Anomalous Drainage of the Right Superior Vena Cava Into the Left Atrium

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A 22 year old man with asymptomatic hypoxemia was found to have a large right to left shunt due to a rare congenital anomaly: total drainage of the right superior vena cava into the left atrium. The anomaly was first suspected after radionuclide angiography was performed using technetium-99m macroaggregated albumin and was confirmed by cardiac catheterization. Contrast echocardiographic and surgical findings are discussed. Other reports on this anomaly are reviewed.

Although anomalies of the great veins are not uncommon, isolated drainage of the right superior vena cava into the left atrium is very rare (1–3). Park et al. (4) reported the first adult case of the right superior vena cava draining into the left atrium diagnosed by radionuclide studies. This is a rare congenital anomaly which causes a total right to left shunt of the venous blood returning through the superior vena cava. However, patients are generally asymptomatic because a greater volume of venous blood returns by way of the inferior vena cava, empties into the right atrium and is oxygenated normally. Another case with this anomaly that we encountered is described and other reports on this anomaly are reviewed.

Case History

A 22 year old black man presented to the emergency room of the Indiana University Medical Center after experiencing several grand mal seizures. He had had a chronic seizure disorder since resection of a brain abscess in 1978 at another institution. He had recently been noncompliant with his Dilantin therapy.

The patient was admitted to the hospital after arterial blood drawn in the emergency room demonstrated unexplained hypoxemia that was not corrected with supplemental oxygen. He was completely asymptomatic with regard to the cardiopulmonary system. Past history revealed that he enjoyed normal growth and development and was able to participate in strenuous sports activities without limitation. He experienced no cyanotic episodes, chest pain or syncope.

Physical examination revealed a muscular, well developed young black man. Blood pressure was 115/80 mm Hg and the pulse rate was 60 beats/min. There was a residual scar from the right temporal craniotomy. There was mild clubbing of the fingers but no cyanosis. Examination of the heart revealed a normal apical impulse, a grade 1/6 ejection murmur at the left sternal border and a soft third heart sound. Carotid artery pulses and jugular vein examinations were normal. The chest X-ray film revealed borderline cardiomegaly. The electrocardiogram revealed normal sinus rhythm. The P wave configuration and axis were normal. The QRS voltage, axis and configuration were also normal. ST segment elevation consistent with early repolarization was noted. The patient’s hemoglobin was 14.9 g/dl and his red blood cell count was 4.9 million/mm³.

Arterial blood gases on room air revealed a partial pressure of oxygen (PO₂) of 57 mm Hg and a partial pressure of carbon dioxide (PCO₂) of 42 mm Hg. With the patient breathing 100% oxygen by mask, the PO₂ increased to 81 mm Hg with a calculated venous admixture of 27 to 40%. A complete battery of pulmonary function tests was normal except for a mild restrictive lung disease pattern.

A lung scan was obtained to investigate the hypoxemia. Results of a xenon-133 ventilation study were normal.

Radionuclide angiography. A radionuclide angiogram, as a part of perfusion lung scan, was performed with 5 mCi of technetium-99m macroaggregated albumin injected into
a right arm vein. The study revealed flow of radioactivity through the right superior vena cava into the left heart chambers and then out the aorta (Fig. 1, upper panel). The pulmonary circulation was completely bypassed and the radionuclide particles were localized in the systemic organs; that is, brain, thyroid, myocardium, liver, spleen and kidneys (Fig. 2). These findings indicated that the right to left shunt was due to drainage of the right superior vena cava into the left atrium.

To investigate the drainage of the inferior vena cava, 20 mCi of technetium-99m pertechnetate was injected into a dorsal foot vein with the scintillation camera over the anterior chest. This study showed a normal flow of radioactivity through the inferior vena cava to the right heart chambers, through the pulmonary arteries, then to both lungs (Fig. 1, lower panel). There was no evidence of a persistent left superior vena cava when a third radionuclide angiogram was performed using 5 mCi of technetium-99m sulfur colloid injected into the left antecubital vein. The liver and spleen were in normal positions.

**Echocardiography.** An M-mode echocardiogram revealed a moderately dilated left ventricle with normal wall motion and a dilated left atrium. A two-dimensional echocardiogram using an injection of agitated saline solution into both arm veins revealed prompt appearance of the contrast effect in the left atrium and left ventricle (Fig. 3). After injection of saline solution into the right femoral vein, there was prompt appearance of the contrast effect in the right atrium and right ventricle without appearance in the left atrium or left ventricle.

