Infarction Patterns in Endocardial Fibroelastosis

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Primary endocardial fibroelastosis may be difficult to differentiate from the syndrome of anomalous origin of the left coronary artery, arising from the pulmonary artery. There is a similar clinical course of progressive cardiac disability and each of the two conditions may present an electrocardiographic pattern of left ventricular hypertrophy with inversion of the T waves over the left precordium. Noren and associates recently stated that the vectorcardiogram helped differentiate the two entities. They showed that patients with an anomalous origin of the left coronary artery exhibited a posteriorly oriented clockwise loop in the horizontal plane of the vectorcardiogram. This pattern of an anterior myocardial infarct was never encountered in patients with endocardial fibroelastosis in their experience.

It is the purpose of this report to present three cases of endocardial fibroelastosis in which the vectorcardiogram displayed such a loop.

The paper is of further interest in that two of these three patients were identical twins who were under observation since birth and were thought to have no significant heart disease, prior to the clinical onset of their ailment, at the age of 2½ years. The electrocardiographic progression from normal to abnormal findings during the development and progression of endocardial fibroelastosis has never been shown before to the best of our knowledge.

Report of Cases

Case 1

M.G., a 3 8/12-year-old Indian girl, born after a normal pregnancy, had remained in good health until the age of 2 years and 11 months, when she developed congestive heart failure, which improved after digitalization.

At 3 8/12 years the patient was hospitalized with congestive heart failure. Physical examination revealed normal blood pressures in both upper and lower extremities, pulse of 136, and respirations of 36 per minute. A grade II/VI blowing pansystolic murmur was audible at the apex. No diastolic murmurs were heard. There were decreased breath sounds posteriorly at the left base, but no rales. The liver was palpated 5 cm. below the right costal margin. No edema was evident.

The hematocrit value was 41 per cent. A mumps skin test was negative. Chest films revealed marked cardiomegaly with atelectasis of the left lower lobe. The electrocardiogram showed left ventricular hypertrophy and borderline p–mitrale. The R/S progression in the precordial leads, from a QS in V₁ through a minute R in V₄ and V₅, was suggestive of an anterior myocardial infarction (fig. 1). The vectorcardiograms demonstrated a counterclockwise loop in the frontal and sagittal planes (fig. 1). In the horizontal plane the initial forces were predominantly posterior and the loop was in a clockwise direction, reversing terminally. There was considerable terminal slowing. The vectorcardiographic pattern was thought to be indistinguishable from anterior myocardial infarction.

Right heart catheterization revealed no shunts but moderately elevated pressures in the pulmonary artery (60/40 mm. Hg with a mean of 50), and pulmonary "capillary" position (27 mm. Hg). Aortography demonstrated normal coronary arteries.

The patient developed increasingly severe congestive heart failure and died 1 week later. Postmortem examination revealed a very large heart. There were hypertrophy and dilatation of both left and right ventricles. The coronary arteries were normal. A diffuse opacification of the
left ventricular endocardium was noted. Microscopic examination revealed marked thickening of the endocardium of the left ventricle, with an increase in elastic tissue. No evidence of fibrosis or acute inflammation was present in the myocardium. Gross and microscopic changes in other organs were compatible with congestive heart failure.

Case 2
This patient, an identical twin to case 3, was the second born to a healthy Negro mother. The pregnancy was uncomplicated except for a 20-Kg. weight gain and an upper respiratory infection in the last month of pregnancy. The baby's course was benign after a rapid recovery from initial respiratory distress. At 1 month of age, a grade-II/VI blowing, pansystolic murmur was heard for the first time at the lower left sternal border. Blood pressures were normal in both upper and lower extremities. Chest films with barium swallow and electrocardiogram (fig. 2), done at the age of 2 months, were interpreted as probably within

