Two-dimensional Echocardiographic Visualization of the Left Coronary Artery in Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

Pre- and Postoperative Studies

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SUMMARY Three young children with anomalous origin of the left coronary artery (LCA) from the pulmonary artery were studied by two-dimensional echocardiography. The LCA was shown to be in confluence with the left posterior aspect of the pulmonary artery root in the two patients studied preoperatively. In one of these patients, and in another patient 2 years after surgery, studied after direct surgical implantation of the LCA to the aorta, the LCA was shown to be confluent with the left anterior aspect of the aortic root. In all, the LCA could be followed beyond the branching point. This study demonstrates the feasibility of noninvasive diagnosis of anomalous origin of the LCA from the pulmonary artery by direct visualization with two-dimensional echocardiography.

NONINVASIVE VISUALIZATION of the left coronary artery (LCA) by two-dimensional echocardiography has been described in adults, infants and children. We examined one infant and two children known to have anomalous origin of the LCA from the pulmonary artery and were able to visualize the origin and course of the LCA before surgery in two and after direct surgical implantation to the aorta in two.

Materials and Methods

The three patients described below were studied by two-dimensional echocardiography and form the basis of this report.

Patient 1 initially presented with respiratory difficulty believed to be due to pneumonia at 3 weeks of age. At 3 months of age she was found to be in congestive heart failure. She had no murmurs, but a loud third heart sound was heard. The ECG was consistent with an anterolateral infarction. The chest x-ray showed cardiac enlargement, and the M-mode echocardiogram revealed a greatly enlarged left ventricle with very poor contraction. Aortography confirmed the diagnosis of anomalous origin of the LCA from the pulmonary artery. One day before surgery, when the patient was 18 months old, a two-dimensional echocardiogram was obtained. The study was repeated 3 days and 3 months after direct surgical implantation of the LCA to the aorta by a previously reported technique. Four months after surgery, the patient was still in chronic heart failure.

Patient 2 developed severe respiratory distress and diaphoresis by 2 months of age. On examination, no significant murmur was heard, but the liver was greatly enlarged. The ECG suggested anterolateral infarction, the chest x-ray showed an enlarged heart, and the M-mode echocardiogram showed an enlarged, poorly contracting left ventricle. Aortography showed anomalous origin of the LCA from the pulmonary artery. One day before surgery, when the patient was 9 months old, a two-dimensional echocardiographic study was done. At surgery, the short left main coronary artery could not be mobilized sufficiently to allow direct implantation to the aorta. A Gortex graft was anastomosed end-to-end to the LCA and end-to-side to the aortic root to establish antegrade flow from aorta to LCA. The infant could not be successfully removed from cardiopulmonary bypass. At autopsy, the graft was patent. The LCA divided into three major branches approximately 6 mm from the anastomotic site (fig. 1).

Patient 3 presented at 3 months of age with cough, tachycardia and tachypnea. There was radiographic cardiac enlargement and the ECG showed left ventricular hypertrophy. The M-mode echocardiogram demonstrated a large, poorly contracting left ventricle. Because of repeated respiratory infections, aortography was not done until the patient was 9 months of age. By then, he had ECG findings of anterolateral infarction. The study (figs. 2A and B) revealed anomalous origin of the LCA from the pulmonary...
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FIGURE 1. View of the base of the heart at autopsy in patient 2. The short left main coronary artery (LCA) has been detached from the posterior aspect of the pulmonary artery (PA) at the suture line (S). A Gortex graft (G), which was patent at autopsy, was anastomosed (wavy arrows) with the LCA and aorta (AO). The LCA divided into three major branches 6 mm from the anastomotic site: 1 = left circumflex; 2 = lateral oblique; 3 = left anterior descending. Branches 2 and 3 are labeled in areas where the vessels were dissected for identification. The aortic root (AoR) is to the right (R) and posterior (P) to the PA and site of origin of the LCA (S). A = anterior; L = left; LAA = left atrial appendage; PV = pulmonary valve.

artery. Two-dimensional echocardiography was not available at our institution at that time. He underwent direct surgical implantation of the LCA to the aorta.78 Aortography (fig. 2C) 6 months postoperatively showed antegrade filling of the small LCA from the aorta. At 36 months of age, 2 years postoperatively, the patient underwent two-dimensional echocardiography. This patient has been reported in detail.9 An infant with normal coronary arteries was also examined by the techniques described below and is presented for comparison.

