were blunt ended and had a coarse speckling of chromatin, others
TABLE 2 Gross findings

<table>
<thead>
<tr>
<th>P.M. No.</th>
<th>Heart weight (g.)</th>
<th>Pericardial effusion</th>
<th>Pericardial adhesions</th>
<th>Hypertrophy</th>
<th>Dilatation</th>
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<td>RV</td>
<td>LA</td>
<td>LV</td>
<td>RA</td>
<td>RV</td>
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<tr>
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<td>-</td>
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<td>610</td>
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<td>-</td>
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<td>-</td>
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<tr>
<td>5482</td>
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<td>560</td>
<td>-</td>
<td>-</td>
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Diabetics

| 5752     | 635              | -                    | -                    | +           | +         | +         | |
| 5933     | 580              | -                    | -                    | -           | -         | -         | |

Alcoholic cardiomyopathy

| 5244     | 515              | -                    | -                    | +           | -         | -         | |
| 5391     | 430              | -                    | -                    | +           | -         | -         | |
| 5892     | 390              | -                    | -                    | -           | +         | +         | |

RAA, Right atrial appendage; LAA, Left atrial appendage; RA, Right atrium; LA, Left atrium; RV, Right ventricle; LV, Left ventricle.

were elongated and wormlike, while staghorn forms predominated in some areas. Parasites were invariably absent.

Fibrosis appeared in a variety of patterns. It was most frequent in the atria and consisted of dense, poorly cellular, fibrous tissue separating single muscle fibres. The ventricles showed a different pattern, the commonest being a mild diffuse, reticular fibrosis between the muscle bundles, with occasional confluent areas. Two cases, however, showed peri-vascular fibrosis while a third case showed widespread patchy fibrosis.

The two hearts with grossly visible lesions had coagulation necrosis of the myocardium with organization and all the features of infarction but without coronary artery occlusion; there was no large coronary artery disease. Myocytolysis was seen in hearts in variable amounts.

FIG. 1 A typical case of idiopathic cardiomegaly (heart weight 850 g.).
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**Conduction system and small blood vessels** The sino-atrial node showed an increase in fibrous tissue in 3 cases though in none was there complete destruction. Recent haemorrhage was seen in one node. The adjacent right atrial myocardium was frequently the site of patchy fibrosis but changes in the node were usually less obvious. The atrioventricular node showed similar changes with some fibrosis in 3 hearts, and haemorrhage and lymphoid cell infiltrate in 1, but in none was there any recent necrosis.

The bundle of His showed similar changes those seen in the atrioventricular node. The left bundle-branch was the site of widespread lesions in 13 of the 15 hearts. These included interruptions by fibrous tissue of the areas of the bundle of His as they enter the bundle-branch, patchy fibrosis along the bundle in short and long strips, and sometimes actual necrosis. Sometimes these changes were associated with endocardial thickening in which the bundle was involved. The right bundle showed patchy fibrosis in 4 cases (Fig. 4) and necrosis in one.

The investigation of the conduction system required skip sections of the upper and middle portion of the interventricular septum. In three of the hearts so examined (Nos. 5482, 5333, and 67/3) we noticed that many

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</table>

2 An angiogram with the heart und illustrated the straight, smooth, coronary arteries.
FIG. 3  *Bizarre nuclei in the myocardium. (H. and E. × 500.)*

FIG. 4  *Fibrosis of the right branch of the bundle of His (indicated by arrows). (H. and E. × 390.)*
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**FIG. 5** (a) Intramural vessel showing thickened media and reduction of lumen. (H. and E. × 115.) (b) Intramural vessel showing intima thickened by fibro-elastic tissue with reduction of lumen. (Elastic van Gieson. × 115.) (c) Greatly reduced lumen, the wall of the vessel consisting of fibro-elastic tissue. (Elastic van Gieson. × 115.) (d) Normal intramural vessel. (H. and E. × 115.)
intramural vessels extending over several millimetres showed thickening. In nearly all
the thickened vessels there was a reduplication of the elastic tissue which sometimes
extended into the media or into the intima. Some vessels showed besides this a reduction
of muscle in the media with or without increased fibrous tissue and increased fibrosis
of the adventitia; others showed a medial thickening of both circular and longitudinal
muscle but the adventitia showed less fibrosis; still others showed intimal thickening made
up of concentric rings of elastic and connective tissue (Fig. 5). There was consequently
a narrowing of the lumen, very obvious in some instances.

It should perhaps be reiterated that these changes in the intramural vessels occurred
without a concomitant lesion in the large coronary arteries.

Alcoholic cardiomyopathy The three cardiomegalies believed to be of alcoholic aetiology
were in men with an average age of 43 years. Symptoms were few, and only 1
case presented with congestive cardiac failure in life. The other 2 cases were discovered at
necropsy. The average heart weight was 444
and 1 heart showed thrombi in the left ventricle and mild generalized thickening of the
endocardium.

The histology was similar to that described above, except that it was not so severe, and
damage to the conduction system was limited to some mild patchy fibrosis of the left bundle
of one heart.

Discussion

Idiopathic cardiomegaly as defined at the
WHO Conference on Cardiomyopathy, Ja-
maica, 1967 (World Health Organization,
1968) continues to be seen at U.H.W.I.
(Summerell et al., 1968).

