Persistent left ventricular disease in clinically "cured" primary endocardial fibroelastosis

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SUMMARY We studied by serial cardiac catheterisation eight patients with the dilated form of primary endocardial fibroelastosis in whom congestive heart failure disappeared with treatment. All remained without symptoms for at least three years before recatheterisation. Four patients showed regression of the abnormal electrocardiographic findings, three showed persistence, and one showed progression of electrocardiographic left ventricular overload pattern.

On first cardiac catheterisation all patients had a dilated left ventricle with a mean ejection fraction of 0.36. In six of the patients repeat cardiac catheterisation showed left ventricular dilatation with a diminished ejection fraction (mean 0.32). Left ventricular end-diastole pressure was raised (12 to 28 mmHg, mean 19 mmHg). In this group were included the three patients with persistence and one with progression of the abnormal electrocardiographic findings, and two of the four patients with regression of these findings. The highest left ventricular end-diastolic pressure was found in a patient in whom the abnormal electrocardiographic findings almost reverted to normal. In the two remaining patients with reversion of the electrocardiographic abnormalities repeat cardiac catheterisation showed nothing abnormal.

Our findings indicate that "cure" in primary endocardial fibroelastosis is incomplete. These findings may be the cause of sudden death or late clinical deterioration in some reported patients with "cured" primary endocardial fibroelastosis. The electrocardiogram is of little value in assessing these processes.

The mortality rate of the dilated form of primary endocardial fibroelastosis has significantly decreased during the past 20 years. Patients in whom congestive heart failure disappeared with treatment and who survive to the age of 5 are considered cured. In addition, if the abnormal electrocardiographic findings regress, there is no clinical remnant of the disease.¹ It is the purpose of this paper to report serial cardiac catheterisation in eight patients with the dilated form of endocardial fibroelastosis, all of them with distinct clinical improvement.

Subjects and methods

Eight patients were studied. In all of them clinical and angiocardigraphic diagnosis of primary endocardial fibroelastosis was made before the age of 1 year. All presented with congestive heart failure in the first year

of life. Clinical history, physical examination, electrocardiograms, thoracic x-rays, and data of two serial cardiac catheterisations were available in all cases. The first cardiac catheterisation included right and left heart studies in six patients and left heart studies only in two patients.

Diagnosis of the dilated form of primary endocardial fibroelastosis was based on the presence of all the following features: (1) congestive heart failure in the first year of life; (2) electrocardiographic signs of left hypertrophy with inversion or flattening of T waves in leads V5 to 6 (Fig. 1 to 2); (3) a dilated poorly contracting left ventricle on angiocardigraphy (ejection fraction less than 0.4) (Fig. 2); (4) absence of shunts (by complete oxygen measurements); (5) absence of pressure gradients across the aortic and pulmonary valves.

The eight patients were studied in the last 15 years. Most of them were catheterised before cross-sectional echocardiography became available to us, so that only three of them were studied in this manner.

Fig 1 (A) Electrocardiogram before first cardiac catheterisation, showing deep S waves in the right praecordial leads and tall R waves in the left praecordial leads. (B) Electrocardiogram six years later, showing a distinct decrease in the voltage of the S waves in the right praecordial leads and R waves in the left praecordial leads.

Results

Five of the patients were female and three were male. In all congestive heart failure disappeared after treatment with digoxin and diuretics. Seven patients remained without symptoms until the second cardiac catheterisation at the age of 5 to 11 years. One patient remained symptomless for six years, but developed dyspnoea and fatigue at the age of 7 years, before repeat cardiac catheterisation.

All patients, when first studied, showed electrocardiographic signs of left ventricular hypertrophy. In four of them these abnormalities improved and at the time of repeat catheterisation the electrocardiogram was almost normal. In three patients these abnormal findings persisted and in one patient they had progressed at the time of the second cardiac catheterisation

(Fig. 1 to 3). At the first cardiac catheterisation all patients were receiving digoxin. Two of them, though improving clinically, still had congestive heart failure at the time of catheterisation and in six of them signs of congestive heart failure disappeared with treatment. The mean left ventricular ejection fraction in the eight patients was 0.36. The left ventricular end-diastolic pressure ranged between 9 and 20 (mean 13) mmHg.

No patients were receiving digoxin at the time of second catheterisation. Repeat cardiac catheterisation in six patients showed abnormalities, including left ventricular dilatation (Fig. 4 and 5), decreased systolic contraction (mean ejection fraction 0.32), and raised left ventricular end-diastolic pressure (12 to 28 mmHg, mean 19 mmHg).