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**Figure 1**

*Case 1. Electrocardiogram and vectorcardiogram in a 3 8/12-year-old girl with endocardial fibroelastosis. The P waves show the notching of p-mitrale but are only 0.08 second in duration. The major forces are inferior and posterior. The R in V_6 exceeds the limits of normal. The precordial R/S progression is abnormal: there is a QS pattern in V_2, and failure of a progressive increase in the amplitude of the R wave from V_2 through V_5. A dominant R wave is present only in V_6. The vectorcardiogram shows a very minimal anterior vector, which quickly swings markedly posteriorly, in a clockwise loop in the horizontal plane. The loop reverses itself to terminate in a counterclockwise inscription, with considerable terminal slowing. (The amplification for this and all other vectorcardiograms in this series is the same. The dots are comet-shaped, with the tail behind.)*
Figure 2

Case 2. Electrocardiogram at 2 months of age, interpreted as within normal limits. (The ST-segment elevation in V₆ was not consistently present.)

Figure 3

Case 2. Electrocardiogram and vectorcardiogram at 2½ years of age. The scalar electrocardiogram shows left ventricular hypertrophy on the basis of the R/S ratio in V₁ and T-wave changes. The frontal loop of the vectorcardiogram is partly counterclockwise; the horizontal loop is markedly leftward and normally counterclockwise. The T loop is rightward and anterior, suggesting "strain."
normal limits. The clinical impression was a small ventricular septal defect.

The child, at 4 months of age, had what was described by the mother as "mumps." This apparently consisted of bilateral swelling in the parotid areas. The child's twin and another sibling had a similar disease.

At 6 months of age the physical examination was unchanged. The electrocardiogram and x-rays were again interpreted as being within normal limits.

The child was seen again at the age of 2 1/2 years. She was in the fortieth percentile for weight and the fiftieth percentile for height. A grade-II/VI blowing pansystolic murmur was heard over the entire precordium with maximum transmission at the base and the upper left sternal border. The electrocardiogram revealed left ventricular hypertrophy with "strain" (fig. 3), and x-rays revealed cardiomegaly. Vectorcardiogram showed normal total voltage, but marked leftward orientation and T-vector deviation to the right and anterior suggesting left ventricular hypertrophy with "strain" (fig. 3).

One month later the child entered the Emergency Room because of progressive edema, rapid respirations, dyspnea, and anorexia of 1 week's duration. Marked peripheral and periorbital edema and an enlarged liver were noted. A gallop rhythm was audible at the apex. X-rays showed cardiomegaly in addition to pulmonary vascular congestion. The electrocardiogram revealed left ventricular hypertrophy and T-wave changes (fig. 4). The vectorcardiographic pat-

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**Figure 4**

Case 2. Electrocardiogram and vectorcardiogram at 2 7/12 years. The electrocardiogram reveals a relatively normal R/S progression in the precordial leads, with extremely deep S in V1, and tall R in V5, plus ST- and T-wave changes. The vectorcardiogram has changed markedly in the horizontal and sagittal planes, which are now inscribed in a predominantly clockwise direction in spite of the marked posterior orientation of forces.
terns in the horizontal and sagittal planes had changed considerably and suggested anterior myocardial infarction (fig. 4). The patient was digitalized and given diuretics with gradual improvement.

Three months later she entered the hospital with more severe congestive heart failure and pneumonitis, and died in 12 hours.

Postmortem examination revealed evidence of chronic congestive heart failure and pneumonitis. The coronary arteries were normal. The left ventricle had diffuse endocardial thickening, with an increase in elastic tissues. No ventricular septal defect was present.

Case 3

The patient was the first-born twin of case 2. She recovered rapidly from mild respiratory distress in the neonatal period.

A grade-I/VI blowing systolic murmur was first noted at the lower left sternal border at 1 month of age. X-rays at 2 months of age were interpreted as being normal. At 6 months of age the electrocardiogram and x-rays were within normal limits. She had had a disease clinically compatible with "mumps" at the age of 4 months.

