DIAGNOSTIC METHODS
CONGENITAL HEART DISEASE

Study of the infradiaphragmatic total anomalous pulmonary venous connection with cross-sectional and pulsed Doppler echocardiography

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ABSTRACT We studied neonates with the infradiaphragmatic form of total anomalous pulmonary venous drainage by a combination of cross-sectional echocardiography and pulsed Doppler ultrasound. The diagnosis by ultrasound was made prospectively in all six patients. Three large vascular channels could be observed passing through the diaphragm from the subcostal parasagittal plane. The vessels were identified as the descending aorta (to the left), the inferior vena cava (to the right), and the anomalous pulmonary venous channel (in the center). The vessels were insolated in turn, with pulsed Doppler ultrasound, and the characteristic normal flow signals in the aorta and inferior vena cava were obtained. The signal from the anomalous pulmonary vein was a continuous venous signal, the direction of flow being away from the heart. Pulsed Doppler ultrasound allows accurate recognition of the anomalous pulmonary venous channel without the use of contrast echocardiography.


CROSS-SECTIONAL ECHOCARDIOGRAPHY has proven useful in the rapid diagnosis of infradiaphragmatic total anomalous pulmonary venous connection (TAPVC) in the sick newborn.1,2 We have shown previously that the rapid recognition of the anomalous pulmonary venous channel can be enhanced through the use of contrast echocardiography.3

Range-gated pulsed Doppler ultrasound is unique in its ability to assess the character and direction of blood flow within vessels visualized by two-dimensional echocardiography. Unlike contrast echocardiography, the use of this method requires no venous access, and we therefore evaluated the combined use of cross-sectional echocardiography and range-gated pulsed Doppler ultrasound for the rapid noninvasive identification of infradiaphragmatic TAPVC in six infants.

Methods

From March 1 to July 30, 1983, we examined six consecutive infants with infradiaphragmatic TAPVC with cross-sectional echocardiography/range-gated pulsed Doppler ultrasound. The infants were referred for evaluation because of a variety of clinical, electrocardiographic, and radiologic findings and ranged in age from 1 to 30 days at the time of diagnosis. In all six the diagnosis of infradiaphragmatic TAPVC was confirmed on catheterization.

The cross-sectional scans were obtained with an ATL Mark 500 mechanical scanner with full spectral output interfaced with a 5 MHz transducer. The transducer was placed in the subcostal region and the scan was oriented in a sagittal body plane and swept from left to right to scan the thoracoabdominal vessels. In a sagittal plane from the subcostal view the aorta was identified to the left and posterior, the inferior vena cava was noted in an anterior position and to the right, and the descending anomalous pulmonary vein was identified in an intermediate position (figure 1).

The scanner could be stopped along any line within the image and the Doppler sample volume could be positioned at any depth along the line for sampling. The sample volume was teardrop shaped and was approximately 1.25 mm long and 1.5 mm wide. The operational mode of the scanner was switched from real-time imaging to spatially oriented Doppler sampling with simultaneous M mode and electrocardiographic display for timing. The pulse repetition frequency rate varied depending on the depth, with the maximum frequency display (Nyquist limit) ranging between 6.4 and 3.2 KHz. There were two outputs of the Doppler signal. The first was an audio signal that allowed differentiation between arterial and venous signals. The arterial signal had a characteristic phasic quality, whereas the venous signal had a more steady blowing quality. The inferior vena cava frequently had a superimposed phasic quality due either to
TABLE I

