Evaluation of Infradiaphragmatic Total Anomalous Pulmonary Venous Connection with Two-dimensional Echocardiography

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SUMMARY In three newborn infants with infradiaphragmatic total anomalous pulmonary venous connection, the common pulmonary vein was visualized with two-dimensional echocardiography and validated with saline contrast injections. The transducer was placed in the subcostal region in a sagittal body plane so as to image the descending aorta and the vertebral column. The common pulmonary vein, which connected the pulmonary veins to a systemic vein in the abdomen, was seen lying parallel and anterior to the descending aorta and to the left of the inferior vena cava. With peripheral venous contrast injections, contrast echoes first filled the inferior vena cava and then the descending aorta because of obligatory right-to-left atrial shunting. The common pulmonary vein was the only structure that remained free of contrast echoes. Thus, contrast echocardiography provided a method for identifying the site of pulmonary venous drainage and for distinguishing the common pulmonary vein from other structures such as the inferior vena cava. In cyanotic infants with respiratory distress, two-dimensional contrast echocardiography permits a rapid diagnosis of infradiaphragmatic total anomalous pulmonary venous connection.

IN PATIENTS with infradiaphragmatic total anomalous pulmonary venous connection (TAPVC), the pulmonary veins drain into a systemic vein below the diaphragm rather than into the left atrium. The anomalous connection is made through a common pulmonary vein, which descends into the abdomen anterior to the aorta and through the esophageal hiatus of the diaphragm. Pulmonary venous obstruction is common in this disorder, and infants with infradiaphragmatic TAPVC are usually critically ill with cyanosis and pulmonary edema. Infradiaphragmatic TAPVC can mimic pulmonary disease, persistent pulmonary artery hypertension, and other forms of cyanotic heart disease; therefore, in cyanotic infants with respiratory distress, a method of rapidly detecting or excluding infradiaphragmatic TAPVC is essential. In this report, we describe a simple technique for identifying and validating the common pulmonary vein with two-dimensional contrast echocardiography.

Methods
We examined three infants, ages 1, 2 and 12 days, with two-dimensional contrast echocardiography. In two infants, the echocardiographic diagnosis was made before cardiac catheterization and surgery. In one infant, the echocardiogram was performed after cardiac catheterization at another institution and with knowledge of the catheterization results. In all three patients, the diagnosis of infradiaphragmatic TAPVC was confirmed by a complete cardiac catheterization and by direct visualization of the pulmonary veins at surgery.

The two-dimensional echocardiograms were performed using a Toshiba SSH 10A Sonolayergraph with a 2.4-MHz transducer. With the patient lying supine, the transducer was placed in the subcostal region and oriented in a sagittal body plane so as to image the descending aorta and vertebral column. The transducer was then angled toward the patient’s right to image the inferior vena cava. Also, the transducer was placed in the subcostal region in a transverse body plane so as to image the vessels in the abdomen in cross section. Two-dimensional contrast echocardiography was performed by rapidly injecting 1–4 ml of sterile saline through a 23-gauge needle positioned in an arm or leg vein.

Results
The transducer was positioned in the subcostal area in a sagittal body plane and swept from left to right in order to scan the abdominal vessels. The descending aorta was imaged slightly to the left and directly in front of the vertebral column (figs. 1–3). In all infants, another vessel was seen parallel to the descending aorta just below the diaphragm. This vessel, which was the common pulmonary vein, was seen anterior to the descending aorta. With angulation of the transducer to the patient’s right, the inferior vena cava was imaged. In two infants, we obtained a plane passing through a portion of the inferior vena cava, common pulmonary vein, and descending aorta simultaneously. However, in most instances, the common pulmonary vein was positioned to the left of the upper inferior vena cava. In one of the patients, the lower portion of the common pulmonary vein could be followed anteriorly to its junction with a dilated venous structure thought to be a hepatic vein (fig. 3).

To verify that the vessel parallel to the aorta was the common pulmonary vein, a contrast echocardiogram was performed with the transducer positioned in the subcostal region so as to image the descending aorta, common pulmonary vein and, if possible, a portion of the inferior vena cava (figs. 1, 2 and 4). In the patient in whom a leg vein injection was used, the inferior vena cava and right atrium filled first with contrast echoes. In the two patients in whom an arm vein injec-

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tent pulmonary artery hypertension or pulmonary disease in the newborn. The M-mode and two-dimensional echocardiograms in all three conditions show four normally positioned cardiac valves and chambers.