The patient was seen again at the age of 2 1/2 years with congestive heart failure. The child had been well until 2 weeks before, when she developed an upper respiratory illness and a cough. Examination revealed an acutely ill child with rapid respiratory rate and pretilial and periorbital edema. A gallop rhythm was present, and a grade-II/VI short, blowing systolic murmur was heard at the upper left sternal border. Auscultation revealed rales at both lung bases.

The electrocardiogram revealed low QRS voltages and S-T shift and T-wave flattening suggestive of myocardial disease. X-rays revealed moderate cardiomegaly and consolidation of the right middle lobe. Digitalis, diuretics, and antibiotics were administered with gradual improvement.

Two months later she developed more severe congestive heart failure. The electrocardiogram revealed left ventricular hypertrophy with S-T and T-wave changes (fig. 5). The vectorcardiogram demonstrated a clockwise loop in the horizontal plane, but with most of the loop posteriorly oriented (fig. 5). X-rays showed increasing cardiomegaly. Because of the possibility of anomalous coronary artery, aortography was proposed but was declined by the mother.

The patient remained in chronic congestive failure despite diuretics and continued digitalis. At the age of 3 years she died suddenly. Postmortem examination was denied.

Discussion

Electrocardiographic or vectorcardiographic patterns of anterior myocardial infarcts are rarely found in early childhood. According to several authorities,3-6 this pattern alone makes the diagnosis of an anomalous left coronary artery quite likely. There are, however, other conditions in which similar electrocardiographic features have been found; specifically in myocarditis,1,5,7-9 as an unusual manifestation of digitalis effect1,10 and in some very complex heart lesions.11,12 Auld and Watson13 in 1957, presented the electrocardiographic findings of an adolescent boy who had died from endocardial fibroelastosis. His electrocardiogram showed a pattern diagnostic of an extensive transmural infarct. The coronary arteries were normal but there was a large mural thrombus present over the anterior wall of the left ventricle. More recently, Linde and Adams14 found two other children with endocardial fibroelastosis whose scalar electrocardiogram was suggestive of an anterior myocardial infarct. We are, however, not aware of any vectorcardiographic studies in primary endocardial fibroelastosis. Noren et al.,2 quite recently presented a series of patients with anomalous origin of the left coronary artery. All of their patients had a posteriorly oriented clockwise loop on the horizontal plane of the vectorcardiogram. This feature was reported to be very reliable in differentiating anomalous origin of the left coronary artery from primary endocardial fibroelastosis. The latter disease was in their experience2 never associated with a posteriorly oriented clockwise loop. Hugenholtz et al.15 stated that in a pattern of an anterior wall infarct, the initial forces on the horizontal plane of the vector had to be directed posteriorly. It is of interest that in three of the patients of Noren et al.,2 the initial forces were directed anteriorly and counterclockwise. This was also found in two of our three patients with endocardial fibroelastosis.

Why patients with endocardial fibroelastosis should have electrical findings suggestive of an anterior myocardial infarct is uncertain. Thomas and associates,16 in their study of autopsy cases of endocardial fibroelastosis, showed that mural thrombosis was a promi...
Figure 5

Case 3. Electrocardiogram and vectorcardiogram at 2½ years. Findings are practically identical to those in figure 4 (case 2).

The electrocardiographic patterns are by themselves not sufficient to differentiate between endocardial fibroelastosis and anomalous origin of the left coronary artery.

Summary

Three cases of endocardial fibroelastosis with vectorcardiographic findings suggestive of an anterior myocardial infarct are presented. It had been previously reported that such vectorcardiographic features were diagnostic of anomalous origin of the left coronary artery. We conclude that the two diseases cannot be differentiated on the basis of electrocardiography or vectorcardiography alone.
Two of the cases presented were in identical twins; serial scalar electrocardiograms and vectorcardiograms were obtained during the development and progression of endocardial fibroelastosis.

References

Ego Protection

Nothing has tended more to retard the advancement of science than the disposition in vulgar minds to vilify what they cannot comprehend. —Samuel Johnson.

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