Congenital dysplastic left ventricle simulating anomalous origin of the left coronary artery[1]

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The authors describe a rare case of congenital dysplasia of the left ventricle. Clinical findings were suggestive of anomalous origin of the left coronary artery from the pulmonary artery. The left ventricle showed areas of endocardial fibroelastosis, with an abnormal and unusually fine, sometimes lacy, filigree-like trabeculation of the internal surface of the endocardium. The endocardium resembled a mat of webbing on the endocardial surface. Many of these fine bands traversed the ventricular cavity from wall to wall. The abnormal Q waves seen on the electrocardiogram may have been caused by an abnormal depolarization process that may have been taking place in the left ventricular trabecular net. Experimental embryologic work in the chick embryo demonstrated that the development of trabecular sheaths in the primitive ventricle that are oriented in a dorsoventral direction are an essential step in ventricular formation. The authors speculate that perhaps an arrest or delay in the full fusion of some trabecular sheaths may give rise to the intracavitary trabeculations demonstrated here.

Index terms: Case reports • Heart defects, congenital

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We describe a case of congenital dysplasia of the left ventricle. The cardiac pathology at postmortem examination was unique, and we could find only one other similar case in the literature.[1] This congenital cardiomyopathy can simulate anomalous origin of the left coronary artery from the pulmonary artery clinically.

Case report

A 15-week-old white girl was a seven-pound product of a full-term and uncomplicated pregnancy and vaginal delivery. The patient was discharged at two days of age in good condition. Five days later, however, she was admitted to another hospital for congestive heart failure, lethargy, and feeding intolerance. A two-dimensional echocardiogram revealed a markedly dilated left ventricle with poor contractility. The child was treated with digoxin, furosemide, and spironolactone with marked improvement. An electrocardiogram showed marked left ventricular hypertrophy. Metabolic screens for inborn errors of metabolism and Pompe's disease were negative. A thallium scan showed diffuse decreased uptake with marked decreased activity in the proximal anterolateral wall of the left ventricle.

At presentation to the Cleveland Clinic, the infant was 13 weeks old and appeared chronically malnourished. She was 60 cm long and weighed 10 pounds. The heart and respiratory rates were regular (150 and 130, respectively). Results of the head, ears, eyes, nose, and throat examination were normal. The chest was clear. The heart demonstrated a left ventricular lift. The first heart sound was normal. The second heart sound was split with a normal pulmonary component. There was a third and questionable fourth heart sound. A grade II/VI systolic murmur was noted at the left lower sternal border. The liver was palpable 3 cm below the right costal margin. Peripheral pulses were normal.

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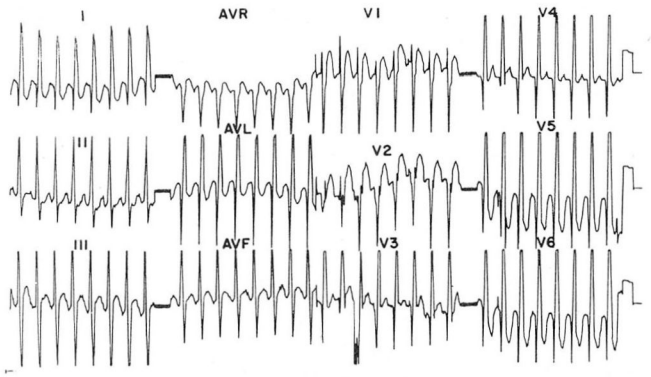

Fig. 1. Twelve-lead electrocardiogram, demonstrating sinus rhythm with deep Q waves in leads I, aV_L, V_5, and V_6 with T and ST segment changes and inverted T waves in V_5 and V_6.

