SESSION II

Chairman: DR. E. G. J. OLSEN

Cardiomyopathies in South Africa – a brief survey of the problem and current therapeutic approaches

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CARDIOMYOPATHY is a common form of heart disease amongst the South African black population (Bradlow, Zion and Fleishmann, 1964; Seftel, Metz and Lakier, 1973), and it is by no means rare in Caucasians.

The aetiology in South Africa, as elsewhere, is totally unknown. The conventional clinical classification into congestive, restrictive, obliterative and hypertrophic forms is at best descriptive of the major haemodynamic features and considerable overlap exists between the various sub-groups. The macro- and microscopical features are equally non-specific. In many instances one does not even know if one is dealing with different entities or varying stages of the same disease. It was hoped that the multi-centre study of this world-wide problem, particularly the electron-microscopic studies, might provide new clues in the search for aetiological agents and eventually a rational approach to prevention and treatment.

The commonest clinical presentation encountered in South Africa is that of congestive cardiomyopathy (Table 1). Patients are noted to have large hypertrophied, dilated, poorly-contracting left ventricles resulting in severe biventricular failure. Murmurs of functional atrioventricular valve incompetence are common. Mural thrombus with systemic embolization and endocardial thickening presumably due to organization of mural thrombus are common. A sub-group of congestive cardiomyopathy, puerperal cardiomyopathy, is identical in all respects except that it carries a slightly better prognosis, some cases recovering completely if subsequent pregnancies are avoided. The congestive cardiomyopathies form the bulk of case material, comprising 62% of the total series.

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Restrictive cardiomyopathy is very much less common and the author has collected eight cases during the last 15 years (2%). The distinctive feature in these cases is the small, normal or only slightly increased heart size with a restrictive haemodynamic pattern mimicking constrictive pericarditis. It usually involves both left and right sides of the heart, and mitral and tricuspid systolic and diastolic murmurs due to valvular involvement by the endocardial fibrous plaques are common. Systolic function of the ventricles is impaired to a variable degree.

The third group, obliterative cardiomyopathy, has been encountered in four cases (1%); three of these were Caucasians who had previously lived in tropical Africa (Beck and Schrire, 1972). All had typical features of endomyocardial fibrosis involving the left ventricle. The most recent case, a young Bantu girl from the Transkei, presented with severe right-sided heart failure due to obliteration of the right ventricular apex, and inflow region with tricuspid valve involvement resulting in both stenosis and incompetence. Systolic function of the left ventricle was normal. Unlike the Nigerian cases of right-sided endomyocardial fibrosis the right atrium was not massively dilated possibly because the thickened endocardium extended into this chamber rendering it non-compliant. Three cases associated with eosinophilia of various causes were encountered and labelled Löffler's eosinophilic carditis. These presented with mixed congestive and restrictive haemodynamic features. It is unlikely that this is a specific disease as it occurs in a wide variety of conditions associated with eosinophilia. It may also represent the acute phase of the restrictive and obliterative cardiomyopathies of non-tropical regions.

Hypertrophic obstructive cardiomyopathy is

TABLE 1. Race and sex distribution of cardiomyopathies

	White male	White female	Coloured male	Coloured female	Black male	Black female	Total	%
Congestive	54	16	48	24	45	27	214	62.0
Hypertrophic	47	25	14	21	0	0	107	31.0
Idiopathic submitral aneurysm	0	0	0	0	8	6	14	4.0
Restrictive	1	1	1	1	2	2	8	2.0
Obliterative	0	3	0	0	0	1	4	1.0

apparently very rare in black patients in Africa. No single case has been encountered but two patients have been reported from the Durban Medical School where a larger black population is cared for (Lewis *et al.*, 1973). Amongst the Caucasian and Coloured population groups hypertrophic obstructive cardiomyopathy is as common in South Africa as elsewhere.

Idiopathic sub-mitral left ventricular aneurysms (14 cases, 4%), is another obscure heart muscle disease common in tropical Africa and occasionally seen in South Africa (Beck and Schrire, 1969). It is a remarkable condition consisting of a small myocardial defect in the sub-annular mitral position communicating with a large false aneurysm. Distortion of the mitral valve apparatus results in severe mitral incompetence; myocardial function is usually not impaired. All the author's patients have been cured by surgery and he has not been able to examine a case at post-mortem.

Therapeutic considerations

Congestive cardiomyopathy

Ignorance regarding aetiology and pathogenesis make preventative measures impossible. However, the sparsity of irreversible structural abnormalities in these cases is one feature suggesting that a potentially reversible situation may at times exist.

In practice, conventional medical therapy has for the most part been palliative and very unsatisfactory. It is believed, however, that therapy should be vigorously pursued because occasionally remarkable recoveries are achieved even in patients with long-standing (longer than one year) congestive heart failure.