Two-dimensional echocardiograms were done using an Advanced Technology Laboratories, Inc., Mark III Real Time Echocardiographic System with a three-element, 5-MHz rotating scan head. This instrument provides optimal resolution at a depth of 5-7 cm. There are 128 lines of true information per 90° image, and signal processing results in a total of 256 lines per image. When operated at a depth of 5-7 cm, as in this study, there are 56-40 frames/sec, respectively. The image was displayed on a video monitor and recorded in real time on ½-inch video cassette tape. Still frames were photographed using a 35-mm camera.

Patients 1 and 2 and the infant with normal coronary arteries were examined in the supine position, and patient 3 was examined in the left lateral position.

FIGURE 2. Aortograms in lateral views in patient 3. Panels A and B were recorded preoperatively; panel C was recorded postoperatively. Opacification of the large right coronary artery (RCA) from the aorta (AO) is seen in an early frame (A). The left coronary artery is not seen. Faint opacification of the anomalous left coronary artery (LCA) is seen later (B). Both coronary arteries fill from the aorta after surgery (C).
The transducer was placed at the left third intercostal space parasternally and positioned to obtain a parasternal long-axis view of the left ventricle (figs. 3A and B, position 1). By rotating the transducer clockwise approximately 90°, we obtained a cross-sectional view through the aortic root. With slight superior angulation, we obtained an approximate cross-sectional view of both great arteries (figs. 3A and C, position 2). A mass of echoes was seen related to the left postero-lateral border of the pulmonary artery and the left anterolateral border of the aorta. From position 2, varying degrees of clockwise rotation of the transducer with slight superior angulation visualized the pulmonary artery root in cross section in all cases allowed tracing of the cloud of echoes from left to right (figs. 3A and D, position 3).

Results

In the two preoperative cases, two-dimensional echocardiography demonstrated a markedly dilated, poorly contracting left ventricle (fig. 4). In all three patients and the normal infant, a linear, branching, echo-free structure was identified within the mass of echoes related to the aortic and pulmonary artery.
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FIGURE 5. Two-dimensional echocardiogram in the parasternal view between cross-sectional and oblique positions in a 3-week-old infant with normal coronary arteries, showing the left coronary artery (LCA) and its confluence with the aortic root (AO). PA = pulmonary artery. A = anterior; P = posterior; R = right; L = left.

In the infant with normal coronary arteries, this structure, believed to be the left coronary artery, was shown to be confluent with the aortic root at about 5 o'clock (fig. 5). In patient 1 preoperatively, the structure could be traced to the right, where it became confluent with the pulmonary artery root at about 5 o'clock (fig. 6). It could be followed to a branching point about 10 mm from the pulmonary artery root and beyond for a total of 30 mm. Distal branches were clearly seen about 20 mm from the pulmonary artery root. The greatest diameter of the vessel visualized was about 2 mm. Postoperatively in this patient (fig. 7), the same branching vessel seen preoperatively within the mass of echoes was now clearly confluent with the aortic root at about 3 o'clock, distinctly more anterior than has been reported in patients with normal origin of the LCA.1-3-

In patient 2 (fig. 8), the left coronary artery turned anteriorly to become confluent with the pulmonary artery somewhat more posteriorly than in patient 1. It could be traced for a total of 17 mm, branching 9 mm

FIGURE 6. Two-dimensional echocardiogram in the parasternal oblique view in patient 1, showing the left coronary artery (LCA) before surgery. This frame shows proximal (arrow) and distal branches (to the right) of the LCA and the confluence with the pulmonary artery (PA) on the left. AO = aorta; LV = left ventricle.

FIGURE 7. Two-dimensional echocardiogram in the parasternal cross-sectional view in patient 1, showing the left coronary artery (LCA) and its confluence with the aortic root (AO) 3 days postoperatively. The scale is unchanged compared with the preoperative study (fig. 6). RVo = right ventricular outflow tract. A = anterior; P = posterior; R = right; L = left.
from its origin. In some views, two branching points were visualized. The greatest diameter of the vessel seen was about 1.5 mm.

