Range-gated Pulsed Doppler Echocardiographic Diagnosis of Supracardiac Total Anomalous Pulmonary Venous Drainage

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SUMMARY Three patients with total anomalous pulmonary venous drainage (TAPVD) into the innominate vein were studied by using M-mode and pulsed Doppler echocardiography. An abnormal continuous flow towards the transducer in a suprasternal notch position detected by pulsed Doppler echocardiography in the left hemithorax leftward of the aortic echo is a sign of blood flow through the left vertical vein into the innominate vein. This finding is highly specific for TAPVD. High-velocity flow across the tricuspid valve, disturbed flow in the right pulmonary artery and abnormal flow in the left atrium are additional important pulsed Doppler echocardiographic findings in TAPVD. Right ventricular enlargement and paradoxical interventricular septal motion by M-mode echocardiography were not specific enough to distinguish TAPVD from other right ventricular volume overload lesions. An echo-free space posterior to the left atrium was not recorded.

THE ECHOCARDIOGRAPHIC features of total anomalous pulmonary venous drainage (TAPVD) are not specific enough to make the diagnosis of this malformation possible. Some echocardiographic signs, including right ventricular diastolic volume overload, an echo-free space behind the left atrium, and an enlarged coronary sinus detected consistently posterior to the left atrium in TAPVD into this structure, are useful, but do not overcome the difficulty of making correct, noninvasive diagnosis of TAPVD.

This study presents the results of an investigation carried out using pulsed Doppler echocardiography in three patients with TAPVD into the innominate vein. Characteristic echocardiographic features have been revealed, allowing diagnosis before catheterization and angiography.

Methods

In this investigation we used a directional pulsed Doppler flowmeter (Pulsed Doppler 400 A, Advanced Technology Laboratories), connected by an Echo-Doppler Interface (900 A, Advanced Technology Laboratories) to an echocardiograph (Echo-cardio-Visor 03, Organon Teknika). We used a 5-MHz piezoelectric transducer with an outer diameter of 13 mm, focused at 5 cm. The transmitter circuit of the equipment emits 1-μsec pulses at a pulse-repetition frequency of 7.7–19.0 KHz, depending on the measurement depth. The reflected signal is used for M-mode echocardiography, as well as for obtaining Doppler flow-velocity information from a preselected, tear-drop-shaped area called the sample volume, whose dimensions are approximately 2 × 4 mm, in focus. By means of a depth control, the sample volume can be located at any position along the ultrasonic beam, with the maximum depth 9 cm. The signal from the sample volume is processed by a special phasedetector circuit and transformed into an acoustic signal corresponding to the Doppler shift spectrum and the blood flow direction. The other output type is a graphic on-line recording of the Doppler spectrum on a recorder (Visioorder LS-6A, Honeywell) or on the M/Q display of an echocardiograph oscilloscope. The M-mode echocardiogram is displayed on the upper part of the memory screen, with a line marking the sample volume location. The lower half of the screen displays the time-interval histogram, a graphic reproduction of the momentary Doppler spectrum. The forward flow (i.e., toward the transducer) is plotted above the baseline, and the reverse flow (i.e., away from the transducer) is plotted under the baseline. At turbulent flow, the blood cells have different directions and velocities, which are represented as a widely dispersed dot pattern (fig. 1). At laminar flow, the sample volume blood cells have almost uniform direction and velocity, which are represented on the time-interval histogram as a narrow dot pattern (fig. 2). Details concerning the method can be found elsewhere.

The patients were examined in the supine position, with the transducer located in the third or fourth intercostal space at the left sternal margin, as in conventional echocardiography. Suprasternal notch echocardiograms were obtained according to the Goldberg method. The beam was directed inferiorly and slightly leftward to visualize the transverse aortic arch and right pulmonary artery. An example of normal flow in these structures is presented in figure 3. A leftward scan starting at this point enables us to detect flow in the left vertical vein (fig. 4).

Right and left ventricular dimensions were measured in end-diastole, and left atrial dimension was measured in end-systole. The results are ex-
pressed in absolute values as well as in values adjusted for body surface area.

