Venous Anomalies of the Coronary Sinus: Detection by M-mode, Two-dimensional and Contrast Echocardiography

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SUMMARY The echocardiographic features of the enlarged coronary sinus are described in eight patients with a left superior vena cava draining to the coronary sinus and two patients with total anomalous pulmonary venous connection to the coronary sinus. The diagnosis was confirmed in each patient by cardiac catheterization and surgery. On the M-mode echocardiogram, a dense echo was present posterior to the mitral valve at the level of the atrioventricular junction. The clear space immediately behind this echo represented the enlarged coronary sinus. On the two-dimensional echocardiographic examination, the enlarged coronary sinus was seen in several planes; however, the structure was imaged best with the parasternal long-axis view. In the long-axis plane in the eight patients with a left superior vena cava, the coronary sinus was visualized as a distinct circular structure lying posteriorly in the atrioventricular junction. In the two patients with total anomalous pulmonary venous connection, the confluence of the pulmonary veins and its connection to the coronary sinus were imaged in the long-axis plane. These findings were substantiated by contrast M-mode and two-dimensional echocardiograms in five patients. The M-mode and two-dimensional echocardiographic examinations allowed detection of the enlarged coronary sinus and diagnosis of certain venous anomalies that caused increased coronary sinus blood flow. The two-dimensional echocardiographic examination easily distinguished the enlarged coronary sinus from other congenital lesions that cause anomalous echoes behind the anterior mitral valve leaflet on M-mode echocardiographic examination.

THE ECHOCARDIOGRAPHIC DETECTION of an enlarged coronary sinus may be helpful in the diagnosis of any congenital anomaly that causes increased coronary sinus blood flow. The most common congenital anomalies of the coronary sinus, a persistent left superior vena cava draining to the coronary sinus and total anomalous pulmonary venous connection to the coronary sinus, cause marked enlargement of the coronary sinus that can be detected by M-mode echocardiographic examination.1-3

We report the M-mode and two-dimensional echocardiographic features of the enlarged coronary sinus in eight patients with a left superior vena cava to the coronary sinus and in two patients with total anomalous pulmonary venous connection to the coronary sinus. Contrast echocardiography is described as an important technique for differentiating persistent left superior vena cava and total anomalous pulmonary venous connection from other congenital cardiac defects.

Materials and Methods

Eight patients with a left superior vena cava draining to the coronary sinus and two patients with total anomalous pulmonary venous connection to the coronary sinus were studied prospectively by M-mode and two-dimensional echocardiography. They ranged in age from 1 day to 18 years. Complete cardiac catheterization, including oximetric shunt determinations and biplane cineangiograms, was performed in all patients. At the time of cardiac catheterization, the catheter was advanced through the coronary sinus into a persistent left superior vena cava in eight patients. The typical appearance on fluoroscopy of the catheter course combined with a determination of oxygen saturation and pressure established the diagnosis of a left superior vena cava connecting to the coronary sinus.4-6

The diagnosis of total anomalous pulmonary venous connection to the coronary sinus was established in two patients at cardiac catheterization by oximetric
data and by the angio graphic demonstration of the confluence of the four pulmonary veins draining into the coronary sinus. The cardiac catheterization findings were confirmed at surgery in all 10 patients.

The M-mode echocardiograms were obtained using an Ekoline 20A Ultrasonoscope interfaced with a strip chart recorder. The patients were in the left lateral decubitus position and the transducer was positioned in the third or fourth intercostal space at the left sternal border. The ultrasound beam was directed in the usual manner to record left ventricle, mitral valve, aorta and tricuspid valve.7

Two-dimensional echocardiograms were recorded with a Varian Associates 80° phased-array sector scanner. A 32-element 3.5-MHz transducer was used. The patients were again examined in the left lateral decubitus position with the transducer in the second or third left intercostal space. The parasternal long-axis view was obtained by directing the ultrasonic plane between the apex and base of the heart. From this position the transducer was rotated 90° clockwise to obtain the short-axis image of the heart. The apical four-chamber view was imaged with the technique previously described.8 The transducer was placed over the cardiac apex and angled so that all four cardiac chambers were visualized simultaneously. The two-dimensional echocardiographic examinations were recorded on videotape for later analysis at real-time or slow speeds. The figures presented here were taken from Polaroid photographs of stop-action, single-frame images from the videotape recordings. As a result, the photographs lack the integrated image and the visual appreciation of motion normally present in the real-time recordings.

