M-mode echocardiographic features of endomyocardial fibrosis

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SUMMARY M-mode echocardiographic findings are described in 21 patients with endomyocardial fibrosis. Features associated with right ventricular endomyocardial fibrosis include: (i) exaggerated motion and thickening of the anterior right ventricular wall; (ii) increased right ventricular end-diastolic dimension; and (iii) paradoxical septal motion.

Pericardial effusion (viz an echo-free space behind the posterior left ventricular wall) was shown in three patients. The tricuspid valve was easily recorded in all. Six patients with left ventricular endomyocardial fibrosis had diminished left ventricular end-diastolic dimension. Three had echo features of pulmonary hypertension (viz reduced e-f slope, absent a wave in sinus rhythm, and systolic notching of the pulmonary valve echogram). Fine fluttering of the anterior mitral valve and tricuspid valve echo was observed in two patients (one of whom was in sinus rhythm) with biventricular endomyocardial fibrosis, and no angiographic evidence of aortic regurgitation.

Endomyocardial fibrosis, a form of obliterative cardiomyopathy, is one of the commonest forms of heart disease in Nigeria^{1,2} and other developing countries in Africa^{3,4} south of the Sahara. The published reports abound in information on the clinical,^{5,6} haemodynamic,^{7,8} angiographic,^{9,10} and pathological^{11,12} features of endomyocardial fibrosis. Information is, however, scanty and sketchy on the echocardiographic features.^{10,13}

While the clinical diagnosis of advanced right ventricular or biventricular endomyocardial fibrosis is easy and straightforward, difficulties often arise in mild to moderate cases, when it may be difficult to distinguish at the bedside between endomyocardial fibrosis, rheumatic valvular disease, constrictive pericarditis, and congestive cardiomyopathy.¹⁴ If characteristic echocardiographic patterns of endomyocardial fibrosis can be defined, this would offer a useful non-invasive diagnostic tool in the hands of clinicians, obviating the need for cardiac catheterisation, with its attendant risks. This is a consideration of great importance, especially since the role of surgery in this disease is not yet clearly defined.

We have studied 21 patients with endomyocardial

fibrosis by M-mode echocardiography, and our observations form the basis of this paper.

Subjects and methods

There were 10 male and 11 female patients, whose ages ranged from 6 years to 70 years. They were referred to the Lagos University Teaching Hospital with diagnoses varying from puerperal cardiomyopathy, endomyocardial fibrosis, ascites of unknown cause, unexplained cardiomegaly, to pericardial effusion. Each had an examination on electrocardiogram and a chest x-ray film. Ultrasound examination was carried out using standard techniques, with an SKI Ekoline 20A Ultrasonograph and a Cambridge strip-chart recorder. A 2.25 MHz transducer with a repetition rate of 1000 impulses/s was placed in the left parasternal position, with the patient supine.

Fifteen patients had cardiac catheterisation and angiocardiography, 10 before and five after the echocardiographic examination. In two patients, phonocardiograms were recorded, and aortic root angiography carried out. One patient, who died, came to necropsy.

For comparison, two patients with massive ascites resulting from portal hypertension, two with constrictive pericarditis, and one with tricuspid stenosis, all

with clinical appearances similar to endomyocardial fibrosis, were also studied echocardiographically.

Results

Nineteen patients presented with the characteristic clinical picture of right ventricular endomyocardial fibrosis, namely, raised jugular venous pressure, massive ascites, and minimal pedal oedema. Two presented with only mild to moderate ascites. One, a 10 year old boy, had unexplained cardiomegaly. Twelve had clinical evidence of left as well as right heart failure, with a pansystolic murmur of mitral regurgitation. Fifteen patients had clinical evidence of tricuspid regurgitation. An early diastolic murmur was heard in two patients. The radiological and electrocardiographic findings were compatible with the clinical diagnosis of endomyocardial fibrosis. Eighteen were in sinus rhythm and three in atrial fibrillation. There was no left bundle-branch block. The haemodynamic and angiographic study indicated that nine had right ventricular, and six had biventricular endomyocardial fibrosis. Five patients were studied echocardiographically before being subjected to cardiac catheterisation. Of these, two who had aortic root angiograms had no evidence of aortic regurgitation.