Results

Using the method described above, we were able to diagnose TAPVD in three children. The first child was male, in whom heart murmur and cyanosis were detected immediately after birth. He was referred to our center at the age of 14 months. At admission, mental retardation and failure to thrive (5500 g, 67 cm) were observed. The patient was mildly cyanotic. Clubbing of fingers was minimal. The cardiac apex was felt in the fifth left interspace in the midclavicular line. The heart rate was 124 beats/min, and the first heart sound at the apex was accentuated. The second heart sound in the pulmonary area was of normal intensity and split. There was a III/VI late ejection systolic murmur heard best in the pulmonary area. Respiratory rate was 48 breaths/min. The liver was palpated 2 cm under the right costal margin. The systolic blood pressure in both the upper and lower limbs was 125 mm Hg. The ECG showed right-axis deviation (+120°) and evidence of right ventricular

**Figure 1.** Pulsed Doppler echocardiographic pattern of a turbulent flow in aortic regurgitation. The upper part of the tracing is the M-mode echo display from the Doppler receiver. The sample volume is placed in the left ventricular outflow tract (LVOT). The sample volume position is indicated by the depth line. Time-interval histogram at the bottom of the tracing shows a widely dispersed dot pattern throughout diastole (indicated by ———X———) as a result of the regurgitant turbulent flow from the aorta. In systole there is normal flow toward the aorta.

**Figure 2.** Pulsed Doppler echocardiogram in a healthy infant. The sample volume is placed in the orifice of the tricuspid valve. Time-interval histogram at the bottom shows normal biphasic diastolic flow through the tricuspid valve. The dots that constitute the flow record are closely organized into distinct waves, indicating a smooth flow. TV = tricuspid valve.

**Figure 3.** Normal pulsed Doppler echocardiogram from the suprasternal notch position. Left) The sample volume is in the aortic arch (Ao) as indicated by the position of the depth line. Normal systolic flow is directed toward the transducer, i.e., above the baseline. Right) The sample volume is placed in the right pulmonary artery (RPA). Normal systolic flow is plotted under the baseline, i.e., the flow is away from the transducer.
and atrial hypertrophy with a slight clockwise rotation in the horizontal plane. On chest x-ray, the heart was moderately enlarged with a large right atrium. An anomalous left vertical vein and dilated left innominate vein and superior vena cava produced a characteristic “figure-of-eight” silhouette. The child was recommended for echocardiographic investigation with the diagnosis of TAPVD.

Echocardiography showed an enlarged right ventricle with A-type paradoxical motion of the interventricular septum\(^1\) (fig. 5 and table 1). The left atrium was of normal size (table 1). No echo-free space posterior to the left atrial wall was found. Pulsed Doppler echocardiography showed high-velocity flow through the tricuspid valve (fig. 6) and atypical flow in the left atrium (fig. 7). Normal findings are presented for comparison in figures 2 and 8. From the suprasternal notch approach, normal flow in the aortic arch and disturbed flow in the right pulmonary artery were recorded. With a leftward deflection of the transducer from the standard suprasternal position, an abnormal, continuous, slightly turbulent forward flow was discovered. The maximum velocity of the flow was reached in early diastole (fig. 9).

At cardiac catheterization the catheter tip entered from the left innominate vein into a vessel with a blood oxygen saturation of 98%. No pulmonary vein could be catheterized from the left atrium. The pulmonary arterial pressure was 38/20 mm Hg (mean 25 mm Hg). The mean pressure in the left atrium was 7 mm Hg. The pressure gradient between the right and left atria was 3 mm Hg. Blood oxygen saturation in the femoral artery was 83%. Angiocardiography proved TAPVD via the left vertical vein into the left innominate vein.

The second patient was a 3-month-old girl recommended for evaluation because of failure to thrive, heart murmur and cyanosis. Clinically, cyanosis was mild and the infant was hypotrophic (3870 g, 58 cm). Respiratory rate was 64 breaths/min, heart rate 140 beats/min. Precordial pulsation was diffuse. A systolic thrill was felt in the second left intercostal space, and a IV/VI systolic murmur was best heard in the same area. The first heart sound in the apical area was closely split and of normal intensity. There were no symptoms of pulmonary congestion. The liver was palpated 2 cm under the right costal margin. The systolic pressure in the upper and lower limbs was 110 mm Hg and 105 mm Hg, respectively. The ECG showed right-axis deviation of +110°, and evidence of right ventricular and atrial hypertrophy. There was moderate cardiomegaly on the chest x-ray and a wide right-upper mediastinum.

Echocardiography showed an enlarged right ventricle (table 1) and A-type paradoxical motion of the interventricular septum. No echo-free space behind the left atrium was found. In the smaller left atrium (table 1), an atypical forward flow was found on a Doppler echocardiogram, with the maximum velocity in systole and especially in end-diastole. High-velocity

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**Figure 4.** Anatomy and a sample volume location in total anomalous pulmonary venous drainage (TAPVD) (case 2). 

