Endocardial Fibroelastosis

Angiocardiographic Studies

By Leonard M. Linde, M.D., Forrest H. Adams, M.D., and Bernard J. O’Loughlin, M.D.

Endocardial fibroelastosis is an important cause of congestive heart failure and death in infancy and early childhood. Four cases of this entity are described, with particular attention to the unique angiocardiographic features. This graphic portrayal of the functional derangement may be of great help in the clinical diagnosis of fibroelastosis.

Endocardial fibroelastosis as a cause of severe cardiac hypertrophy in infants is being recognized with increasing frequency. The diagnosis can be suspected clinically, but final confirmation must be made at necropsy. This report describes an angiocardiographic sign that may be helpful in the antemortem diagnosis of this entity and that may help us to understand better its pathologic physiology.

Method

Four children were studied in whom the chronic form of primary endocardial fibroelastosis was diagnosed clinically. In one of these patients the diagnosis was confirmed at autopsy. In addition to the routine clinical and laboratory procedures, cardiac catheterization and selective angiocardiography were performed on the 4 patients. Biplane angiocardiograms were taken at 6 or 12 frames per second after automatic injection at 5 Kg./cm.² of 50 per cent sodium diprotrizoate (Miokon*) into the main pulmonary artery. Mechanical systole and diastole were determined by inspection and confirmed by reference to simultaneously recorded electrocardiograms.

Results

The angiocardiograms in these patients with endocardial fibroelastosis uniformly showed greatly diminished left ventricular contractility. In 3 patients there was no change in systolic and diastolic volume in consecutive cardiac cycles. Angiocardiography in the fourth patient, performed prior to the development of cardiovascular symptoms, showed slight cyclic changes in the cardiac contour. All studies demonstrated prolonged retention of dye in an enlarged left ventricle.

Case Report

Case 1. C. L. B., a white girl, developed dyspnea, listlessness, vomiting, and pallor at 3½ months of age. (A male sibling had died at age 3½ months with identical symptoms, but no diagnostic or postmortem studies had been performed.) Examination revealed rapid grunting respiration and faint heart tones without a murmur. Congestive heart failure was diagnosed and responded to therapy. The electrocardiogram showed marked left ventricular hypertrophy. A roentgenogram demonstrated gross cardiomegaly involving the left ventricle and left atrium. On cardiac catheterization there were normal right heart pressures, oxygen saturations consistent with decreased cardiac output, and no evidence of left-to-right shunt (table 1). The angiocardiogram showed a large left ventricular cavity with an unchanging contour during systole and diastole and prolonged retention of dye (fig. 1).

In spite of radiation therapy to the heart and general supportive measures, the patient’s condition progressively deteriorated, with fatal heart failure occurring at age 2 years. At postmortem examination a 2-mm. thick, yellow-white endocardium coated the left atrium, left ventricle, and mitral valve. There was biventricular hypertrophy and dilatation with congestion related to left and right heart failure. No other cardiac anomalies were found.

Case 2. D. V. W., a 4½-month-old white boy, previously reported, suddenly developed rapid respirations, irritability, pallor, anorexia, and vomiting. Examination revealed a pale infant in shock, with gross cardiomegaly and marked hepatomegaly. Heart tones were muffled and no murmurs were audible. He improved after appropriate treatment of the shock and congestive heart failure. Electrocardiograms showed left ventricular hypertrophy with inverted T waves over the left precordium. A roentgenogram revealed marked cardiac enlargement, mainly left ventricular, with some left atrial enlargement. The findings on cardiac catheterization were within normal limits (table 1). The

From the Departments of Pediatrics and Radiology, School of Medicine, University of California, Los Angeles, Calif.

* Manufactured by Mallinckrodt Chemical Works, St. Louis, Mo.
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angiocardio gram showed loss of left ventricular contractility with diminished expulsion of dye (fig. 2).

When last seen at 13 months of age, pallor, rapid respirations and cardiomegaly persisted, but the

Table 1.—Cardiac Catheterization in Four Patients with Primary Endocardial Fibroelastosis

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Pressure mm Hg</th>
<th>Oxygen saturation %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Right atrium</td>
<td>Right ventricle</td>
</tr>
<tr>
<td>1</td>
<td>3/1</td>
<td>21/3</td>
</tr>
<tr>
<td>2</td>
<td>7/3</td>
<td>27/0</td>
</tr>
<tr>
<td>3</td>
<td>7/4</td>
<td>26/4</td>
</tr>
<tr>
<td>4</td>
<td>8/1</td>
<td>43/1</td>
</tr>
</tbody>
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Case 3. C. B., a 2-year-old white girl, had an enlarged heart at age 19 months on a roentgenogram that was taken during an episode of pneumonia. For 2 months prior to entry her private physician heard a cardiac murmur intermittently. Examination revealed cardiac enlargement with heart tones of fair quality. A short soft midsystolic murmur in the left second interspace and a higher pitched localized apical systolic murmur were barely audible. The electrocardiogram was consistent with left ventricular hypertrophy. On chest roentgenogram there was moderately severe cardiomegaly involving the left ventricle and left atrium. Cardiac catheterization revealed normal right heart pressures, low oxygen saturations consistent with low cardiac output, and no evidence of left-to-right shunt (table 1). The patient's condition was stable with maintenance digitalis therapy.