We have confined this report to hearts for
which no explanation for hypertrophy has
been found even after considerable investi-
gation. Also excluded were cases of sudden
death before full clinical investigation, thus
avoiding as far as possible cases of undetected
hypertension, anaemia, and other systemic
disease.

The typical idiopathic cardiomegalic heart
is much enlarged (average weight 675 g.)
and globular in shape. Its gross picture is un-
like the hearts described by Hill, Still, and
McKinney (1967) of which several were below
the heart weight for conforming with the
WHO criteria for cardiomegaly. Endocardial
thickening is not a conspicuous picture in
the hearts described by us, though we have
seen it in mild degree in hearts associated
with an alteration of the haemodynamics. In-
deed a heart as depicted in Hill et al. (Fig. 1)
has never been observed in our University
Hospital, and endomyocardial fibrosis as
described originally from Uganda (Davies
and Ball, 1955) is unknown in Jamaica among
cardiologists and pathologists. Hill et al.
(1967) further included 2 cases of pulmonary
tuberculosis; such cases may have to be ex-
cluded by the WHO criteria, i.e. whenever
cardiac lesions develop as a result of the pul-
monary conditions. Again there was a case
where death followed caesarian section in a
19-year-old girl. This would have to be scruti-
nized as possibly being peripartal or other
heart disease.

Clinically, idiopathic cardiomegaly is un-
like nutritional heart disease reported by Gil-
landers (1951) from Africa. None of our
patients was significantly malnourished, and
we have seen no association with cirrhosis of
the liver in this present series, though the
association was noted in the previous paper
by Stuart and Hayes (1963) and in nutritional
disease by Gillanders (1951). Morphologically
there are similarities, the hearts described by
Gillanders being also generally hypertrophied,
dilated, and without significant endocardial
thickening.

Heart muscle disease (Eddington and Jack-
son, 1963), cardiovascular collagenosis (Bec-
ker, Chatgidakis, and van Lingen, 1953),
cryptogenic heart disease (Higginson, Isaac-
on, and Simson, 1960), and primary myo-
cardial disease (Mattingly, 1961) are other
names given to cardiac conditions with many
similarities to each other and to our cases of
idiopathic cardiomegaly. The frequency of
these conditions in tropical and developing
areas suggests that there may be common
factors in their aetiology, and dietary habits
have been considered by many writers. We
have no evidence of any specific aetiology
among our patients.

Three of the patients in our series gave a
clue to history of alcohol excess, and their hearts
showed no morphological difference from the
rest, though microscopically the conduction
systems in all 3 were unaffected. It has been
reported in a recent survey that heavy alcohol
consumption is on the increase in certain
social groups in Jamaica (Beaubrun, 1968).
The clinical and social histories of most of our
patients, however, make an alcohol aetiology
unlikely.

Puerperal cardiomyopathy has been fol-
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lowed clinically in this area for several years (Stuart, 1968). One case, which does not appear in this series, has recently come to necropsy and was found to differ in no way from the cases described above.

A striking feature of our series of cardiomegalic hearts is the excellent state of the large coronary arteries, though many of the cases were over the age of 50 years and 3 were in the 70’s.

- In the 3 cases in which there was conspicuous thickening of the intramural vessels, there was minimal or no atheroma in the large coronaries. The changes were confined to vessels of 80μ-400μ diameter, and the intima, media, and adventitia were variously involved; one of the hearts weighed 1140 g.

- James (1967) has described changes in the small coronary vessels similar to those which “we have seen, but all the cases had a familial hereditary disease. So far we have not been able to demonstrate any familial occurrence in this series. The myocardial arterioles have also been examined by More and Sommers (1962) in angina pectoris, but in all their cases there was an association with atheroma in the large coronary vessels. Donomae et al. (1962) have examined postmortem material with mild atheroma of the large coronary vessels and cardiomegaly and reported many of the changes we have seen, but as the clinical history of many of these cases was complicated by hypertension, syphilis, and other systemic disease we are unable to draw a parallel.

Bundle-branch block is a common electrocardiographic finding, and in all our cases examined histologically there was a correlation with the microscopic finding of fibrosis interrupting conducting tissue. Davies (1967), when examining cases of chronic complete heart block, found fibrosis in the bundle-branches in 36 of 47 cases, and in 19 of these it was unassociated with damage to contractile myocardium. He referred to this entity as ‘isolated disease of the conduction system’ but did not indicate whether these hearts were enlarged.

We have excluded cases of active myocarditis from this investigation, but myocarditis has to be considered as a possible aetiological factor for focal myocardial fibrosis.

- In only one of our cases were virological studies performed in life during the early weeks of illness (PM 5899), and the results were negative. Myocarditis in our necropsy population is not particularly different from other places (Hayes and Summerell, 1966), and if myocarditis were the cause of cardiomyopathy we would have to postulate that the conduction system was almost exclusively affected in some cases. Such a case was described in a Jamaican by Hudson (1965).

We feel that the changes we have described in the intramural coronary arteries may be important in the pathogenesis of idiopathic cardiomegaly, and we propose to concentrate future research on study of the fine vasculature using microangiographic and other techniques.

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