The chest radiograph demonstrated cardiomegaly with biventricular hypertrophy. The electrocardiogram showed sinus rhythm with left ventricular hypertrophy. There were deep Q waves in leads I, aV_L, V_5, and V_6 with T and ST segment changes with inverted T waves in V_5 and V_6 (*Fig. 1*). The vectorcardiogram demonstrated left ventricular and septal hypertrophy and minimal intraventricular conduction abnormality. The vector direction in the horizontal plane was clockwise. A two-dimensional echocardiogram showed a markedly dilated left ventricle that contracted poorly with a shortening fraction of 12%. Some intracavitary band-like formations were seen that did not appear to contract. The origin of the left coronary artery from the aortic root was difficult to assess. A repeat resting thallium study demonstrated an enlarged heart with dilatation of the left ventricle. There were segmental areas of decreased thallium uptake involving the anterior and anterolateral left ventricular segments; the only area that approached normal uptake of thallium was the inferior left ventricular segment (*Fig. 2*).

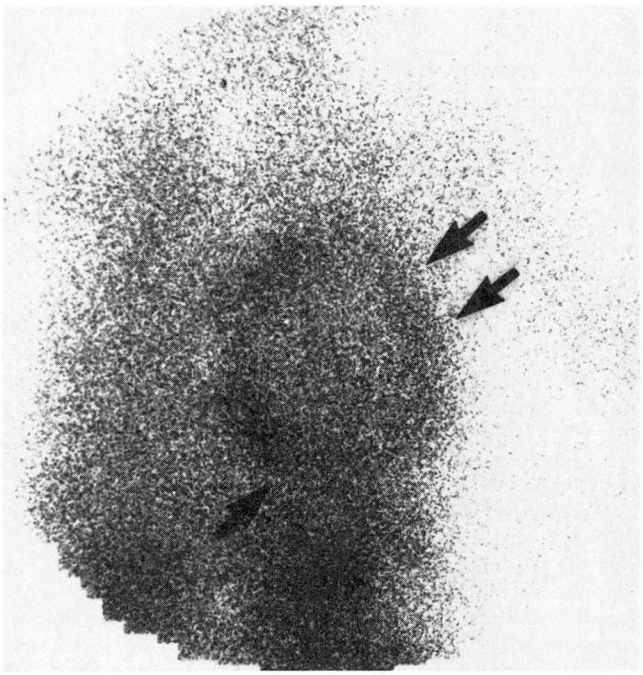

Fig. 2. Resting thallium study, demonstrating an enlarged heart. There are segmental areas of decreased thallium uptake involving the anterior and anterolateral left ventricular segments (double arrow). The only area that approached normal uptake of thallium was the inferior left ventricular segment (single arrow).

Fig. 3. Gross postmortem specimen, demonstrating the appearance of the right ventricle which appears to ride high on the shoulder of the left ventricle.

The infant underwent cardiac catheterization 2½ weeks following initial presentation to completely rule out the possibility of an anomalous origin of the left coronary artery from the pulmonary artery. The catheterization revealed a severe cardiomyopathy with a hugh dilated left ventricle with poor left ventricular function. The left atrium was dilated. The coronary arteries were normal. Right sided pressures were slightly elevated with a right ventricular pressure of 30/3-9 with a pulmonary artery pressure of 30/17 (mean, 23). Left ventricular pressure was 75/10-17 with no gradient demonstrated across the aortic valve. One hour following the catheterization, ventricular fibrillation developed and the infant died 4½ hours later.

Pathologic findings

Other than bilateral pulmonary congestion and edema, the abnormalities at postmortem examination were confined to the heart. The entire heart and lung weighed 180 g. The gross weight of the heart without the lungs was 105 g. The coronary arteries had a normal anatomic distribution with multiple cross sections demonstrating no significant narrowing of the lumen and no evidence of thrombosis. The right atrium and the tricuspid valve were normal, but the right ventricle had an abnormally enlarged moderator

