ENDOMYOCARDIAL FIBROSIS: CLINICAL STUDY OF NIGERIAN CASES

By U. Basile*, R. Carlisle and A. O. Falase

Endomyocardial Fibrosis (EMF), in its endemic form at least, was probably first reported by Bedford and Konstam[1] in 1946: their cases were soldiers seen during the North African campaign but coming from both East and West Africa. J. N. P. Davies[2] has, with co-workers[3] and successors[4,5], provided a series of descriptions of the disease from Makerere, Uganda. Studies on the subject have also been published from Ibadan, Nigeria[6-8].

We have heard hitherto in this symposium, of diseases which are "Primary Myocardial" in that the primary process is located in the myocardium and it is not a matter of disease elsewhere—as in systemic hypertension or congenital shunts—afflicting an otherwise healthy myocardium by haemodynamic overload.

We would not include endomyocardial fibrosis in this nosological group since the process involves mainly and severely the endomyocardium whereas only the inner third of the myocardium shows marked changes.

In endomyocardial fibrosis the problem does seem to be primarily haemodynamic; diastolic restriction or systolic shunting, or both, and the myocardium splinted, probably, but not weakened or stretched as in the other entities we have been hearing about.

We are presenting some aspects of the accumulated clinical experience of EMF as documented in the Ibadan W.H.O. Cardiac Registry. In doing so I will omit the haemodynamic, angiographic and pathologic features since they have already been described by the previous speakers, Prof. Goodwin and Doctor Olsen.

Fig. 1 shows the distribution of diagnosed non-paediatric cases by sex and by age at presentation. Only few paediatric cases are represented. The symbol C indicates the proportion of cases confirmed by catheterisation, operation or necropsy and the symbol + those confirmed by necropsy. It is as well to check these proportions against our totals as we proceed, so that we can be alerted to clinical under or over-diagnosis. A wholly disproportionate number of male teenagers (i.e. ten plus) is present. This excess, absent in other heart diseases, must reflect exposure, at or before this age, to an environmental factor of high sexual predilection.

Dividing the whole group by lesion site there were 77 patients with right ventricular (RV) 76 with biventricular (BV) and 25 with left ventricular (LV) involvement.

Looking at proportions of confirmed cases, we note a deficiency of non-confirmed, i.e. clinically-diagnosed, left ventricular endomyocardial fibrosis. These were, in fact, only 6 whereas there were 31 and 34 non-confirmed cases in the groups with RV and BV disease respectively. This is to be explained in great measure as loss by misdiagnosis to rheumatic heart disease or "functional" mitral incompetence.

The number of patients of stated origin in Ibadan is much lower than that expected on the basis of a control sample of 178 hospital patients randomly selected but matched within the same period (i.e. within the same 1,000 serial numbers) of presentation. Fig. 2 shows the rate observed/expected cases for various areas of the country. In contrast to Ibadan, Ijebu division leaps into prominence as a source of the excess of observed over expected.

This is a forest area, with a high incidence of certain parasitic diseases; among them ascariasis, filariosis and schistosomiasis. This may be relevant in view of our finding of microgranulomata in an undue proportion of postmortem livers in endomyocardial fibrosis[9] and in view of the known association of systemic eosinophilia and intraventricular lesions. Eosinophilia is in fact a feature of Loffler's disease and has as well been found in cases of EMF occurring in Europeans resident in Tropical Africa.[10] Furthermore heart lesions indistinguishable from EMF have been described in cases of eosinophilic leukaemia.[11]

Looking at frequency of symptoms at presentation, lesions of each ventricle appear additive in their effects, with regard to abdominal swelling. Effort dyspnoea is shown as the commonest symptom. In right ventricular cases we must presumably blame at least partially the loss of functioning lung tissue owing to accumulation of ascitic and pericardial effusions.

Proptosis is common only with right sided or bilateral involvement. This is in line with the known association of proptosis and tricuspid incompetence.