**Cardiac catheterization.** The patient underwent cardiac catheterization by way of the right arm and right femoral vein, which confirmed the isolated drainage of the superior vena cava into the left atrium. There was no evidence of a concurrent left to right shunt at either the atrial, ventricular or great vessel level. There was normal take-off of right and left coronary arteries. Selective coronary angiography was not performed.

**Surgical findings and correction.** Subsequently, the patient agreed to undergo surgical correction of the congenital anomaly. At surgery the left atrium appeared to be rotated more cephalad and to the right. The superior vena cava drained entirely into the left atrium. An anomalous left superior vena cava was not present. The right upper and middle lobe pulmonary veins were partially conjoined with the distal end of the superior vena cava. The remaining right and left pulmonary veins drained into the left atrium. The inferior vena cava drained normally.

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Figure 1. Upper panel, Radionuclide angiogram in the anterior projection (with right arm vein injection) showing the anomalous drainage of the right superior vena cava into the left atrium. Note the flow of activity by way of the right subclavian vein and right superior vena cava (SVC) into the left atrium and left ventricle (LV), then out the aorta (A). The pulmonary circulation is completely bypassed. Lower panel, Radionuclide angiogram in the anterior projection (with foot vein injection) showing normal drainage of the inferior vena cava. Note the venous flow from the lower leg by way of the inferior vena cava (IVC) entering the right atrium and the right ventricle (RV) normally and leaving the pulmonary arteries (PA). The lungs are perfused normally.
Figure 2. Systemic distribution of technetium-99m macroaggregated activity after an arm vein injection. Note the activity in the brain, thyroid (left), myocardium, liver (middle), spleen and kidneys (right). There is no activity in the lungs. This unusual distribution of macroaggregated particles in the systemic organs is additional evidence for a total right to left shunt. A = anterior view and P = posterior view of the torso.

Figure 3. Apical four chamber two-dimensional echocardiogram. In the upper panel, all four chambers of the heart and the mitral and tricuspid valves are visualized. (Drop out of echoes in the area of the interatrial septum is due to technique and does not represent an interatrial communication.) In the lower panel after injection of agitated saline solution into an arm vein, a mass of echoes opacifies the left atrium and left ventricle (dark arrows). There is no ultrasound contrast effect in either the right atrium or the right ventricle. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

A vertical incision was made from the superior vena cava across the atroio caval junction, through the atrial septum, creating an atrial septal defect, and was extended onto the anterolateral wall of the right atrium. A patch of Goretex was used to create an intraatrial baffle diverting the superior vena caval blood to the right atrium. The superior margin of the patch was sutured to the posterior wall of the superior vena cava and the inferior margin was sutured to the caudal edge of the created atrial septal defect. A second Goretex patch was used to enlarge the cavo-right atrial junction as well as to prevent future constriction. The patient continued to have normal sinus rhythm postoperatively and is doing well.

Discussion

Isolated anomalous drainage of the right superior vena cava into the left atrium is very rare. A survey of the English language literature revealed a total of eight previously reported cases of this anomaly (3-10). There were six children and two adults (Table I). Patients with this anomaly had normal growth and development and the majority of them had no cardiac murmur.

Diagnosis. The hypoxemia is apparently well tolerated. Cyanosis may be present. If present, it is mild and may be difficult to recognize, especially in black people. Therefore, this congenital anomaly may escape clinical detection altogether as happened during our patient’s first hospitalization.

Cardiac catheterization performed through both arm and femoral veins has been the primary tool in the detection of this anomaly. More recently, however, noninvasive techniques have been used. Park et al. (4) described the use of...
Table 1. Findings in Nine Reported Cases of Isolated Right Superior Vena Cava Draining Into Left Atrium