T = transducer; Ao = aorta; RPA = right pulmonary artery; PA = pulmonary artery; LVV = left vertical vein; SVC = superior vena cava; RA = right atrium; LA = left atrium; CPVC = common pulmonary venous connection.

**Figure 5.** Echocardiogram showing enlarged right ventricular cavity and paradoxical interventricular septal motion in case 1. Horizontal line in the right ventricle indicates the position of the sample volume. The vertical dashes and spaces represent 1 cm and occur every 0.5 second. RV = right ventricle; S = interventricular septum; LV = left ventricle; LVPW = left ventricular posterior wall.
TABLE 1. Echocardiographic Data for Three Patients with Total Anomalous Pulmonary Venous Drainage

<table>
<thead>
<tr>
<th>Case</th>
<th>RV dimension</th>
<th>LA dimension</th>
<th>LV dimension</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Size (cm)</td>
<td>Index (cm/m²)</td>
<td>Size (cm)</td>
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<tr>
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<tr>
<td>3</td>
<td>1.5</td>
<td>6.0</td>
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Abbreviations: RV = right ventricular; LA = left atrial; LV = left ventricular.

Figure 6. Pulsed Doppler echocardiogram in case 1. The sample volume is placed in the orifice of the tricuspid valve (TV). The time-interval histogram underneath the tracing shows high-velocity diastolic forward flow (i.e., above the baseline) with reverse flow components in the tricuspid valve. RA = right atrium.

Figure 7. Pulsed Doppler echocardiogram in case 1. Atypical flow wave forms were recorded from the left atrium. There was a disturbed nonlaminar flow pattern most likely consistent with a right-to-left atrial shunt instead of a discrete flow normally detectable in the left atrium. Ao = aorta; LA = left atrium.

Flow through the tricuspid valve was detected. Echocardiographic examination from the suprasternal notch showed an abnormal, continuous, high-velocity flow in diastole to the left of a normal flow in the aortic arch and a turbulent flow in the right pulmonary artery (fig. 10).

At catheterization, no pulmonary vein could be entered from the left atrium. The pulmonary arterial pressure was 16/6 mm Hg. The mean pressure in the left atrium was 5 mm Hg, and the pressure gradient between the right and left atria was 2 mm Hg. Blood oxygen saturation in the femoral artery was 80%. Angiocardiography showed TAPVD into the left innominate vein (fig. 11).

The third patient was a 2-month-old girl recommended for evaluation because of heart murmur and tachypnea. The infant was acyanotic, respiratory frequency was 64 breaths/min and heart rate was 132 beats/min. Precordial pulsation was diffuse. No thrill was felt. Heart sounds were of normal intensity, and the second heart sound in the pulmonary area was inconstant split. A II/VI short systolic murmur was best heard in the second left intercostal space. There were no symptoms of pulmonary congestion. The liver was palpated 2 cm under the right costal margin. The systolic pressure in the upper and lower limbs was 105 mm Hg and 110 mm Hg, respectively. The ECG showed right-axis deviation of +110° and evidence of
FIGURE 8. Normal flow pattern in the left atrium. Pulsed Doppler echocardiogram in a child with ventricular septal defect. LA = left atrium.

FIGURE 9. Case 1. Pulsed Doppler echocardiogram from the suprasternal notch position with a leftward transducer angulation. Abnormal, continuous, slightly turbulent forward flow was discovered, with maximum velocity in early diastole.

FIGURE 10. Case 2. Pulsed Doppler echocardiogram from the suprasternal notch position with a leftward transducer angulation. There was abnormal, continuous forward flow, with the maximum velocity in diastole.

right ventricular hypertrophy and incomplete right bundle branch block. On chest x-ray the heart was of normal size, with an enlarged mediastinal shadow, interpreted as hyperplastic thymus. Pulmonary arterial vasculature was increased.

Echocardiography showed right ventricular en-
largement (table 1) and A-type paradoxical motion of the interventricular septum. The left atrium was of normal size (table 1). No echo-free space behind the left atrium was found. Pulsed Doppler echocardiography showed high-velocity disturbed flow through the tricuspid valve and atypical forward flow in the left atrium, with maximum velocity in early diastole. From the suprasternal notch approach, the normal flow in the aorta and slightly disturbed flow in the right pulmonary artery were recorded. With a leftward deflection of the transducer from the standard suprasternal position, an abnormal, continuous, disturbed forward flow was found, with the maximum velocity in early diastole (fig. 12).