Contrast M-mode and two-dimensional echocardiograms were obtained in five patients (three with a left superior vena cava and two with total anomalous pulmonary venous connection) by injecting saline into a left arm vein.

### Results

All 10 patients with venous anomalies of the coronary sinus had technically adequate M-mode and two-dimensional echocardiograms. The correct diagnosis was made before cardiac catheterization in each patient. Pertinent clinical data are summarized in table 1.

#### M-mode Echocardiography

An anomalous echo was identified at the level of the atrioventricular junction in the eight patients with left superior vena cava connecting to the coronary sinus (fig. 1A). This echo appeared posterior to the mitral valve echoes and represented the combined echoes arising from the posterior left atrial wall and the anterior wall of the enlarged coronary sinus. The clear space immediately posterior to the anomalous echo represented the coronary sinus. The dense echoes from the anterior wall of the coronary sinus disappeared as the ultrasound beam was directed toward the aortic valve. On a left ventricular sweep, these echoes disappeared below the atrioventricular junction.

The echoes arising from the anterior wall of the coronary sinus had a phasic movement similar to the movement of the anterior mitral valve leaflet. This phasic movement, illustrated in figure 1A, appeared to reflect primarily the volume changes of the left atrium throughout the cardiac cycle. As left atrial filling occurred during ventricular systole, the left atrial volume increased and the anomalous echo was displaced posteriorly. Anterior movements occurred during the phase of rapid ventricular filling and during atrial systole.

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**Table 1. Clinical, Catheterization and Surgical Data in 10 Patients with an Enlarged Coronary Sinus**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (year)</th>
<th>Diagnosis by catheterization</th>
<th>Vena cava present</th>
<th>Operative procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>TJ</td>
<td>1.5</td>
<td>Tetralogy of Fallot</td>
<td>RSVC, LSVC</td>
<td>Complete repair tetralogy of Fallot</td>
</tr>
<tr>
<td>EV</td>
<td>18</td>
<td>ASD</td>
<td>RSVC, LSVC</td>
<td>Repair ASD</td>
</tr>
<tr>
<td>SY</td>
<td>17</td>
<td>Severe subAS</td>
<td>No RSVC, LSVC</td>
<td>Resection subAS</td>
</tr>
<tr>
<td>NL</td>
<td>7</td>
<td>Coarctation of Ao, bicuspid Ao valve, PDA</td>
<td>RSVC, LSVC</td>
<td>Repair coarctation, ligation PDA</td>
</tr>
<tr>
<td>DD</td>
<td>2 mos</td>
<td>Coarctation of Ao</td>
<td>RSVC, LSVC</td>
<td>Repair coarctation</td>
</tr>
<tr>
<td>NA</td>
<td>14</td>
<td>Congenital MS, PAH, mild AS, postop repair coarctation of Ao</td>
<td>RSVC, LSVC</td>
<td>MV replacement</td>
</tr>
<tr>
<td>SB</td>
<td>3</td>
<td>ECD, PAH</td>
<td>RSVC, LSVC</td>
<td>Repair ECD</td>
</tr>
<tr>
<td>DS</td>
<td>15</td>
<td>Supravalvar mitral ring, mild MS, dextroversion</td>
<td>RSVC, LSVC</td>
<td>Resection supravalvar mitral ring</td>
</tr>
<tr>
<td>CM</td>
<td>6 mos</td>
<td>TAPVC to CS</td>
<td>RSVC</td>
<td>Complete repair TAPVC</td>
</tr>
<tr>
<td>BA</td>
<td>1 day</td>
<td>TAPVC to CS</td>
<td>RSVC</td>
<td>Death at attempted complete repair TAPVC</td>
</tr>
</tbody>
</table>

Abbreviations: Ao = aorta; AS = aortic stenosis; ASD = atrial septal defect; CS = coronary sinus; ECD = endocardial cushion defect; LSVC = left superior vena cava; MS = mitral stenosis; MV = mitral valve; PAH = pulmonary artery hypertension; PDA = patent ductus arteriosus; RSVC = right superior vena cava; subAS = subaortic stenosis; TAPVC = total anomalous pulmonary venous connection.
Two-dimensional Echocardiography

In all eight patients with a left superior vena cava to coronary sinus, the enlarged coronary sinus was imaged best with the long-axis view (fig. 3A). The coronary sinus was seen in this view as a discrete circular structure adjacent and slightly superior to the posterior mitral valve leaflet. There appeared to be no phasic changes in the internal diameter of the coronary sinus; however, the entire structure moved anteriorly in diastole with left atrial wall motion. During two-dimensional contrast studies, echoes from a left-arm-vein injection appeared first in the circular coronary sinus and then opacified the right ventricle, confirming the presence of a persistent left superior vena cava draining to the coronary sinus (fig. 3B).