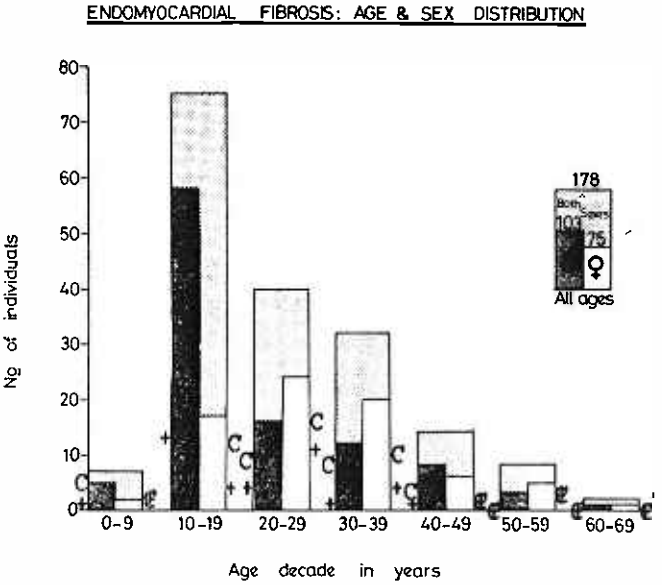

Fig. 1

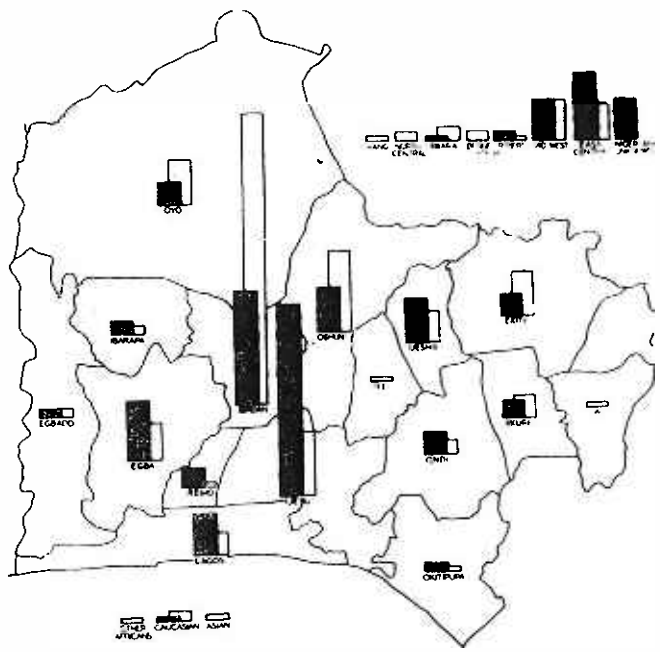

Fig. 2

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It has previously been regarded as a hallmark of endomyocardial fibrosis that ascites is present without oedema. We must ascribe this impression to the relatively higher susceptibility of the oedema to diuretic treatment.

The venous pressure in the veins of the neck and head was noted as elevated in all but 16. It is important, however, to warn that the sign, gross in right ventricular cases, may be missed unless the patient is examined in an upright position as the level may be above the angle of the jaw.

In view of the known enlargement, at times gross, of the pulmonary outflow tract in right ventricular endomyocardial fibrosis, it was expected that pulsation over the pulmonic area would differentiate the ventricle affected. This is so to a certain extent. A right ventricular heave is more frequent in left ventricular disease probably due to secondary pulmonary hypertension.

A third heart sound is usually heard in endomyocardial fibrosis, at times as early and high-pitched as the "knock" of constrictive pericarditis. It was heard in 72% of the patients with RV, 63% with BV and 55% with LV lesion. It was recorded as absent in less than 10% in each of the three groups, and not stated in the remainder.

Pericardial effusion is virtually limited to right ventricular cases—either in isolation or combined. Pleural effusion is non-discriminatory.

Pulmonary conus enlargement in isolated right ventricular cases, where there is no pulmonary hypertension, is perhaps a response on the part of the unaffected portion of the right ventricular myocardium to the loss of function of the remainder. A type of compensatory hypertrophy.

Left atrial enlargement is highly specific—due, we presume, to the mitral incompetence of the left ventricular form. Right atrial enlargement is shown as the result of right ventricular disease or somewhat less commonly, as a result of pulmonary hypertension.

SUMMARY

We have analysed 178 cases of the manifest, late stages of endomyocardial fibrosis. It is seen mostly in adolescent with male preponderance. Unexpectedly large numbers come from Ijebu division.

The venous pressure is raised, there is dyspnoea and, in many, ascites. Peripheral oedema is present. In the left-sided form mitral incompetence causes left atrial enlargement and right ventricular heave. In the right ventricular or biventricular forms there is frequently pericardial effusion. The pulmonary conus may be enlarged in all forms.

Left ventricular endomyocardial fibrosis appear to be underdiagnosed in life. A careful search for a third heart sound would help here, although not absolutely specific in the absence of an opening snap.[12]

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THE BIOCHEMICAL BASIS OF CARDIOMYOPATHY IN THE SYRIAN GOLDEN HAMSTER

By K. G. Nair

The early biochemical events that occur in the myocardium of the Syrian golden hamster with a hereditary form of myopathy (B10 14.6 strain) were studied by the author. Spontaneous cardiac dystrophic lesions appear in the myocardium by the thirtieth day of life. This is followed by cardiac hypertrophy and failure.

The key enzyme in the genetic mechanism of protein synthesis, RNA polymerase, is increased in activity at the time of appearance of the myocardial lesions. The activities of the two major forms of RNA polymerase in the cell, nucleoplasmic and nucleolar, were studied. Alpha-amanitin, a toxic peptide from the common poisonous mushroom, was used to distinguish between the activities of the two enzymes. Elevated adenyl cyclase activity was also noted in heart muscle. The possible interrelationship between catecholamines, adenyl cyclase and RNA polymerase will be discussed. Increased catecholamine content in the myocardium may play an important role in this form of hereditary cardiomyopathy. Animal models of cardiomyopathy give us a valuable insight into the biochemical and cellular mechanisms of the disease process.

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