In patient 3 (fig. 9), studied 2 years postoperatively, the mass of echoes related to the left anterolateral border of the aortic root was easily recorded. The LCA was small (maximal diameter 0.6 mm) and could be traced for 20 mm from its confluence with the aortic root on the right and beyond its branching point on the left. The confluence with the aortic root was at about 3 o'clock.

In patients 1 (fig. 10) and 2, the right coronary artery was recorded at about 10 o'clock on the aortic root. It had a distinctly larger diameter than the LCA in both patients.

Discussion

The clinical diagnosis of anomalous origin of the LCA from the pulmonary artery is difficult because the lesion is rare and the clinical and ECG findings may be indistinguishable from those of several other conditions affecting the myocardium and commonly presenting with congestive heart failure in infancy.10, 11 In those even rarer patients with associated intracardiac defects, findings are masked by those of the associated lesion.12-14 This lesion has a very high mortality in untreated cases.10, 11, 13 Early diagnosis and definitive surgery before the occurrence of extensive myocardial damage appear to be the main hope for long-term survival.9, 10, 12 In support of this view are reports documenting remarkable improvement in left ventricular function after successful surgery in a few infants.9, 15, 17

Definitive diagnosis depends on aortography, which carries a significant risk in these sick infants.18 Formanek et al.18 recently suggested selective right coronary arteriography in patients in whom aortography is not diagnostic. In patients with associated lesions, the diagnosis may be missed or delayed despite the performance of such invasive diagnostic procedures.12-14 Caldwell et al.9 examined three patients with anomalous origin of the LCA from the pulmonary artery by two-dimensional echocardiography. The authors could not visualize the LCA in these patients and concluded that nonvisualization of the LCA os-

**Figure 8.** Two-dimensional echocardiogram in the parasternal view between cross-sectional and oblique positions in patient 2, showing the confluence of the left coronary artery (LCA) with the pulmonary artery (PA) 1 day before surgery. AO = aorta; A = anterior; P = posterior; R = right; L = left.

**Figure 9.** Two-dimensional echocardiogram in the parasternal cross-sectional view in patient 3, showing the small left coronary artery (LCA) and its confluence with the aortic root (AO) 2 years after surgery. PA = pulmonary artery; A = anterior; P = posterior; R = right; L = left.
tium could be used to differentiate this lesion from "myocardopathy." This approach provides little basis for definitive diagnosis. In the present report, we demonstrate the feasibility of definitive noninvasive diagnosis of anomalous origin of the LCA from the pulmonary artery by direct visualization with two-dimensional echocardiography. We noted, in addition, that the mass of echoes containing the LCA is related to the left aspect of the aortic root more anteriorly than has been described in patients with normal origin of the LCA and more anteriorly than in the infant with normal coronary arteries.

Various two-dimensional echocardiographic instruments have been used to visualize the LCA.\(^1\)\(^-\)\(^6\) We were able to trace the LCA in our patients farther than has been reported with use of the parasternal approach\(^1\)\(^-\)\(^3\)\(^-\)\(^6\) and visualized the branching point in all cases. The instrument used provides 256 lines per 90° image, more than twice the number of lines on the instruments used in previous reports.\(^1\)\(^-\)\(^2\)\(^-\)\(^6\) The availability of a 5-MHz transducer with its superior resolution may also have facilitated recognition and tracing of the LCA.

Although we have no documentation that the echo-free structure shown is in fact the LCA, good indirect evidence supports our interpretation. The mass of echoes containing the LCA was located in the approximate position described by previous groups,\(^1\)\(^-\)\(^6\) one of which proved by contrast injection that the echo-free structure observed within this mass of echoes was the LCA. Echocardiographic change from preoperative confluence with the pulmonary artery to postoperative confluence with the aorta after aortic implantation of the LCA in patient 1 is probably the best evidence that this structure is indeed the LCA.

In conclusion, a definitive noninvasive diagnosis of anomalous origin of the left coronary artery from the pulmonary artery can be made by direct visualization with two-dimensional echocardiography. Although the findings described are specific when recognized, until further experience confirms the sensitivity of this method, aortography (and possibly right coronary arteriography in selected cases) remains the definitive diagnostic procedure. The sensitivity of the method depends in large part upon operator experience and availability of an instrument and transducer capable of providing resolution adequate for visualization of 1-2-mm vessels.

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