Patient data

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex</th>
<th>DOB</th>
<th>Age at presentation</th>
<th>Weight</th>
<th>Clinical findings</th>
<th>ECG findings</th>
<th>X-ray findings</th>
<th>IVC sat. (%)</th>
<th>Systemic sat. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>2/83</td>
<td>1 month</td>
<td>2.9 kg</td>
<td>Cyanosis, dyspnea, single second sound, Goldenbar’s syndrome (ventilated)</td>
<td>Axis 210°</td>
<td>Bilateral pneumothorax, right upper lobar consolidation</td>
<td>41</td>
<td>74</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>4/83</td>
<td>1 day</td>
<td>3.4 kg</td>
<td>Cyanosis, split second sound</td>
<td>Poor LV forces</td>
<td>Small heart, interstitial edema</td>
<td>100</td>
<td>98</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>5/83</td>
<td>3 days</td>
<td>2.8 kg</td>
<td>Cyanosis, split second sound</td>
<td>Within normal limits</td>
<td>Within normal limits</td>
<td>62</td>
<td>77</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>3/83</td>
<td>1 day</td>
<td>3 kg</td>
<td>Cyanosis</td>
<td>RVH</td>
<td>Cardiomegaly, pulmonary edema</td>
<td>69</td>
<td>74</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>6/83</td>
<td>1 month</td>
<td>3.2 kg</td>
<td>Cyanosis, poor perfusion, split second sound</td>
<td>RVH</td>
<td>Mild cardiomegaly, increased pulmonary vascular markings, pulmonary edema</td>
<td>56</td>
<td>46</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>6/83</td>
<td>1 day</td>
<td>3 kg</td>
<td>Cyanosis</td>
<td>QR right precordium</td>
<td>Moderate pulmonary edema, increased pulmonary vascular markings</td>
<td>92</td>
<td>83</td>
</tr>
</tbody>
</table>

DOB = date of birth; IVC = inferior vena cava; LV = left ventricle; RVH = right ventricular hypertrophy; sat. = saturation.

FIGURE 1. Top. Image of subcostal sagittal plane obtained from patient No. 2 in the orientation used for performing pulsed Doppler examination. The anomalous pulmonary vein (APV) is seen behind the left atrium (LA) and anterior to the aorta (AO) as they pass through the diaphragm. The spine, lying behind the aorta, is labeled. The right pulmonary artery (PA) can be seen lying superior to the left atrium. RA = right atrium; RV = right ventricle. Bottom. Diagrammatic representation of the anatomy seen from the subcostal parasagittal position shown in figure 1, left. The image is shown in anatomic orientation. The sample volume (small bar on the scan line in each of the vessels insonated) and resulting Doppler spectral outputs are displayed diagrammatically. Flow toward the transducer is the signal above the baseline and flow away from the transducer is the signal below the baseline. The aortic signal has the characteristic pulsatile quality representing flow toward the transducer. The inferior vena cava signal shows a phasic flow away from the baseline and transducer. The signal from the anomalous pulmonary vein shows amorphous flow away from the heart, i.e., toward the transducer and above the baseline.
Results

The clinical findings and those of cardiac catheterization are listed in table 1. With cross-sectional echocardiography from the subcostal sagittal position, the anomalous channel could be traced from the abdomen through the diaphragm to a position adjacent to the inferior margin of the left atrium in all six infants (figure 1).

While the cross-sectional echocardiographic display was being monitored, the sample volume was positioned in each of the abdominal vessels and the Doppler signal was recorded (figures 2 to 4). The angle between the direction of flow and position of the pulsed Doppler insonation was between 15 and 48 degrees. In all six patients a typical tracing of antegrade pulsatile aortic flow was recorded from the descending aorta (figure 2), while in the inferior vena cava approximately 45 degrees to the angle of flow. Bottom, Doppler display showing an amorphous flow signal away from the transducer (toward the heart). The signal is represented below the baseline. The direction display and scale are as in figure 2.

FIGURE 2. Top, Cross-sectional echocardiogram showing the descending thoracic aorta near the diaphragm. The sample volume in the aorta is at an angle of 48 degrees. Bottom, The aortic Doppler recording showing pulsatile forward flow. The markers show Doppler frequency shift of 1 KHz. Flow is toward the transducer, i.e., above baseline.

respiratory movement or to the sample volume being placed close to the atria. The signal in the anomalous vein was uniform and lacked any phasic variation with the exception of that due to respiratory movements.

The second output was a quantitative fast-Fourier transform spectral analysis of the Doppler frequency shift sampled at 100 times/sec. The direction of the signal was defined from the middle of the recording strip. Flow toward the transducer was displayed above the baseline, whereas flow away from the transducer was displayed below it.