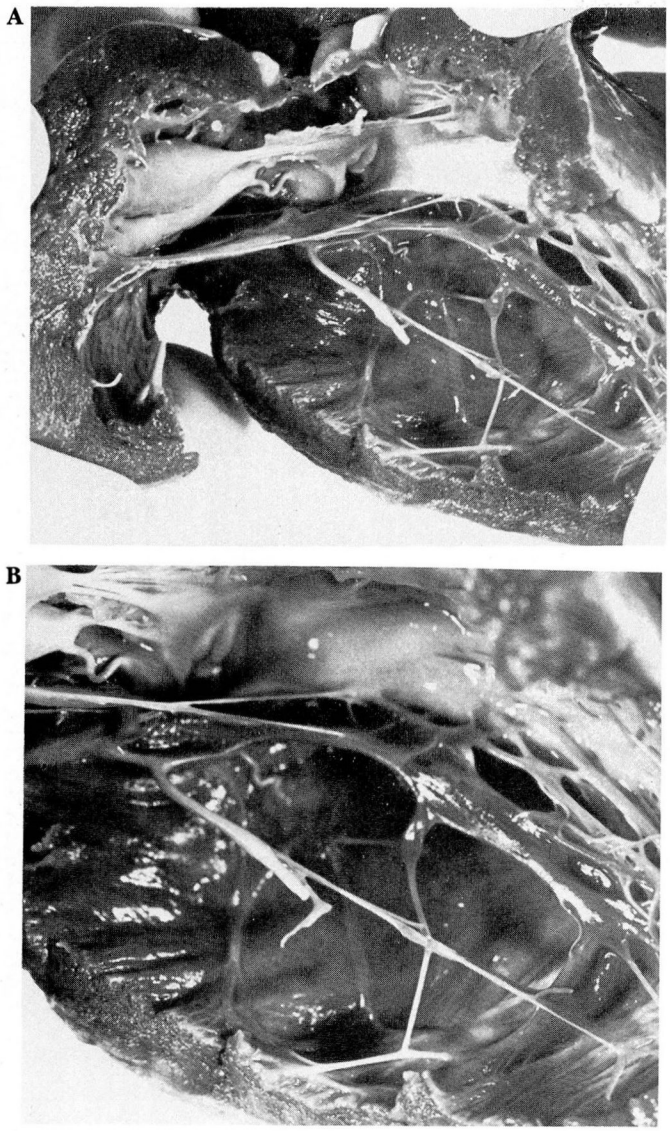

Fig. 4. A. Close-up view of the superior portion of the left ventricle. The top part of the picture shows the mitral valve, which is dysplastic and is displaced posteriorly and superiorly. Note the lacy, filigree-like trabeculations of the internal surface of the endocardium inferior to the mitral valve apparatus.

B. Close-up view of the region of the mitral valve and the left ventricle, demonstrating the superior displacement of the mitral valve and more clearly delineating the lacy, filigree-like trabeculations of the endocardial surface of the ventricle.