The following echocardiographic features were common to all 21 patients.

(1) Increased right ventricular dimension (mean

47 mm; SD 11 mm: normal range 7 to 23 mm) (Table), (Figs. 1 and 2).

(2) Paradoxical septal motion (Fig. 1).

(3) Thickening of the right ventricular anterior wall (mean 9 mm; SD 2 mm: normal < 5 mm), with increased right ventricular anterior wall motion (Fig. 3).

(4) Easily recordable tricuspid valve (Fig. 4). These features were associated with right ventricular involvement. Other features found associated with right sided endomyocardial fibrosis were as follows: (a) Two patients (each with an early diastolic murmur) had fine fluttering of the anterior tricuspid valve leaflet (Fig. 4) (one of these patients was in sinus rhythm); (b) three patients had posterior pericardial effusion (Fig. 5). Features associated with left ventricular endomyocardial fibrosis were as follows: (a) Reduced left ventricular dimension (mean 24 mm; SD 6 mm: normal range 35–56 mm) (Fig. 5), (Table). This was found in all 12 patients with biventricular endomyocardial fibrosis. (b) Two patients, who had early diastolic murmur, had fine fluttering of the anterior mitral valve leaflet (Fig. 6). One of these patients was in sinus rhythm. Two other patients in atrial fibrillation who had anterior mitral valve fluttering did not have an early diastolic murmur. (c) Five patients had left atrial enlargement, and (d) three of them showed echocardiographic features of pulmonary hypertension, viz reduced e-f slope, absent a

Table Clinical, echocardiographic, and haemodynamic data in 21 patients with endomyocardial fibrosis

No.	Age (y)	Sex	Echocardiographic data			Haemodynamic data		Clinical data				EMF involvement (ventricle)
			RVEDD (mm)	EDARVWT (mm)	LVEDD (mm)	RVEDP (mmHg)	LVEDP (mmHg)	MR	TR	ECG	EDM	
1	25	F	40	7	34	18–19	15–20	+	+	AF	+	R+L
2	13	F	38	13	32	13–16	15–18	+	+	SR	–	R+L
3	6	M	70	10	19	—	—	+	+	SR	–	R+L
4	32	F	37	10	24	—	—	+	+	SR	–	R+L
5	21	F	37	8	50	12–17	6–10	–	–	SR	–	R
6	56	F	45	9	45	15–20	0–3	–	+	SR	–	R
7	23	M	46	7	36	18–21	11–12	–	–	SR	–	R
8	50	F	59	7	30	—	—	+	+	SR	–	R+L
9	10	M	50	12	55	16–22	9–12	–	+	SR	–	R
10	19	F	40	6	37	10–15	6–10	–	+	SR	–	R
11	25	F	62	14	22	17–19	20–24	+	+	SR	–	R+L
12	21	M	34	6	42	16–18	0–6	–	–	SR	–	R
13	64	F	50	10	30	—	—	+	+	SR	–	R+L
14	21	M	47	8	15	—	—	+	–	SR	–	R+L
15	45	M	38	10	50	20–23	6–7	–	+	AF	–	R
16	22	M	60	7	20	21–22	23–27	+	+	SR	–	R+L
17	15	M	54	12	40	18–22	10–12	–	+	SR	–	R
18	13	F	30	9	38	12–14	9–12	–	–	SR	–	R
19	45	M	56	6	21	17–25	16–20	+	+	SR	+	R+L
20	70	F	35	9	27	—	—	+	–	AF	–	R+L
21	16	M	58	11	19	21–24	18–22	+	+	SR	–	R+L

RVEDD, right ventricular end-diastolic dimension; EDARVWT, end-diastolic anterior right ventricular wall thickness; LVEDD, left ventricular end-diastolic dimension; RVEDP, right ventricular end-diastolic pressure; LVEDP, left ventricular end-diastolic pressure; MR, mitral regurgitation; TR, tricuspid regurgitation; AF, atrial fibrillation; SR, sinus rhythm; EDM, early diastolic murmur.