The enlarged coronary sinus could be seen in the short-axis view in all eight patients at the level of the anterior mitral valve leaflet and just below the standard short axis plane through the aorta. Figure 4A shows the enlarged coronary sinus, which appeared as a crescent-shaped structure posterior to the left ventri-
Figure 2. Contrast M-mode echocardiogram from a 1-day-old infant (BA) with total anomalous pulmonary venous connection to the coronary sinus (CS). Echoes from a peripheral left-arm-vein injection are seen in the left atrium (LA) and right ventricle (RV) almost simultaneously due to obligatory right-to-left atrial shunting. The aorta (Ao) fills subsequently due to forward flow from the LA. The CS, because of its pulmonary venous drainage, is the only structure that remains free of echoes from the contrast injection. The structure that fills deep to the CS probably represents the descending Ao.

Figure 3. A) Long-axis image from a 17-year-old female (SY) with severe subaortic stenosis and a left superior vena cava draining to the coronary sinus (CS). The right superior vena cava was absent in this patient. The CS is seen in cross-section as a discrete circular structure (black arrows) lying in the atrioventricular junction. B) Contrast two-dimensional echocardiogram in the long-axis plane in a 7-year-old female with coarctation of the aorta and a persistent left superior vena cava to the CS. Contrast filled the CS (posterior black arrows), then appeared in the right ventricle (RV) (anterior black arrows). On the scale to the right of each image, 1 cm is the distance from the top of one white line to the top of the next line. Ao = aorta; LA = left atrium; LV = left ventricle.

cle and continuous with the right atrium. Echoes from a left-arm-vein saline injection appeared posterior to the left ventricle and flowed in the coronary sinus toward the right atrium (fig. 4B). The contrast echoes entered the right atrium and subsequently the right ventricle.

In the apical four-chamber view, the enlarged coronary sinus appeared as an oval-shaped structure along the lateral border of the left atrium (fig. 5A). During two-dimensional contrast studies from a peripheral left arm vein, contrast echoes first opacified the oval-shaped coronary sinus and then appeared in the right atrium as a stream arising near the atrial septum (figs. 5B and 5C).

In the two patients with total anomalous pulmonary venous connection to the coronary sinus, the enlarged coronary sinus was seen in the standard long-axis view just superior to the mitral valve (fig. 6A). With slight superior and medial angulation of the transducer, the confluence of the pulmonary veins and its connection to the coronary sinus were imaged. The pulmonary venous confluence coursed posterior to the true left atrium, which appeared small, to join the coronary sinus just superior to the anterior mitral valve leaflet.

In the patients with total anomalous pulmonary venous connection to the coronary sinus, the confluence of the pulmonary veins was imaged in the short-axis view of the heart at the level of the aortic valve (fig. 6B). The confluence of the pulmonary veins was imaged posterior and just to the right of the aortic root and was continuous with a large right atrial chamber.

In the two infants with total anomalous pulmonary venous connection to the coronary sinus, we could not clearly identify the pulmonary venous confluence or the enlarged coronary sinus in the apex view. Contrast
two-dimensional echocardiograms were obtained in both patients from left-arm-vein injections. The echoes from the contrast injection were seen filling the right atrium, right ventricle, left atrium, and left ventricle due to right-to-left atrial shunting. Because the microcavitations are cleared by passage through the lung, the coronary sinus and pulmonary venous confluence were the only cardiac structures that were not opacified by contrast echoes. During cardiac catheterization in one patient, the catheter was advanced into the coronary sinus and pulmonary venous confluence, and a contrast injection was performed. In both the long- and short-axis views, the structures described above as representing the pulmonary venous confluence and coronary sinus opacified immediately with echoes. In addition, dense echoes arising from the catheter could be seen lying in the coronary sinus and pulmonary venous confluence.

Discussion

The most common congenital anomaly involving the coronary sinus, a persistent left superior vena cava connecting to the coronary sinus, occurs in 3–10% of children with congenital heart disease and in approximately 0.5% of children without congenital heart disease.4, 5, 9, 10 Total anomalous pulmonary venous connection to the coronary sinus occurs in less than 1% of children with congenital heart disease.11 Other congenital anomalies of the coronary sinus are extremely rare.9 In cardiac catheterization performed from the femoral vessels in children, the left superior vena cava is detected only when the catheter is accidentally advanced into the enlarged coronary sinus ostium and up into the left superior vena cava. When the diagnosis is not made preoperatively, the presence of a left superior vena cava may cause difficulty for the cardiac surgeon. Special venous cannulation may be required for cardiopulmonary bypass.12 For these reasons, a noninvasive means of detecting a left superior vena cava connecting to the coronary sinus would be extremely useful.