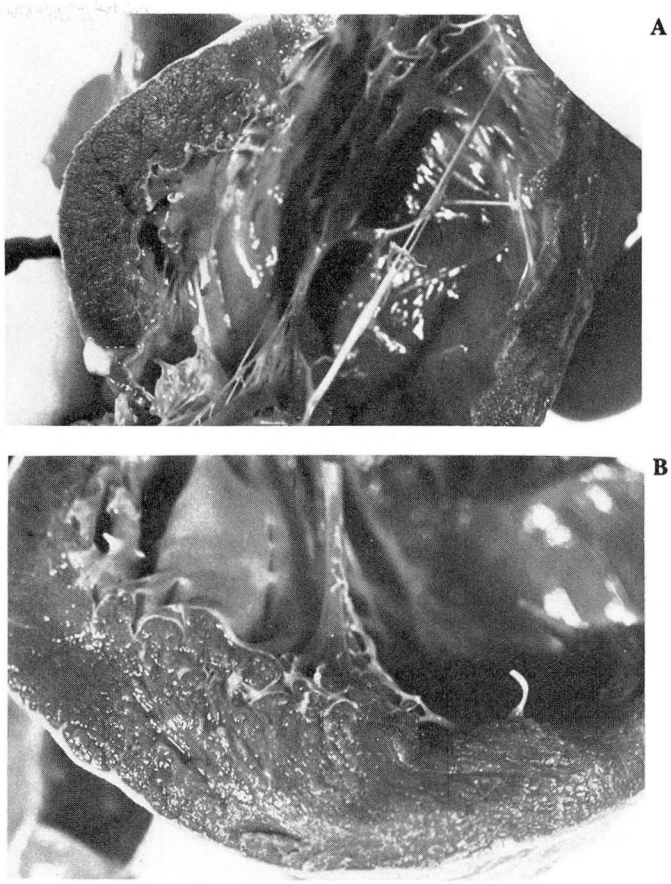

Fig. 5. A. Illustration of the endocardial fibroelastosis seen in the left side of the picture with marked hypertrophy of the ventricular wall in this region compared to the thinner dilated area of the right ventricle on the right side of the picture.

B. Illustration of the marked hypertrophy in the basilar, superior, and posterior region of the left ventricle. The remainder of the left ventricle on the right side of the picture is dilated with much thinner walls.

band. The right ventricle was cephalad to the left ventricle, riding high on the bulge of the left ventricle in a manner analagous to a hypoplastic right ventricle (*Fig. 3*), but was of normal size and appeared relatively small compared to the massively dilated left ventricle. The left atrium was normal. The mitral valve was dysplastic with the entire mitral valve apparatus (including the valve cusp, chordae tendineae, and papillary muscles) displaced posteriorly and superiorly in such a way that the papillary muscles inserted into the posterior wall and the chordae tendineae were somewhat shortened (*Fig. 4*). In the basilar, superior, and posterior region of the left ventricle, the white and thickened endocardium demonstrated the changes of fibroelastosis (*Fig. 5*). The left ventricular myocardium of this area was also thickened (up to 1 cm). The remainder of the left ventricle was markedly dilated and had much thinner walls than the perimitral valvular region (*Fig. 5*), as well as exhibiting an abnormal and unusually fine, sometimes lacy, filigree-like trabeculation of the internal surface of the endocardium (*Fig. 4*). This structure resembled a mat of webbing on the endocardial surface. Many of