<table>
<thead>
<tr>
<th>Age (yr) &amp; Sex</th>
<th>Reference (first author &amp; Year)</th>
<th>Growth &amp; Development</th>
<th>History, Physical Examination</th>
<th>Primary Diagnostic Tool</th>
<th>Surgery, Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10F</td>
<td>Wood (3), 1956</td>
<td>Normal</td>
<td>Cyanosis, clubbing</td>
<td>Cardiac cath</td>
<td>Not done</td>
</tr>
<tr>
<td>2F</td>
<td>Kirsch (5), 1961</td>
<td>Normal</td>
<td>Cyanosis, clubbing</td>
<td>Cardiac cath</td>
<td>+, well</td>
</tr>
<tr>
<td>3F</td>
<td>Braudo (6), 1968</td>
<td>Below 3rd percentile</td>
<td>Cyanosis, clubbing, murmur</td>
<td>Cardiac cath</td>
<td>+, well</td>
</tr>
<tr>
<td>7/12M</td>
<td>Vázquez-Pérez (8), 1979</td>
<td>Normal</td>
<td>Cyanosis, murmur</td>
<td>Cardiac cath</td>
<td>Not done</td>
</tr>
<tr>
<td>1F</td>
<td>Tomoe (10), 1980</td>
<td>Normal</td>
<td>Cyanosis, clubbing</td>
<td>Contrast echo</td>
<td>Not done</td>
</tr>
<tr>
<td>14M</td>
<td>Thivolle (9), 1980</td>
<td>Unknown</td>
<td>Brain abscess × 2, polycythemia</td>
<td>Radionuclide study</td>
<td>+, unknown</td>
</tr>
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| Adults      |                               |                     |                             |                         |                 |
| 34F         | Park (4), 1973                | Normal              | Dyspnea, cyanosis, no clubbing | Radionuclide study      | Not done        |
| 52M         | Ezekowtz (7), 1978            | Normal              | Cyanosis, clubbing, polycythemia | Radionuclide study      | +, died 3 wk postop |
| 22M         | Present study                 | Normal              | Brain abscess × 1, no cyanosis, +, clubbing | Radionuclide study      | +, well         |

+ Corrective surgery performed; cardiac cath = cardiac catheterization, contrast echo = contrast echocardiography, F = female; M = male; postop = postoperatively.

radionuclide studies for detection of this anomaly in an adult in 1973. Tomoe et al. (10) used contrast echocardiography in an infant in 1980. Both modalities are simple to perform, safe and readily available.

**Brain abscess.** Brain abscesses are known to occur frequently in patients with right to left shunt (11). When the brain abscess is nontraumatic in origin or occurs in patients without chronic middle ear, sinus or pulmonary infection, one should look for a right to left shunt. The brain abscess in our patient several years earlier was caused by a penicillin-sensitive anaerobic streptococcus, which is one of the normal oral flora and one of the most frequently isolated organisms in nontraumatic brain abscesses in adults. Our patient maintained good oral hygiene and had no dental work done before the development of his brain abscess. There was no apparent primary septic focus, which is found in 5 to 10% of reported brain abscesses (11). Surgical correction of the right to left shunt should certainly reduce the chance of developing a brain abscess or paradoxical emboli in patients with such an anomaly.

**Embyology and incidence.** The embryologic abnormality resulting in this anomaly is not entirely clear. Kirsch et al. (12) proposed that this anomaly may result from a malposition and aberrant development of the right horn of the sinus venosus with a relative leftward and cephalad movement of the aperture of the future superior vena cava. In our patient, the P waves had an entirely normal configuration and axis, indicating a relatively normal position of the sinoatrial node.

**Incidence.** The incidence of this anomaly is not known, but it appears to be very rare. Though a large number of patients undergo lung perfusion scans, only three adult cases including the present case have been reported in which the anomaly was demonstrated. In patients with this anomaly, the technetium-99m macroaggregated albumin lung scan should show the obvious right to left shunt and total lack of activity in the lungs.

**Surgical result.** The surgical procedure to correct the anomaly was well tolerated by our patient. He has continued to have normal sinus rhythm postoperatively despite the incision across the atrio caval junction. One potential postoperative complication is transient right heart failure due to sudden volume overload in the relatively hypoplastic right ventricle that may be present in young children with this anomaly (13).

**Effect on coronary circulation.** It has been proposed that chronic systemic hypoxemia may have beneficial effects on the coronary circulation by enhancing coronary artery dilation and neovascularization (14). In our patient, the coronary arteries were not selectively catheterized, but their take-off and proximal portions appeared normal after an aortic root dye injection, and there was no visible dilation of the coronary arteries at surgery. Surgical creation of a right to left shunt (15) in human subjects as a preventive or
therapeutic measure for coronary artery disease may be dif­
ficult to justify in view of the increased risk of brain abscess
as discussed.

We believe that evaluation of patients with paradoxical
emboli, nontraumatic brain abscess or unexplained hypox­
emia should include the noninvasive screening tests de­
scribed in this report to rule out congenital right to left shunt.

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