The M-mode echocardiographic findings of total anomalous pulmonary venous connection to the coronary sinus have been described previously.2, 3 The report of Aziz and colleagues3 described M-mode findings in one patient with a left superior vena cava to the coronary sinus and a secundum atrial septal defect. The M-mode findings in our 10 patients are in agreement with these earlier works.

Although the M-mode echocardiogram is useful for detecting the enlarged coronary sinus, it does not distinguish between congenital anomalies with increased coronary sinus blood flow and other structures that cause left atrial echoes. These structures include the descending aorta, pulmonary veins, and left atrial appendage, as well as rare congenital anomalies such as cor triatriatum or supravalvar mitral ring. Saline contrast echocardiography is helpful in some instances in delineating the origin of anomalous left atrial echoes.

The two-dimensional sector scan, however, allows differentiation of echoes due to an enlarged coronary sinus from those due to descending aorta, pulmonary veins, cor triatriatum, or supravalvar mitral ring. In the parasternal long-axis view, the thoracic aorta and pulmonary veins are visualized posterior to the posterior left atrial wall. The coronary sinus is positioned at the same level as the posterior left atrial wall and has a distinct circular appearance. In one patient with both a supravalvar mitral ring and a left superior vena cava to the coronary sinus, we could distinguish easily the supravalvar mitral membrane, which was distinctly different in appearance and location in the left atrium from the coronary sinus. The
The appearance of the coronary sinus on the two-dimensional apex echocardiogram has been previously reported and is distinct from our apex image of the enlarged coronary sinus.

Two-dimensional echocardiography did not distinguish a patient (SY) with an absent right superior vena cava from the seven patients with bilateral superior vena cavae. In addition, we would predict that two-dimensional echocardiography alone could not differentiate left superior vena cava to the coronary sinus from other congenital anomalies that cause increased coronary sinus blood flow such as partial anomalous hepatic venous connection to the coronary sinus, continuity of the inferior vena cava to the hemiazygous vein to a left superior vena cava, and coronary artery to coronary sinus fistula. However, a saline contrast study using the left arm vein could distinguish the patient with a left superior vena cava to coronary sinus. We have seen several patients with two-dimensional echocardiographic findings of an enlarged coronary sinus who were not included in this report because they did not undergo cardiac catheterization or surgery. These patients have had cardiomyopathy, chronic right-heart failure, or left ventricular hypertrophy that would explain the enlargement of the coronary sinus. In over 2000 two-dimensional echocardiographic studies, we have not imaged the coronary sinus in any normal pediatric patient.

In all 10 patients with enlarged coronary sinus, the long axis was the easiest plane for detecting the coronary sinus. The coronary sinus courses around the heart in the atrioventricular junction in a plane perpendicular to the long-axis plane of the heart; therefore, the coronary sinus was seen in the long-axis view as a discrete circular structure. The coronary sinus was most difficult to visualize in the short-axis plane where careful use of gain and reject controls was required to image the area behind the left ventricle. Its crescent shape in this view mimicked a posterior pericardial effusion; however, the absence of echocardiographic evidence of a pericardial effusion in the other precordial planes substantiated that this structure was indeed an enlarged coronary sinus. The coronary sinus was seen with difficulty in the apex four-chamber view. Careful transducer angulation to "open" the left atrial cavity and strict attention to gain settings were necessary to image the coronary sinus in this view. We could not visualize the enlarged coronary sinus in the apex view in the two small infants with total anomalous pulmonary venous connection. This difficulty may have been related to technical difficulties encountered in apex imaging in small infants. When all views were used, however, the coronary sinus was seen without difficulty in patients DD, CM and BA, who all weighed less than 4 kg. Patient age and size were not limiting factors in detecting the coronary sinus by two-dimensional echocardiography.

The two-dimensional echocardiogram, particularly
the long-axis view and saline contrast studies, allows precise imaging of the enlarged coronary sinus, facilitating the detection of certain congenital anomalies of the coronary sinus. The finding of a venous anomaly of the coronary sinus provides important information for planning cardiac catheterization or open heart surgery.

References

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