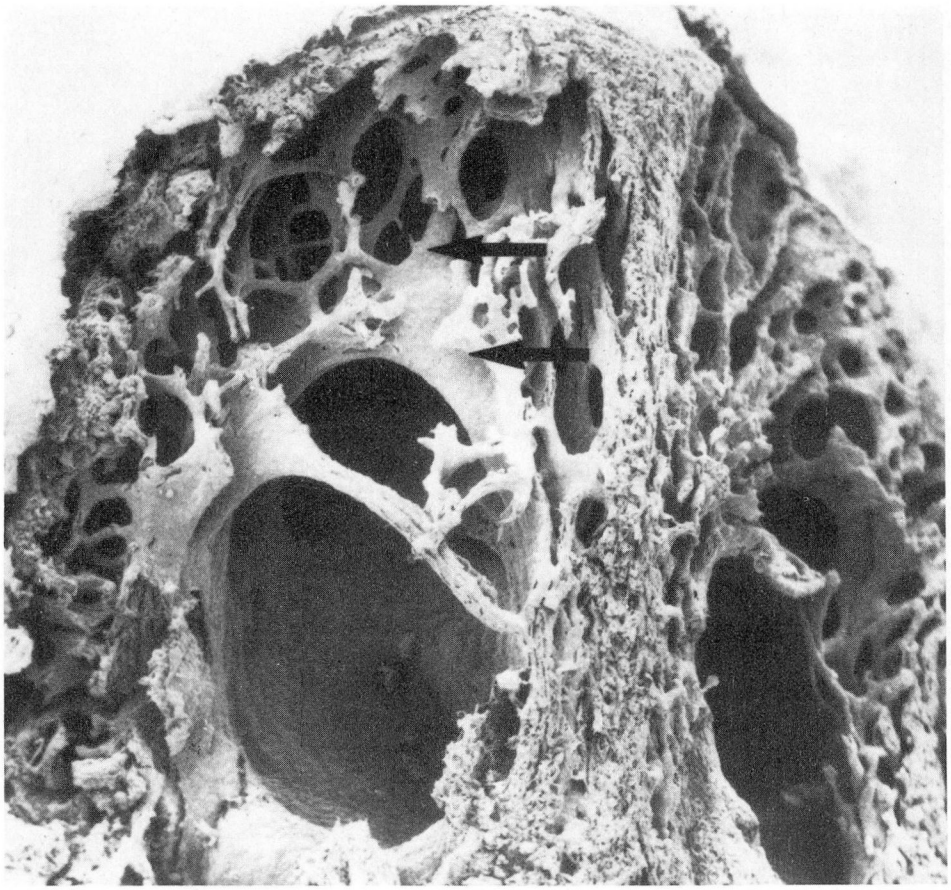

Fig. 6. Scanning electron micrograph of a chick embryo heart at Hamburger-Hamilton stage 26 (coronal section). The ventral wall has been removed. Trabecular sheaths (arrows) fill the left ventricular cavity and are oriented in the dorsoventral direction (×90).

these fine bands traversed the ventricular cavity from wall to wall. The aortic valve was normal.

Microscopic examination did not shed much light on either the etiology or the timing of this entity. The myocytes did not appear unusual. Focal myocardial fibrosis was noted in the thin portion. The thickened portion contained large, thin-walled vascular channels within the myocardium. There was also some endocardial fibrosis overlying the thick muscle, comparable to what was seen on gross examination.

Discussion

The unusual pathologic features of the left ventricle in this case are unlike any condition known to the authors except for the report by Ruttenberg et al.[1] It is interesting that both our case and theirs clinically suggested an anomalous origin of the left coronary artery from the pulmonary artery on the basis of the electrocardiogram, and yet both had the same pathologic left ventricular findings. The fibrous endocardial networks, as well as the marked thinning and dilatation of the left ventricle with focal areas of endocardial fibroelastosis in the hypertrophic portion of the left ventricle, were similar in both cases. Interestingly, the two-dimensional echocardiogram we obtained showed much fewer cavitary trabeculations than were found postmortem, and possibly only the major trabeculations reflected the echo beams.

The differentiation of complex cardiomyopathy from an anomalous origin of the left coronary artery in our patient was difficult. The existence of mitral regurgitation by physical examination, the abnormal Q waves in the 12-lead electrocardiogram, and the findings of the thallium scan were all suggestive of the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery. Only the vectorcardiogram did

not support this diagnosis. We speculate that the abnormal Q waves seen in the patient described by Ruttenberg et al[1] and our patient may have been caused by an abnormal depolarization process taking place in the left ventricular trabecular net. This type of complex cardiomyopathy should be considered in the differential diagnosis of anomalous origin of the coronary artery.

Franck[2] suggested that the ventricular bands may be related to the long muscular layers of the primitive heart. Experimental work with the chick embryo demonstrated the development of trabecular sheaths in the primitive ventricle that are oriented in a dorsoventral direction as an essential step in ventricular formation.[3] The coalescence of some of these sheaths form the muscular ventricular septum whereas the rest are fused with the free wall of the left ventricle (*Fig. 6*), thereby forming the left ventricular cavity. We speculate that perhaps an arrest or delay in the full fusion of such trabecular sheaths may give rise to intracavitary trabeculations, as was demonstrated in our patient. Therefore, it would seem more appropriate to refer to these left ventricular elements as residual ventricular trabeculations and not as false chordae tendineae or moderator bands.

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