FIGURE 3. Top, Doppler sample volume has been placed in the inferior vena cava approximately 45 degrees to the angle of flow. Bottom, Doppler display showing an amorphous flow signal away from the transducer (toward the heart). The signal is represented below the baseline. The direction display and scale are as in figure 2.
cava, a low-velocity tracing of retrograde flow was noted (figure 3). In the vessel identified as the infradiaphragmatic common pulmonary vein, we recorded a low-velocity venous flow tracing oriented towards the transducer (figure 4). Thus, in all six patients two venous vessels were defined in the combined cross-sectional echocardiography/range-gated pulsed Doppler ultrasound study; in the inferior vena cava flow was toward the heart and away from the subcostally positioned transducer and in the common pulmonary vein the flow was toward the transducer and away from the heart.

**Discussion**

Infants with infradiaphragmatic TAPVC are usually cyanotic and may present with severe respiratory distress associated with the development of increasing pulmonary venous obstruction. This defect can be difficult to differentiate clinically from persistent pulmonary hypertension of the newborn or pulmonary disease. Cross-sectional echocardiography has proved useful in the diagnosis of the supracardiac forms of TAPVC, which appear as a large common pulmonary venous chamber behind the left atrium, a large vertical vein, or an enlarged coronary sinus. Cross-sectional echocardiographic assessment of infradiaphragmatic TAPVC is somewhat more difficult since the pulmonary veins may converge at a great distance from the atrium just above the diaphragm. A subcostal transducer position may allow imaging of the infradiaphragmatic common pulmonary vein in such cases. With the use of contrast echocardiography, this vessel can be differentiated from the inferior vena cava and descending aorta because the common pulmonary vein remains echofree due to filtration of the contrast echoes in the pulmonary capillary bed. The obligatory right-to-left shunt causes the aorta to fill with contrast medium and reflux causes filling of the inferior vena cava. Contrast echocardiography requires rapid injection of 1 to 2 ml of sterile saline into a peripheral vein with simultaneous imaging of the abdominal vessels to detect the presence or absence of contrast microcavitations. Insofar as continuous imaging of the abdominal vessels in a critically ill infant who is hyperventilating may be difficult, repeated injections of saline may be required to define the anatomy. In addition, because there is a potential risk of air embolus in the presence of a large right-to-left shunt, a safer means of validation of structure is desirable.

Once these vessels have been imaged by cross-sectional echocardiography, pulsed Doppler echocardiography offers an effective alternative method to contrast echocardiography for the identification of the common pulmonary vein, the descending aorta, and the inferior vena cava in infants with infradiaphragmatic TAPVC. By assessing the quality of the Doppler flow signal, the pulsatile high-frequency signal indicating descending aortic flow can easily be differentiated from the low-frequency venous signal. The directionality of flow in the two venous vessels can then be used to differentiate these structures. Flow away from the heart occurs in a common pulmonary vein, whereas flow...
toward the heart occurs in the inferior vena cava. There are few structures that might be confused with an anomalous channel descending through the diaphragm. The infradiaphragmatic esophagus shares the same hiatus in the diaphragm as the common pulmonary vein, but is usually imaged for a shorter distance than the anomalous vein and does not produce a flow signal on Doppler insonation. A large azygos vein may also be imaged but its position is posterior and to the left of the aorta. Doppler insonation of this vessel shows flow toward the heart. Although in these six patients all the pulmonary veins were found to drain into a common pulmonary venous channel, cases of mixed anomalous drainage with one vein draining into a vertical vein, into the coronary sinus, or directly into an atrium may give an identical echocardiographic/Doppler pattern. This must be considered when other investigative or surgical procedures are performed.

These echocardiographic findings are specific for the infradiaphragmatic anomalous pulmonary venous channel. Indeed, since the previous report on contrast echocardiography in infants with TAPVC, use of the technique has resulted in the correct diagnosis in 14 consecutive patients.

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References
Study of the infradiaphragmatic total anomalous pulmonary venous connection with cross-sectional and pulsed Doppler echocardiography.
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