In this group were three patients with persistently

Fig. 2 (A) Electrocardiogram before cardiac catheterisation, showing left ventricular hypertrophy and inverted T waves in the left praecordial leads. (B) Electrocardiogram five years later, showing regression of left ventricular hypertrophy and positive T waves in the left praecordial leads.

Fig. 3 Serial electrocardiograms (lead V6) showing progression of left ventricular hypertrophy pattern despite complete clinical recovery.

abnormal electrocardiographic findings; the abnormalities in one patient progressed and the electrocardiogram reverted to normal in the other two. In two of these patients both angiocardiograms showed mitral regurgitation which was not found in one of them at the first study. The highest left ventricular end-diastolic pressure (28 mmHg) was found in one patient who had symptoms at the time of second car-

diac catheterisation. The electrocardiogram of this patient showed pronounced regression of the abnormalities at this time (Fig. 2). Cross-sectional echocardiography in two patients with abnormal electrocardiograms showed left ventricular dilatation in both patients and concentric thickening in one of them.

In the remaining two patients repeat cardiac catheterisation showed left ventricular end-diastolic pressures of 5 and 8 mmHg, respectively, and ejection fractions of 0.52 and 0.58, respectively. The left ventricular wall was not thickened. Cross-sectional echocardiography in one of these patients showed nothing abnormal.

Discussion

There is some disagreement concerning the possibility of the clinical diagnosis of primary endocardial fibroelastosis. Many investigators,¹⁻⁷ however, claim to be able to make this diagnosis accurately. Even if the exact diagnosis may be controversial it is agreed that the syndrome of congestive heart failure in infancy with electrocardiographic signs of left ventricular hypertrophy with inversion or flattening of T waves in leads V5 to 6, and without clinical signs of a shunt or obstructive cardiac anomaly, forms a distinct entity, which may be termed primary endocardial fibroelastosis. Cardiac catheterisation improves the diagnosis but cannot exclude the rare possibility of primary congestive cardiomyopathy. Despite this fact it is agreed that primary endocardial fibroelastosis may be diagnosed by clinical findings and cardiac catheterisation. The commonest form of primary endocardial fibroelastosis is the dilated form.¹

The reported mortality has decreased significantly in the past 20 years, for several reasons.^{1,7} Disappearance of congestive heart failure with treatment and

(A)

(B)

Fig. 4 Left ventriculography of a patient who recovered completely clinically and has been without symptoms for five and a half years. (A) diastole, (B) systole showing a dilated left ventricle with poor contraction.

Fig. 5 Left ventriculography of a patient who developed dyspnoea after nine years without symptoms. (A) Diastole, (B) systole, showing a dilated left ventricle with poor contraction.

lack of symptoms after the age of 5 years are considered as indicating a cure. In about 75% of these long term survivors the electrocardiogram is normal after five years, thus leaving no apparent sequelae of the disease. These data collected mainly in the last decade evidenced optimism for the long term prognosis of primary endocardial fibroelastosis.¹ This optimism was obscured, however, by several reports of late clinical deterioration or sudden death in patients who were considered to be cured by the above criteria.¹⁷

Our findings indicate that cure is incomplete, as abnormal features were found at cardiac catheterisation in 75% of the cases. The decreased ejection fraction and the raised left ventricular end-diastolic pressure may result from persistent disease (incomplete cure) or from some secondary process. It is not clear whether the thickening of the ventricular wall results from endocardial proliferation and fibrosis only or also from myocardial hypertrophy. It is possible that myocardial hypertrophy is secondary to the primary disease and develops as a compensatory mechanism, preventing further decrease in left ventricular function. These abnormal findings may be responsible for the late complications reported in primary endocardial fibroelastosis. This hypothesis is supported by recurrence of symptoms in one of our patients.

Our data also indicate that the electrocardiogram has little significance in evaluating long term prognosis. Abnormal haemodynamic findings were present in all patients with a persistently abnormal electrocardiogram and in 50% of the patients with a normal electrocardiogram. The number of patients, however, is too small to draw conclusions.

Danilowicz⁸ performed repeat cardiac catheterisations in four survivors of primary endocardial fibroelastosis at less than 5 years of age. In three of them the studies were normal, and in one (25%) there

was impaired left ventricular function. The difference between these data and ours may be because our patients were older.

Most information obtained by repeat cardiac catheterisation and angiocardiology may be obtained from cross-sectional echocardiograms. Moreover, left ventricular wall thickening can be assessed better by echocardiography than by angiocardiology. We believe that in the near future, cross-sectional echocardiography will be the method of choice for the evaluation and long-term follow-up of endocardial fibroelastosis.

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