Cross-Sectional Echocardiographic Diagnosis of Coronary Artery Aneurysms in Patients with the Mucocutaneous Lymph Node Syndrome

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SUMMARY Real-time cross-sectional echocardiographic studies revealed the presence of coronary artery aneurysms in five patients with mucocutaneous lymph node syndrome. These lesions appeared as circular echo-free spaces with clearly-defined borders in sites corresponding to angiographically proven aneurysms. In 15 normal subjects who were studied only by noninvasive methods, and in 17 who had normal coronary arteriograms (including eight with the mucocutaneous lymph node syndrome), we found no similar echo findings. The aneurysms were in both right and left coronary arteries in three patients, and were confined to the left side in two. The mucocutaneous lymph node syndrome is an increasingly common condition in Japan which may be fatal due to myocardial infarction occurring in a setting of coronary aneurysm with thrombosis. Therefore, the ability to demonstrate coronary aneurysms noninvasively is of prognostic and potentially therapeutic value in this inadequately understood syndrome.

CROSS-SECTIONAL ECHOCARDIOGRAPHY provides, noninvasively, spatial information about cardiac structures. Although the coronary arteries have been visualized extensively only by coronary arteriography, noninvasive recording of the left main coronary artery using cross-sectional echocardiography has recently been achieved.1, 2 This approach stimulated our interest in noninvasive diagnosis of aneurysms of the coronary arteries using cross-sectional echocardiography. The purpose of this study was to use this technique to detect coronary artery aneurysms in the mucocutaneous lymph node syndrome. This syndrome is widespread in Japan, with more cases reported each year.3-6 The cause of death is almost invariably myocardial infarction related to coronary artery aneurysm.7, 8

Material and Methods

Cross-sectional echocardiographic examination of the main coronary artery was performed in 15 normal subjects who did not have invasive studies, 17 patients with angiographically proven normal coronary arteries and five patients with coronary artery aneurysm in whom the diagnosis was established by retrograde aortography or selective coronary arteriography. Normal subjects were children or young adults without coronary artery disease. The patients with normal main coronary arteries included eight patients with mucocutaneous lymph node syndrome and nine with anterior chest pain. All the patients with coronary artery aneurysm had the mucocutaneous lymph node syndrome. Four of the five patients were referred for cardiac evaluation because of clinical features of an acute febrile mucocutaneous lymph node syndrome, fulfilling the diagnostic criteria proposed by a specially appointed Japanese investigative committee.3, 5 Four of the patients ranged in age from 11 months to 11 years. The remaining patient was 28 years old. He had a history consistent with this syndrome at age 11, and suffered an acute myocardial infarction at age 27. Coronary artery aneurysms were demonstrated angiographically, and he underwent double aortocoronary saphenous vein bypass grafting to the left anterior descending and right coronary arteries. Three patients had aneurysms in both the left and right main coronary arteries, and two patients only in the left main coronary artery. Cross-sectional echocardiographic examination was performed using an Aloka SSD-111 Echograph with a
FIGURE 1. Cross-sectional echogram of the aorta and coronary ostia. No abnormal structures in the areas of the left and right main coronary arteries are found. The aorta is viewed from the left ventricular chamber, so the patient’s right side is to the left of the figure. Ao = aorta; LA = left atrium; R and L = right and left coronary ostia.

Pulse repetition rate of 4.3 kilocycles/sec. A 2.25 MHz transducer mechanically driven through an approximately 60° sector at a variable rate of 5–40 frames/sec was used. The sector angle was manually adjustable from 30°–90°. The system was routinely operated at a frame rate of approximately 30 frames/sec, which yielded a line density of 110 lines/frame. Images were permanently recorded on 8 mm cinefilm and videotape, while phase-selected single frames were recorded on Polaroid film using the electrocardiograph gated circuit.

The technical details in cross-sectional echocardiographic visualization of the left main coronary artery were consistent with the recommendations of Weyman et al. The approach used in this study was from the long axis scan of the aorta. The transducer was then rotated 90° to a position parallel to the short axis of the aorta. The probe, with the plane of the sweep maintained perpendicular to the short axis of the aorta, was swept in a superior-inferior arc. During the course of this sweep, any abnormal structure in the areas of both the left and right main coronary arteries was investigated. The transducer was then rotated parallel to the short axis or horizontal cross-section of the body in an attempt to align the plane of the ultrasonic scan more closely to the course of the left main
coronary segment. In addition, the probe was rotated to a position close to the perpendicular section of the body rather than the short axis of the aorta to align the transducer sweep parallel to the long axis of the right main coronary artery.

**Results**

No abnormal structures in the area of the left or right main coronary artery were found in normal patients or in patients with normal main coronary arteries. It was possible to record the ostia of the coronary arteries in all (fig. 1). The left main coronary segment was visualized in 29 of the 37 patients (fig. 2), and the proximal right main coronary artery in 17. In the two patients with aneurysm in the left main coronary artery, there was an area of outward bending of the anterior and posterior wall echoes in the region of the left main coronary artery.

Figure 3 is a cross-sectional echogram from a patient with an aneurysm in the middle portion of the left main coronary artery. The circular echo-free space within clearly defined boundaries in communication with the inferolateral margin of the aorta is visualized.
This cross-sectional echocardiographic pattern corresponded to the angiographically demonstrated aneurysm in the left main coronary artery (fig. 4). A similar pattern was observed in the other patient with exclusively left-sided involvement, while bilateral areas of this type were noted in the region of the left and right main coronary arteries in each of the three patients with two lesions. Figure 5 is a cross-sectional echocardiogram from a patient with a large aneurysmal dilatation in both coronary arterial systems. Extensive areas of luminal dilatation distal to the coronary ostia were observed. Both the left and right main coronary arteries communicating with the lesions were visualized. In figure 6, an angiocardiogram corresponding to the cross-sectional echocardiographic study in figure 5 is shown. Large aneurysmal formation in both the left and right main coronary arteries was observed.