Fig. 1. Case 1. Angiocardiogram with dye in the left heart shows no measurable change in ventricular volume in systole (right) and diastole (left). Atrial change is less than normal.

Fig. 2. Case 2. Angiocardiogram demonstrating unchanging left ventricular volume and poor aortic filling.
angiocardiogram showed a poorly emptying, enlarged left ventricle with an unchanging contour (fig. 3).

**Case 4.** M. G., a 6-month-old white boy, had cardiac enlargement on a roentgenogram taken because of "repeated colds, chest congestion, and rapid breathing." Examination revealed a rapid respiratory rate. Cardiomegaly was associated with a loud high-pitched apical systolic murmur, transmitted to the left axilla. Electrocardiographic findings consisted of left ventricular hypertrophy with inverted T waves over the left precordium. An enlarged left ventricle was the main cause of the cardiomegaly seen on the chest film. Findings on cardiac catheterization were elevated right ventricular pressure and low oxygen saturations consistent with decreased cardiac output. There was no evidence of a left-to-right shunt (table 1). The angiocardiogram demonstrated reduced but measurable systolic and diastolic variation in left ventricular volume (fig. 4).

**DISCUSSION**

The clinical aspects of endocardial fibroelastosis have been covered in an excellent extensive review of the subject by Dennis and coworkers. Suspected etiologic agents have been numerous but the final result is a thickened endocardium that limits expansion and contraction of the ventricle. This splinting action leads to heart failure in most patients.

Others have described the radiologic picture to consist of great cardiomegaly, predominantly of the left ventricle. On angiocardiography left ventricular enlargement and delayed systolic expulsion have been described.

In our experience reciprocal atrial and ventricular filling is easily seen in normal patients (fig. 5) and in patients with cardiac conditions other than endocardial fibroelastosis (figs. 5 and 6). Atrial systole accompanies ventricular diastole, while atrial filling occurs simultaneously with ventricular emptying. Figure 5 shows the respective chamber emptying to be fairly complete in the normal, with marked

**FIG. 3.** Case 3. Lateral and anteroposterior angiocardiographic views depicting absent left ventricular contractility and reduced left atrial contractility.
change in systolic and diastolic volume during the cardiac cycle.

This is in sharp contrast to what is seen in patients with endocardial fibroelastosis (figs. 1–4). In such patients, no systolic and diastolic change in volume occurs. Dye enters an enlarged left ventricular cavity and remains there, as muscular contraction is apparently impaired by a splinting action of the thickened endocardium. In case 4, studied when he had few cardiovascular symptoms, minimal systolic and diastolic change occurred. Perhaps this instance represented an early stage in the development of the typical angiocardiographic picture.

Angiocardiographic studies were also performed in patients with left ventricular enlargement and left ventricular failure without endocardial fibroelastosis. Figure 6 shows marked systolic and diastolic change in volume on the angiocardiogram of a patient with a ventricular septal defect, left ventricular hypertrophy, and left ventricular failure.

Before the specificity of this interesting angiocardiographic sign can be fully evaluated, studies must be performed on the other rarer types of heart disease formerly classified as "idiopathic cardiac hypertrophy." Glycogen storage disease of the heart, aberrant left coronary artery disease, and idiopathic myocarditis can all clinically resemble endocardial fibroelastosis, but at the present time we have not studied patients with these other diseases angiographically. It is hoped, however, that patients with an aberrant left coronary artery will be recognized by the technic of injecting the radiopaque material selectively into the main pulmonary artery. If such a vessel is present, it should be well seen in the lateral view originating from the main pulmonary artery.
Summary

Selective biplane angiocardiography was performed in 4 patients with the chronic form of primary endocardial fibroelastosis. The diagnosis was made clinically in all and confirmed at autopsy in 1. A striking finding was the unchanging size and contour of the left ventricle. Evidence was presented to indicate that this was not due to heart failure or left ventricular hypertrophy. These observations may help in understanding the pathologic physiology in endocardial fibroelastosis and may be of aid in the antemortem diagnosis of this entity.

Summario in Interlingua

Selective angiocardiographia biplan esseva executate in 4 patientes con le forma chronic de primari fibroelastosis endocardial. Le diagnose esseva facite clinicamente in omne casos e confirmate al necropsia in 1. Un constatation frappante esseva le nonalterate dimension e contorno del ventriculo sinistre. Es documentate le these que isto non resultava de disfallimento cardiac o hypertrophia sinistroventricular. Iste observationes va possibilemente promover le comprension del pathophysiologia de fibroelastosis endocardial e
assister in establir le diagnose de iste entitate ante le morte.

REFERENCES


9 Carter, B.: Personal communication, as quoted by Greaves et al. (ref. 8).


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LEONARD M. LINDE, FORREST H. ADAMS and BERNARD J. O'LOUGHLIN

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