Superior and Inferior Vena Cava Flow Velocity in Patients with Anomalous Pulmonary Vein Connection

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SUMMARY
The superior and inferior vena cava flow velocities (SVC and IVC flow velocities) of 3 patients with anomalous pulmonary vein connection were recorded with the Doppler flowmeter catheter. The cases consisted of a patient with partial anomalous pulmonary vein connection to the SVC (Case 1), one with total anomalous pulmonary venous connection to the SVC (Case 2), and one with partial anomalous pulmonary vein connection to the IVC (Case 3). The SVC and IVC recordings of these patients were compared with those of 10 normal subjects, 20 patients with atrial septal defect and 150 patients with other heart diseases.

The SVC and IVC flow velocities in the latter group of 180 patients showed a biphasic pattern, having systolic (S) and diastolic (D) waves, the peak S wave occurring around midsystole. In Cases 1 and 2, the SVC flow velocity showed a markedly delayed peak S wave, similar to the pattern of the pulmonary vein flow velocity; this pattern was also seen in the IVC flow velocity in Case 3. This abnormal pattern could be useful in diagnosing anomalous pulmonary vein connection.

Additional Indexing Words:
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About 10 to 15% of patients with atrial septal defect have associated anomalous pulmonary vein (PV) connection.1) Although clinically it is important to detect such an anomaly before operation, correct diagnosis

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is sometimes difficult even with routine cardiac catheterization study and angiocardiography.\textsuperscript{2,3} We have been studying flow velocity in the cardiac chambers and various vessels with a bidirectional Doppler flowmeter catheter and have found this approach to provide valuable diagnostic informations.\textsuperscript{4-12} In the present study, we examined the superior vena cava (SVC) and inferior vena cava (IVC) flow velocities in 3 patients with partial or total anomalous PV connection to the SVC or IVC. All patients showed a characteristic SVC or IVC flow velocity pattern that could be useful in diagnosing anomalous PV connection.

**Materials and Methods**

A 37-year-old man with atrial septal defect (ASD) associated with partial anomalous PV connection to the superior vena cava (Case 1), a 16-year-old boy with total anomalous PV connection to the SVC (Case 2), and a 6-year-old boy with ASD associated with partial anomalous PV connection to the IVC (Case 3, Scimitar syndrome) were studied. The control group was composed of 20 patients with an uncomplicated secundum type of ASD, 150 patients with other heart diseases, and 10 normal subjects admitted to Nagasaki University Hospital. The clinical diagnosis was confirmed by right and left heart catheterization and selective angiocardiography. The cardiac catheterization data in the 3 patients with anomalous PV connection (Cases 1, 2, and 3) are shown in Table I. The X-ray films, which show the course of the catheter passing through the anomalous PV in Cases 1 and 2, and a selected film from angiocardiogram in Case 3, are shown in Fig. 1. SVC and IVC flow velocities were measured with a bidirectional Doppler flowmeter catheter (supplied by Southwest Research Institute, San Antonio, Texas), developed by Stegall et al.\textsuperscript{13} A detailed description of this technique has been previously described.\textsuperscript{11} After routine cardiac catheterization study, the flowmeter catheter was advanced from a saphenous vein to the SVC or IVC under

<table>
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<th>Case No.</th>
<th>Patient</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>RA press. (mmHg)</th>
<th>LA press. (mmHg)</th>
<th>RV press. (mmHg)</th>
<th>PA press. (mmHg)</th>
<th>LV press. (mmHg)</th>
<th>Aortic press. (mmHg)</th>
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<tr>
<td>2</td>
<td>KH</td>
<td>16</td>
<td>M</td>
<td>7 3 6 4 4</td>
<td>7 3 7 4 4</td>
<td>50/0-6</td>
<td>50/12</td>
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<td>7 3 8 5 5</td>
<td>9 4 8 4 6</td>
<td>25/0-6</td>
<td>20/8</td>
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RA press. = right atrial pressure; LA press. = left atrial pressure; PA press. = pulmonary artery pressure; LV press. = left ventricular pressure.
Fig. 1. Chest X-ray films during cardiac catheterization. Upper left: Case 1. The catheter was passed from the superior vena cava to the partial anomalous pulmonary vein. Lower left: Case 2. The catheter was passed from the superior vena cava to the total anomalous pulmonary vein. Right: Case 3 (angiocardiogram). Contrast medium was injected into the anomalous pulmonary vein. The anomalous pulmonary vein which is connected to the inferior vena cava is seen (arrows).

continuous fluoroscopic control and monitoring of the flowmeter audiosignal by a loud speaker. The flowmeter catheter tip was positioned about 1 cm above the junction of the SVC with the right atrium to measure the SVC flow velocity, and about 1 cm below the junction of IVC with the right atrium to measure the IVC flow velocity.