In four of five patients the left main coronary artery communicated with an aneurysm. In the remaining patient the left main coronary segment and aneurysm were visualized individually. In one patient with a right coronary aneurysm, we saw the right main coronary artery communicating with an aneurysm.

During the cardiac cycle there was both superior-inferior and anteroposterior motion of the aortic root. The main coronary arteries and aneurysm followed this motion. At end-diastole they were in their most
posterior-superior position, and at approximately end-systole in their most anterior-inferior position.

Discussion

Recent reports using a real-time cross-sectional echocardiographic scanner have demonstrated the feasibility of recording the left main coronary artery. The artery appears as two dominant parallel linear echoes, with a clear space representing the lumen of the vessel. Significant obstruction of the left main coronary artery was associated with almost complete obliteration of the echo-free arterial lumen. Weyman et al. have demonstrated the ultrasonic features of a case with aneurysmal formation in the left main coronary artery. This stimulated our interest in the ultrasonic diagnosis of coronary disease in the mucocutaneous lymph node syndrome, which is known to be associated frequently with coronary artery aneurysm. This syndrome is a specific arteritis with a predilection for the main coronary arteries. Since Kawasaki first described the clinical features of the syndrome in 1967, nearly 10,000 cases have been reported in Japan. Recently, attention has been drawn to this syndrome by American pediatricians, and reports of cases in various parts of the US are increasing. Although the majority of infants with this syndrome survive, coronary arterial lesions have frequently been demonstrated by coronary arteriography. The natural history of coronary aneurysms in this syndrome may be to regress or to occlude as a result of thromboendarteritis. Fatal cases are the result of myocardial infarction due to this pathologic mechanism. Noninvasive visualization of the lesion could therefore be of great clinical value, not only in the initial pre-angiographic assessment of infants with this syndrome, but also in the longitudinal follow-up of infants with proven coronary artery aneurysm. This study demonstrates the ability of real-time cross-sectional echocardiography to detect an aneurysm of the right or left main coronary artery. Visualization of aneurysms is facilitated when they are relatively large, and by their location and motion, which are analogous to those of the coronary arteries. The aneurysmal lesion appears as a large, circular, free area with clearly definable boundaries.

There are several limitations or problems in the assessment of coronary artery aneurysms using cross-sectional echocardiography. First, the technique can visualize only proximal portions of the coronary arteries, while the left anterior descending or circumflex coronary branches or the distal right coronary artery may be the site of the lesion. It is not known whether lesions in these areas can be detected using this technique. However, most of the aneurysms in the mucocutaneous lymph node syndrome are located in the left main coronary artery or the proximal portion of the right main coronary artery. Therefore, the technique could be useful in patients with this syndrome, and might prevent unnecessary coronary cineangiography.

Secondly, several structures commonly visualized in areas close to the main coronary arteries should be differentiated from a coronary artery aneurysm. Therefore, to determine convincingly the presence of the lesion, we must observe a distinctive area within clearly definable boundaries which has a motion and location similar to that of the coronary arteries. In the area of the left main coronary segment, the posterior wall of the pulmonary artery and left atrial appendage should be excluded. In the area of the right main coronary artery, echoes from the tricuspid valve and its
FIGURE 5. Cross-sectional echogram from a patient with a large aneurysmal formation in both the left main coronary artery and right main coronary artery. Extensive areas of luminal dilatation distal to the coronary ostia are visible. Both the left and right main coronary arteries communicating with the lesions are visualized. There are dense echoes from the aneurysmal walls of the left main coronary artery, which may be related to severe wall change or calcification due to endoarteritis. PA = pulmonary artery; Ao = aorta; LA = left atrium; L and R = left and right coronary aneurysm.

apparatus should be avoided. Weyman et al. emphasized that to be sure one is visualizing the left main coronary artery, its origin from the aorta and continuity between the lumina of the two vessels must be recorded. Similarly, in patients with coronary artery aneurysm, continuity between the lumen of the aorta and the aneurysm, which was observed in most of our patients, is an important factor in the diagnosis. However, this continuity is not always perceived, since the main coronary arteries are not perfectly straight in every patient. In addition, echoes of a coronary artery aneurysm should be differentiated from those of sinus of a Valsalva aneurysm. Well-delineated saccular echoes originating directly from the aorta would support diagnosis of the latter. Frequent changes in transducer angulation are therefore necessary in the differential diagnosis.

Finally, since a coronary artery aneurysm moves in and out of the plane of the cross-sectional scan, analysis of a number of still frames is required to visualize the lesion clearly.

This study dealt primarily with the validity of cross-sectional echocardiography in detecting the presence of a coronary artery aneurysm. Size of the aneurysms could only be approximated, since it was not always possible to align the transducer parallel to the long axis of the aneurysms and their angiographic contours were too irregular for quantitation of the size. Therefore, we did not attempt to quantitate size by cross-sectional echocardiography for comparison with angiography.

Our report supports the feasibility of this approach in patients with the mucocutaneous lymph node syndrome. However, this study provides no information
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Figure 6. Aortograms corresponding to the cross-sectional echogram in figure 5. Huge aneurysmal formation is observed in the left main coronary artery (left) and in the proximal portion of the right main coronary artery (right). The aneurysm in the right main coronary artery is indicated by arrows.

Concerning the sensitivity and reliability of this technique in routine clinical practice. It may be far less useful in the detection of aneurysms related to atherosclerotic coronary disease which may be more distal in the coronary artery and may be considerably smaller in both absolute and relative terms than those related to the mucocutaneous lymph node syndrome.

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