Burch, Walsh and Black (1963) claimed good results from treatment with prolonged bed-rest (1 year). Many patients, however, relapsed on resumption of normal activities and in under-developed areas such treatment is impracticable.

It has also been claimed that in cardiomyopathies where alcohol abuse is a factor, abstinence may result in a return to normal function in early cases

(McDonald, Burch and Walsh, 1971). Reversibility may also be seen in the puerperal cases.

The role of systemic hypertension (Falase, 1977) in cardiomyopathy has been difficult to evaluate. Many patients present initially with considerable hypertension but this may be reactive and a compensatory response to the low-output state. In others it is more persistent but is seldom accompanied by other evidence of hypertensive vascular disease. Pathologically the hearts frequently show macro- and microscopic features of hypertrophy. These considerations suggest that long-term oral vasodilator therapy may be an appropriate form of therapy and there is a plan to investigate this shortly.

Some patients manifest with a disproportionate tachycardia, and good results have recently been achieved by Swedish investigators (Waagstein *et al.*, 1975). One brilliant result was obtained, by the author and his colleagues, using β -blockade in a woman with long-standing congestive cardiac failure and disproportionate tachycardia (Fig. 1), but in another it caused more severe myocardial depression ending in a state of cardiogenic shock. Clearly, careful haemodynamic monitoring is required while assessing the effect of β -blockade initially, as in those patients where the tachycardia is a compensatory mechanism in maintaining the cardiac output, disastrous results may be expected.

In a few carefully selected patients cardiac transplantation has proved to be a satisfactory form of treatment. Three of four cardiomyopathy patients are alive and well 12 and 6 months after heterotrophic transplantation. The last case was of interest in that severe irreversible rejection of the transplanted heart occurred three months after surgery. At that time the patient's own heart had shown such tremendous improvement that the rejected heart could be removed without the need for a new implant. He continues to be well and asymptomatic on a small dose of steroid (Fig. 2).

It is interesting to speculate whether the recovery of the heart failure in this patient was, (1) unrelated

FIG. 1. PA chest radiographs of patient treated with β -blockers. (a) before therapy, (b) after 4 months of therapy, (c) 2 months after discontinuing therapy, (d) 2 months after restarting beta-blockers.

to the procedure; (2) related to temporary haemodynamic unloading; (3) related to steroid therapy; (4) due to some undefined effect of the procedure.

Cardiomyopathies with endocardial involvement

Patients presenting with the obliterative and restrictive forms of the disease are in many ways a totally different therapeutic problem. Pathologically

the emphasis falls on the ventricular endocardium and atrioventricular values. In many patients the disease is apparently confined to either the left or the right side of the heart. The major portion of the myocardium may be little affected and systolic function of the affected ventricle may be remarkably well preserved (Beck and Schrire, 1972).

These features suggest that surgical stripping of the obliterative endocardial fibrous mass combined

FIG. 2. PA chest radiograph of patient subjected to heterotopic cardiac transplantation. (a) before surgery, showing massive cardiomegaly. (b) 1 month after surgery, showing the heterograft in the right hemithorax. (c) 3 months after surgery, showing reduction in heart size of bypassed heart. (d) following removal of the rejected heterograft – a sustained reduction in heart size is evident.

FIG. 3. Right heart ciné-angiogram from a patient with endomyocardial fibrosis of the right heart. (a and b) systolic and diastolic pictures showing the small right ventricular cavity with apical obliteration, normally contracting outflow portion and narrowed tricuspid orifice. (c and d) the same patient post-operatively following excision of the fibrous endocardial fibrous mass and tricuspid valve replacement with a Bjorck-Shiley prosthetic valve. Note that the right ventricular cavity has been only marginally enlarged and that the prosthetic valve is rather small.

FIG. 4. Chest radiographs of patient with a subannular left ventricular aneurysm, (a) before surgery, (b) after surgery, (c) the pre-operative left ventricular angiogram shows the normal left ventricular cavity size, the narrow neck and the large double-chambered false aneurysm filling with contrast.

with prosthetic replacement of the affected atrio-ventricular valve may be an appropriate form of therapy. Successful operations of this kind have already been reported (Lepley *et al.*, 1974; Weyman, Rankin and King, 1977). This therapeutic approach was recently attempted in a young African patient with a right-sided lesion. Unfortunately, a poor result was obtained owing to incomplete excision of the right ventricular endocardial fibrous mass with inadequate relief of the restrictive features and increase in right ventricular cavity size. In addition the extremely small shrunken tricuspid valve ring necessitated the placement of a very small prosthetic valve (Fig. 3). Further experience with this procedure may well show that left ventricular disease is more amenable to surgical treatment.

Idiopathic left ventricular aneurysms

These are ideal cases for surgical treatment (Schrire and Barnard, 1963). Resection of the aneurysm and suture of the ventricular wall defect results in complete cure of most cases (Fig. 4). Residual mitral incompetence is uncommon. Only one out of fourteen cases has needed a mitral valve replacement.

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