In 10 patients with ASD and 5 patients with other heart diseases, the Doppler flowmeter catheter was advanced to the left upper PV through the ASD or the foramen ovale. The tip of the catheter was positioned 1 cm above the junction of the left atrium to measure the PV flow velocity. This flowmeter catheter was connected to a Doppler ultrasonic flowmeter, Model 806, and the output signals were recorded on a mingograph operated at a paper speed of 5 cm/sec or 10 cm/sec. Lead II of the electrocardiogram, the phonocardiogram at the 3rd left sternal border, and intracardiac pressure were also recorded simultaneously. All the recordings were made during expiration. Zero flow velocity was obtained by briefly disconnecting the in-
put signal to the frequency meter, as zero frequency shift corresponds to zero velocity. The recording was calibrated by taking a fixed frequency from a signal generator (Hewlett Packard Model 651 A) and applying it to the input of a voltage converter.

**RESULTS**

_The normal pattern of the SVC and IVC flow velocity_

The SVC and IVC flow velocities were recorded in 10 normal subjects. Fig. 2 shows the recording of the SVC flow velocity in a 16-year-old normal subject. From the top, it shows lead II of the electrocardiogram (ECG), the phonocardiogram at the 3rd left sternal border, the SVC flow velocity, and the right atrial pressure. At the bottom, the schema of the blood flow in the cardiac phase is shown. In the SVC flow velocity recording, the flow above the zero line represents forward flow to the right atrium (flow toward the catheter tip), and the flow below the zero line represents reversed flow to the right atrium (flow away from the catheter tip).

As shown in Fig. 2, the normal pattern of the SVC flow velocity was characterized by two distinct positive waves (S and D waves) and one nega-

![Image of Fig. 2 showing the SVC flow velocity curve and the schema illustrating blood flow in each cardiac phase.](image-url)
The notched point between the S and D waves is named O point. At the end of the diastole, there was a negative A wave caused by atrial contraction. About 10 to 20 msec after the Q wave of the ECG, there was a rapid rise and a round peak S wave. The peak of the S wave occurred around the middle of the ventricular systole and might be due to both atrial relaxation and downward movement of the tricuspid floor during ventricular contraction. The D wave occurred during ventricular diastole, perhaps caused by the rapid filling phase of the right ventricle.

In normal subjects, the peak S wave was greater than the D wave, the peak S wave being between 15 and 35 cm/sec. Although the IVC flow velocity was much influenced by respiration, the pattern and phase of the flow velocity was similar to that of the SVC flow velocity. For convenience, we adjusted our instruments so that forward flow to the right atrium would always be above the zero line, thereby inverting the IVC pattern to match the SVC pattern.

The pattern of the PV flow velocity

The PV flow velocity was recorded in 10 patients with ASD and 5 patients with other heart diseases. Fig. 3 shows the recording of the PV flow velocity in aortic insufficiency.
velocity (upper), and the SVC flow velocity (lower) in a patient with aortic insufficiency. In the PV flow velocity recording, the flow above the zero line represents forward flow to the left atrium (flow toward the catheter tip) and the flow below the zero line represents reversed flow to the left atrium (flow away from the catheter tip). As shown in Fig. 3, the patterns of the PV and SVC flow velocities were similar, having two distinct positive S and D waves. The peak S wave of the PV flow velocity, however, occurred in late systole or early diastole, in contrast to the peak S wave of the SVC flow velocity, which occurred in midsystole.

**SVC and IVC flow velocities in patients with and without ASD**

The SVC and IVC flow velocities were recorded in 20 patients with an

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**Fig. 4.** Venous flow velocity in atrial septal defect. The recordings of the superior vena cava flow velocity (upper), the inferior vena cava flow velocity (middle), and the pulmonary vein flow velocity (lower) in a 17-year-old girl with atrial septal defect. Right: the schema of the position of the Doppler flowmeter catheter tip.
uncomplicated secundum type of ASD. Fig. 4 shows the recording of the SVC, IVC, and PV flow velocities in a 17-year-old female in this group. The characteristic pattern of SVC and IVC flow velocities in these patients was a precipitous fall of the descending limb of the S wave, a slight negative O point and a diminished D wave. The peak systolic S wave occurred around midsystole, whereas the peak S wave of the PV flow velocity appeared near end systole or early diastole.

The SVC and IVC patterns among the 150 patients without ASD were varied, but nevertheless did display a midsystolic peak S wave, as did the patients with ASD.