At catheterization no pulmonary vein could be catheterized from the left atrium. The catheter tip entered from the left innominate vein a vessel with blood oxygen saturation of 96%. The pulmonary arterial pressure was 25/10 mm Hg (mean 17 mm Hg). The mean pressure in the left atrium was 2 mm Hg and the pressure gradient between right and left atria was 2.5 mm Hg. Blood oxygen saturation in the femoral artery was 86%. Angiocardiography proved TAPVD via the left vertical vein into the left innominate vein.

Discussion

The range-gated pulsed Doppler method improves substantially the diagnostic capability of echocardiography in congenital heart disease. Our experience based on the investigation of more than 450 children with congenital heart disease has proved the advantage of range-gated pulsed Doppler method mainly in newborns and infants. Its value in detecting regurgitation or poststenotic flow turbulence has been documented repeatedly. Presence or absence of flow in the hypoplastic left heart and demonstration of the diastolic turbulent flow in the pulmonary artery in patent ductus arteriosus are only two examples of an application of the pulsed Doppler echocardiography in newborns. The diagnosis of abnormal blood flow in the left hemithorax described in this paper represents a new application of this method. The direction of the flow toward the transducer is a more important feature than its turbulent character. Both features found in the typical location seem to be specific for TAPVD into the left innominate vein, and discriminate TAPVD from a persistent left superior vena cava, in which the flow is directed towards the heart and away from the transducer. The same character and direction of the flow located in the left hemithorax could be seen with the extremely rare levoatriocardinal vein. However, other pulsed Doppler and M-mode echocardiographic findings are entirely different.

Other nonspecific signs of TAPVD on a pulsed Doppler echocardiogram in TAPVD include high-velocity flow through the tricuspid valve and turbulent systolic flow in the right pulmonary artery. However, these findings are also common in other heart diseases with a hyperkinetic circulation. Turbulence in the right pulmonary artery alone is often present in congenital heart diseases with a significant left-to-right shunt. An atypical flow in the left atrium, observed in all patients, is probably a manifestation of a right-to-left shunt through a defect in the atrial septum.

The M-mode echocardiographic signs of TAPVD reported in the literature, that is, enlargement of the right ventricle and a paradoxical motion of the interventricular septum, were found in our patients. However, these two findings are not specific. Signs of right ventricular volume overload can be found also with other heart malformations, e.g., in atrial septal defects or in tricuspid and pulmonary regurgitation. In TAPVD with obstruction or with pulmonary hypertension, the size of the right ventricle and the movement of the interventricular septum can be normal. Similarly, the importance of the discovery of an echo-free space behind the left atrium is limited and we have not been able to prove the latter in our patients. Furthermore, the common pulmonary venous trunk can be mistaken for nonspecific linear echoes, occasionally recorded in the left atrium, that may originate from the mitral annulus or from some part of the pulmonary veins. An echo-free space behind the left atrium can also be caused by coronary sinus. The common pulmonary venous trunk will be recorded only when located immediately behind the left atrium, which occurs only in some cases of supracardiac pulmonary venous drainage. The absence of an echo-free space on the echocardiogram, therefore, does not eliminate a diagnosis of TAPVD.

Glaser et al. and Meyer and Kaplan reported a
 diminutive left atrium in TAPVD. In two of our patients the left atrium was of normal size and in one it was small. The size of the left atrium seems to be function of the size of the interatrial communication. For the purpose of diagnosing TAPVD, pulsed Doppler echocardiography cannot be replaced by conventional suprasternal notch echocardiography. An echocardiographic recording requires a right-angle relation between the structure and the ultrasonic beam that in our patients could be achieved only in the most cranial part of the left vertical vein. However, in this area it is possible to mistake the anomalous trunk for the aortic arch or the right pulmonary artery. Pulsed Doppler echocardiography, in which the frequency shift of the reflected ultrasound depends on the cosine of the angle formed by the ultrasonic beam and the reflecting medium, will reproduce ideally the flow in the vessel running parallel to the transmitted ultrasonic beam.

Pulsed Doppler echocardiography may markedly improve our ability to diagnose, and it can be extremely helpful in infants with an atypical clinical picture, as was seen in our third patient. Diagnosis of TAPVD by this method does not offer detailed information about the hemodynamics and anatomy of the malformation. We do not think that pulsed Doppler echocardiography can replace cardiac catheterization and angiocardiography, but it can help to choose the most appropriate technique to shorten the procedure and to decrease the total amount of a contrast material injected.

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