Discussion
Infants with infradiaphragmatic TAPVC are usually cyanotic and in severe respiratory distress. This defect can be difficult to differentiate clinically from persis-
Inferiorly, the CPV curves anteriorly to its junction with a dilated venous structure, which is probably a hepatic vein. A = anterior; I = inferior.

two normally positioned great arteries, and right heart dominance. In most instances, TAPVC can be diagnosed on the basis of echocardiographic examination by identifying the common pulmonary venous chamber into which the pulmonary veins drain posterior to the left atrium.\(^4\) In the supracardiac or cardiac forms of TAPVC, the common pulmonary venous chamber is usually a large structure immediately posterior or slightly superior to the left atrium\(^6\) and is, therefore, usually visible in several precordial echocardiographic planes. In infracardiac TAPVC, the pulmonary veins usually converge like the branches of a tree just above the diaphragm; therefore, the common pulmonary venous chamber is often small and inferior to the left atrium or may not exist as a distinct, separate chamber. Because of its size, shape and location, the common pulmonary venous chamber in infradiaphragmatic TAPVC can be difficult to image directly. Also, because of the technical limitations associated with examining small infants with respiratory distress, it can be difficult to visualize with certainty the wall separating the true left atrium from the common pulmonary venous chamber. If the pulmonary veins can be seen clearly draining into a common pulmonary venous chamber, which is separated from the left atrium, then TAPVC can be diagnosed with certainty.

The most common form of supracardiac TAPVC, TAPVC to the superior vena cava through a left vertical vein and innominate vein, can be diagnosed on the two-dimensional echocardiogram by direct visualization of the anomalous connection in the suprasternal views.\(^5\)\(^6\) Also, the most common form of cardiac TAPVC (TAPVC to the coronary sinus) can be diag-

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**FIGURE 3.** Subcostal long-axis view from a 12-day-old infant with infradiaphragmatic total anomalous pulmonary venous connection. The descending aorta (DAO) is seen in front of the vertebral column (V). The common pulmonary vein (CPV) in the upper abdomen is anterior to and parallel to the DAO. Inferiorly, the CPV curves anteriorly to its junction with a dilated venous structure, which is probably a hepatic vein. A = anterior; I = inferior.

**FIGURE 4.** (top) Subcostal view in a transverse abdominal plane from the same infant as in figure 3. The descending aorta (AO) is seen in cross section to the left and in front of the vertebra (V). The inferior vena cava (IVC) is seen to the right. The common pulmonary vein (CPV) is seen anterior to the AO and to the left of the IVC. Because the plane is taken where the CPV curves anteriorly in this patient (see figure 3), two portions of the CPV can be seen. (middle) After a leg vein contrast injection, contrast echoes first fill the IVC. (bottom) Subsequently, because of obligatory right-to-left shunting and forward flow in the left heart, the AO fills with contrast echoes. The only vascular structure free of contrast echoes is the CPV.
nosed on the two-dimensional echocardiogram by direct visualization of a greatly enlarged coronary sinus in several echocardiographic views. In infants in whom infracardiac TAPVC is suspected and in whom a common pulmonary venous chamber cannot be visualized with certainty, direct visualization of the common pulmonary vein below the diaphragm is a rapid, safe method for diagnosing this disorder. In two of our three patients, the common pulmonary venous chamber could be imaged from the subcostal four-chamber view as described by Sahn and colleagues; however, in the third infant, the common pulmonary venous chamber could not be seen with certainty from any view. The common pulmonary vein could be imaged easily in all three infants from a subcostal sagittal body plane.

Contrast echocardiography is an effective method for differentiating the common pulmonary vein in the abdomen from the inferior vena cava and from the descending aorta. In the infants in whom an arm vein injection was used, contrast echoes refluxed into the inferior vena cava and hepatic veins during vigorous atrial contractions. Reflux of contrast echoes into the inferior vena cava during atrial contraction occurs in some normal patients, during deep inspiration, and in patients with pulmonary artery hypertension, right atrial hypertension or right atrial volume overload. In infants with TAPVC, inferior vena caval reflux after an arm vein injection is probably related to several factors, such as right atrial volume overload, pulmonary artery hypertension, and tachypnea and respiratory distress. Although we have not examined an infant suspected of having TAPVC who did not have reflux of contrast echoes into the inferior vena cava from an arm vein injection, we believe that the contrast injection should be made when possible from a leg vein so that the inferior vena cava is completely filled with contrast echoes. With leg vein contrast injections, it should be easy to determine if a large venous structure in the abdomen is the inferior vena cava (which will fill with contrast echoes) or the common pulmonary vein (which will remain free of contrast echoes).

In infants with infradiaphragmatic TAPVC, the common pulmonary vein in the abdomen can be identified by two-dimensional echocardiography and validated by contrast echocardiography. Identification of the common pulmonary vein can be extremely useful for substantiating the two-dimensional echocardiographic diagnosis of infracardiac TAPVC when other echocardiographic features are present or can be the only echocardiographic finding suggesting the diagnosis of infracardiac TAPVC.

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