The SVC and IVC flow velocities in patients with anomalous PV connection

Case 1. Anomalous PV connection to the SVC

Fig. 5 shows the flow velocities recorded at various positions in the SVC and IVC in Case 1. The SVC flow velocity (No. 3, upper right) had the peak of the S wave in late systole (nearly coinciding with the second heart sound), being quite similar to the normal PV flow velocity pattern. In contrast, the IVC flow velocity (No. 4, lower right) showed the normal pattern, in which the peak of the S wave occurred in midsystole. The flow velocity of the anomalous PV (No. 2, lower left) displayed a late systolic peak S wave,
similar to the pattern seen in normal PV flow velocity. SVC flow velocity at the peripheral portion of the junction of the anomalous PV (No. 1, upper left) also had a late systolic peak S wave.

In summary, a delayed peak S wave (the pattern of the PV flow velocity) was observed in the SVC flow velocity, while the IVC flow velocity showed a normal IVC flow velocity pattern in this patient.

Case 2. Total anomalous PV connection

Fig. 6 shows the SVC flow velocity in a patient with total anomalous PV connection. The peak of the S wave occurred in late systole or early diastole, resembling the pattern of the PV flow velocity, and was probably due to the flow of the total anomalous PV. The patient became dyspneic after this recording, so we could not record the IVC flow velocity.

Case 3. Partial anomalous PV connection to the IVC (Scimitar Syndrome)

Fig. 7 shows the recording of the SVC, IVC, and left PV flow velocities in a patient with Scimitar syndrome. The SVC flow velocity (upper) displayed a peak S wave around midsystole. The IVC flow velocity (middle) was similar to the mirror image pattern of the PV flow velocity (lower); the peak S waves in both recordings occurred in late systole or early diastole. We adjusted our instruments here, too, so that forward flow to right atrium would always be above the zero line. Anomalous PV flow velocity was thus recorded as a mirror image pattern of the usual PV flow velocity in the IVC flow velocity recording.

QSc, QOc, TDe intervals, and S/D, O/D ratios

Fig. 8 shows the results of various measurements of the SVC flow velocity.

![Fig. 6. Superior vena cava flow velocity in total anomalous pulmonary venous connection (Case 2). Left: lead II of the electrocardiogram, phonocardiogram of the third sternal border and superior vena cava flow velocity. Right: schema of the superior vena cava flow velocity. Dotted line represents the normal pattern of the superior vena cava flow velocity. Note the peak S wave of the superior vena cava flow velocity is much more delayed than the normal peak S wave.](image)
in 20 patients with uncomplicated ASD and corresponding data from Cases 1 and 2. The QS interval is the interval between the Q wave of the ECG and the peak S wave of the SVC flow velocity; the QO interval is the interval between the Q wave of the ECG and the O point of the SVC flow velocity; and the TD interval is the interval between the end of the T wave of the ECG and the peak D wave of the SVC flow velocity. Since these intervals are influenced by the cardiac cycle, each measurement was divided by $\sqrt{RR}$ of the ECG and named corrected QS (QSc), corrected QO (QOc) and corrected TD (TDc). The amplitude of the peak S wave divided by the amplitude of the D wave (S/D ratio) and the amplitude of the O point divided by the amplitude of the D wave (O/D ratio) were also calculated. The
shaded bars show the mean ± standard deviation in 10 normal subjects. As shown in Fig. 8, QSc is more markedly delayed (delayed peak S wave) in Cases 1 and 2 than in the 20 patients with uncomplicated ASD.

**DISCUSSION**

In the present study, the SVC and IVC flow velocities in normal subjects showed a biphasic pattern, having systolic S and diastolic D waves, the peak of the S wave occurring around midsystole. The patients with partial (Case 1) and total (Case 2) anomalous PV connection to the SVC, however, displayed a SVC flow velocity pattern similar to that of the PV, in which a delayed peak S wave occurred in late systole or early diastole. The IVC flow velocity of Case 1 showed the same pattern (no delayed peak S wave) as seen in ASD patients without an associated anomaly. In the patient with anomalous PV connection to the IVC (Case 3), the IVC flow velocity mirrored the pattern of the PV flow velocity, but the SVC flow velocity pattern was the same as that in ASD patients without an associated anomaly.²¹,³¹
Diagnosing an anomalous PV connection in ASD patients is sometimes difficult even after cardiac catheterization and angiocardiography. On the other hand, the SVC and IVC flow velocities can be recorded quite easily during routine cardiac catheterization and appear to be a useful diagnostic tool. Although the present study focused on only 3 patients, we believe the abnormally delayed peak S wave in the SVC or IVC pattern will prove helpful in determining the presence of an anomalous PV connection.

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REFERENCES