RVEDD: normal range 7 to 23 mm. Right ventricular endomyocardial fibrosis mean 47 mm, SD 11 mm.

LVEDD: normal range 35 to 56 mm. Left ventricular endomyocardial fibrosis mean 24 mm, SD 6 mm.

EDARVWT: normal <5 mm. Right ventricular endomyocardial fibrosis mean 9 mm, SD 2 mm.

Fig. 1 *Echocardiogram in right ventricular endomyocardial fibrosis showing (i) increased right ventricular dimension, and (ii) paradoxical septal motion. RV, right ventricle; IVS, interventricular septum; LV, left ventricle.*

Fig. 2 *M-mode scan in right sided endomyocardial fibrosis showing increased dimension of the outflow tract. IVS, interventricular septum; Ao R, aortic root; LA, left atrium.*

Fig. 3 Echocardiogram in right ventricular endomyocardial fibrosis showing thickened and strongly contracting anterior right ventricular wall.

Fig. 4 Tricuspid valve echocardiogram showing fine fluttering of the anterior tricuspid valve leaflet. Electrocardiogram shows sinus rhythm.

Fig. 5 Echocardiogram in biventricular endomyocardial fibrosis. The features of right ventricular endomyocardial fibrosis are (i) dilated right ventricle (RV), (ii) pericardial effusion (PE), and (iii) paradoxical septal motion. The reduced left ventricular dimension of left sided involvement is shown.

Fig. 6 Fine fluttering of anterior mitral valve leaflet (AML). (See text.)

Fig. 7 Right ventricular end-diastolic pressure (RVEDP) and echocardiographic right ventricular end-diastolic dimension (RVEDD) relation in 15 patients with right ventricular endomyocardial fibrosis. The end-diastolic pressure and echocardiographic dimension were raised in all 15. There was a significant degree of correlation between the RVEDP and RVEDD ($r=0.683$ $p<0.005$).

Fig. 8 Left ventricular end-diastolic pressure (LVEDP) and echocardiographic left ventricular end-diastolic dimension (LVEDD) relation in 15 patients with endomyocardial fibrosis (nine with right ventricular and six with left ventricular involvement). The LVEDP was raised, and LVEDD reduced in patients with left ventricular endomyocardial fibrosis; the LVEDP and LVEDD were normal in right ventricular endomyocardial fibrosis. There was a highly significant correlation between LVEDD and LVEDP ($r=0.82$ $p<0.005$).

wave (the patients were in sinus rhythm), and systolic notching of the pulmonary valve echocardiogram.

The echocardiograms in the two patients with constrictive pericarditis, and in two patients with gross ascites caused by portal hypertension were normal. One other patient showed echocardiographic features of tricuspid stenosis, namely, reduced e-f slope of the anterior tricuspid leaflet.

The 15 patients studied haemodynamically and angiographically showed the characteristic dip and plateau right ventricular pressure pattern with raised right atrial and right ventricular end-diastolic pressures. The six patients with left ventricular endomyocardial fibrosis had raised left ventricular end-diastolic pressures (Table). The right atrial angiograms showed a grossly dilated right atrial chamber with stasis of the contrast medium in the right atrium. The inflow tract of the right ventricle was obliterated and/or irregular, with distension of the outflow tract. The left ventricular angiogram showed changes consistent with left ventricular endomyocardial fibrosis in six patients (viz small, irregular chamber). There was also mitral regurgitation. Fig. 7 shows the relation of the right ventricular end-diastolic pressures to the echocardiographic right ventricular end-diastolic dimensions. Both were increased in all. The relation of the left ventricular end-diastolic pressures to the echocardiographic left ventricular end-diastolic dimension is shown in Fig. 8. The left ventricular end-diastolic pressures and left ventricular end-diastolic dimensions were normal in the nine patients with lone right ventricular endomyocardial fibrosis. The left ventricular end-diastolic pressures were raised and left ventricular end-diastolic dimensions diminished in patients with left ventricular involvement.

Discussion

Endomyocardial fibrosis, first described from Uganda in 1948 by Davies,¹⁵ is characterised by fibrosis of the endocardium and the subendocardial layers of either the left or right ventricle, or both. The main feature^{11 12} is deep scarring of the endocardium usually starting at the apex and spreading to the inflow tract, eventually affecting the papillary muscles, chordae, and posterior cusps of the atrioventricular valves, leading to mitral or tricuspid regurgitation. Macroscopically, the heart shows distortion and partial obliteration of the ventricles. There is normally moderate hypertrophy, and some hearts are greatly dilated, the dilatation mostly being the result of an enlarged right and/or left atrium. In right sided endomyocardial fibrosis, though there is apical obliteration and distortion of the cavity, the right ventricular outflow tract is dilated. There may be hydropericardium. Angio-

graphic studies^{9 10} have shown vigorous pulsation of the infundibulum of the right ventricle, tricuspid regurgitation, and a grossly dilated right atrium.

The echocardiographic features of increased right ventricular dimension and paradoxical septal motion in our patients with right sided endomyocardial fibrosis are not specific for endomyocardial fibrosis, and may in part result from the associated tricuspid regurgitation and/or pulmonary hypertension. This does not, however, explain these findings in those patients without tricuspid regurgitation or pulmonary hypertension. The paradoxical echocardiographic finding of increased right ventricular dimension in a disease that is characterised by cavity obliteration is difficult to explain. A possible explanation may be that, as a result of distortion of the heart and its axis, the ultrasound beam may be partly traversing the dilated right atrium as well as the right ventricle. The dilated right ventricular outflow tract and the vigorous anterior right ventricular wall contractions are in keeping with the pathological and angiographic findings in this disease, as is the reduced left ventricular dimension in left sided endomyocardial fibrosis. Apical obliteration of the cavity in right ventricular endomyocardial fibrosis, and strong echoes from the right ventricular endocardium have been reported by Hess *et al.*¹⁰ and Hernandez-Pieretti,¹³ respectively.

The aortic and pulmonary valves are not affected in endomyocardial fibrosis. Early diastolic murmurs have, however, been reported in a few cases of haemodynamically and angiographically proven endomyocardial fibrosis.^{5 10} In the series reported by Davies and Ball,¹⁶ in five cases the aortic valve had subacute bacterial endocarditis. In the two patients in our series with biventricular endomyocardial fibrosis in whom an early diastolic murmur was heard, aortic root angiograms were normal. Fine fluttering of the anterior mitral valve and tricuspid valve echogram was observed in these two patients.

Fine fluttering on an atrioventricular valve in the absence of aortic regurgitation, atrial fibrillation, or a high flow, is likely to be the result of distortion of the ventricular inflow tract and/or the valve apparatus by fibrosis. This mechanism may underlie the early diastolic murmur present in our two cases.

The echocardiographic features of left ventricular involvement are worth noting. These include a small left ventricular dimension, an increase in left atrial dimension, and the rather interesting inverse relation between the left ventricular end-diastolic pressure and end-diastolic dimension. The last is not seen in other types of left ventricular disease. Though the echocardiographic features found in our patients are not specific for endomyocardial fibrosis, they nevertheless are useful in resolving, non-invasively, any problems in diagnosis in these circumstances. Patients with

constrictive pericarditis have similar clinical and haemodynamic features with right ventricular endomyocardial fibrosis. The echocardiographic features observed in our patients help to distinguish between right ventricular endomyocardial fibrosis and constrictive pericarditis.

Endomyocardial fibrosis was diagnosed in all clinically, and confirmed haemodynamically and angiographically in 15 and at necropsy in one. An attempt is made to correlate the echocardiographic findings with the haemodynamic findings, and to relate the echocardiographic findings to the angiographic findings. Though the echocardiographic features are not specific for endomyocardial fibrosis, they are characteristic, and useful in distinguishing non-invasively between endomyocardial fibrosis and other diseases which present with similar clinical features. In such circumstances, it is suggested that these echo features may be considered diagnostic of endomyocardial fibrosis, especially since these other conditions, for example rheumatic valvular disease and congestive cardiomyopathy, have characteristic echo features.

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