Superior Vena Caval Obstruction
After Mustard’s Operation: Detection
by Two-dimensional Contrast Echocardiography

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SUMMARY To assess superior vena caval (SVC) obstruction after the Mustard operation for transposition of the great arteries, we performed two-dimensional contrast echocardiography (2-D contrast echo) in 18 patients, ages 1–9 years. Sterile saline was injected into a peripheral scalp or arm vein while the junction of the inferior vena cava (IVC) and the systemic venous atrium (SVA) was imaged from the subcostal long-axis plane. The results of 2-D contrast echo were compared with those obtained at cardiac catheterization. In nine patients, contrast passed from the SVC to the SVA and the IVC remained free of contrast echoes. At catheterization, these patients had no SVC obstruction by angiography and minor SVC-SVA mean pressure differences (0–4 mm Hg). In five patients, contrast passed from the SVC into the IVC and within a few cardiac cycles, contrast from the IVC flowed in the IVC toward the SVA. At catheterization, these patients had partial SVC obstruction by angiography and SVC-SVA mean pressure differences of 9–13 mm Hg. In four patients, the SVA was filled by contrast arriving from the IVC by way of azygos-IVC collateral vessels. At catheterization, these patients had complete SVC obstruction and SVC-SVA mean pressure differences of 14–20 mm Hg. Two-dimensional contrast echocardiography is a simple, accurate, non-invasive method for detecting SVC obstruction after Mustard’s operation and allows differentiation of complete from partial SVC obstruction.

PATIENTS with superior vena caval (SVC) obstruction after Mustard’s operation may be asymptomatic or may develop facial edema, delayed closure of the fontanelles, widening of the cranial sutures, increasing head circumference, pleural effusions or chylothorax.1–11 Recently, the detection of SVC obstruction by transcutaneous range-gated Doppler ultrasound has been reported;12, 13 however, this Doppler technique has not been used to differentiate partial from complete SVC obstruction. Using two-dimensional contrast echocardiography (2-D contrast echo),14 we have developed a simple method to detect the presence of complete or partial SVC obstruction after the Mustard operation.

Materials and Methods
The study group included 18 patients, ages 7 months to 9 years, who had undergone both cardiac catheterization and 2-D contrast echo after the Mustard procedure. The interval between the Mustard operation and the postoperative cardiac catheterization ranged from 3 months to 5½ years. Contrast two-dimensional echocardiography was usually performed 24 hours before cardiac catheterization as part of the routine echocardiographic evaluation for baffle leaks. In two patients, however, the echocardiographic study was performed after the cardiac catheterization with knowledge of the catheterization results. In patient 11, the echocardiographic study was performed after the

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TABLE 1.  Clinical, Echocardiographic and Cardiac Catheterization Data in 18 Patients After Mustard's Procedure

<table>
<thead>
<tr>
<th>Pt</th>
<th>Sex</th>
<th>Age at operation</th>
<th>Clinical findings at time of echo</th>
<th>2-D contrast echo</th>
<th>Cardiac catheterization</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Age</td>
<td>SVC obstruction</td>
</tr>
<tr>
<td>1</td>
<td>M</td>
<td>2 y 6 mos</td>
<td>Asymptomatic</td>
<td>3 y</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>13 mos</td>
<td>Asymptomatic</td>
<td>14 mos</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>7 mos</td>
<td>1 head circumference, cyanosis</td>
<td>3 y</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>8 mos</td>
<td>Asymptomatic</td>
<td>9 mos</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>13 mos</td>
<td>Asymptomatic</td>
<td>3 y 6 mos</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>3 wks</td>
<td>CVA, left hemiparesis</td>
<td>2 y 3 mos</td>
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<tr>
<td>7</td>
<td>M</td>
<td>8 mos</td>
<td>Asymptomatic</td>
<td>20 mos</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>5 days</td>
<td>Cyanosis with crying</td>
<td>18 mos</td>
<td>None</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>3 mos</td>
<td>Asymptomatic</td>
<td>1 y</td>
<td>None</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>1 wk</td>
<td>Asymptomatic</td>
<td>2 y 8 mos</td>
<td>Partial</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>1 y 6 mos</td>
<td>Asymptomatic</td>
<td>6 y 9 mos</td>
<td>Partial</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>5 mos</td>
<td>Asymptomatic</td>
<td>2 y</td>
<td>Partial</td>
</tr>
<tr>
<td>13</td>
<td>M</td>
<td>2 y 6 mos</td>
<td>Asymptomatic</td>
<td>9 y</td>
<td>Partial</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>7 wks</td>
<td>1 head circumference</td>
<td>7 mos</td>
<td>Partial</td>
</tr>
<tr>
<td>15</td>
<td>F</td>
<td>5 wks</td>
<td>Asymptomatic</td>
<td>3 y</td>
<td>Complete</td>
</tr>
<tr>
<td>16</td>
<td>F</td>
<td>1 mo</td>
<td>Prominent upper body veins</td>
<td>4 y 6 mos</td>
<td>Complete</td>
</tr>
<tr>
<td>17</td>
<td>F</td>
<td>3 wks</td>
<td>1 head circumference,</td>
<td>16 mos</td>
<td>Complete</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>prominent upper body veins</td>
<td></td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>M</td>
<td>1 mo</td>
<td>1 head circumference,</td>
<td>18 mos</td>
<td>Complete</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>prominent upper body veins</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Bilateral ileofemoral vein thrombosis prevented venous catheterization.

Abbreviations: CVA = cerebrovascular accident; 2-D contrast echo = two-dimensional contrast echocardiography; SVC = superior vena caval; SVA = systemic venous atrium.

FIGURE 1.  The three echocardiographic contrast patterns in 18 patients after the Mustard operation.  (left)  A contrast injection is made into an arm or scalp vein while the transducer (Tr.) is applied in the subcostal long-axis plane to image the junction of the inferior vena cava with the systemic venous atrium. In nine patients without superior vena caval obstruction, the contrast echoes (black arrows) filled the systemic venous atrium from above. No contrast echoes were seen in the lower inferior vena cava.  (middle)  In five patients with partial superior vena caval obstruction, contrast echoes filled the systemic venous atrium from above. Subsequently, contrast echoes appeared in the lower inferior vena cava, flowing toward the systemic venous atrium. These contrast echoes arrived in the inferior vena cava by way of azygos-inferior vena cava collateral vessels.  (right) In four patients with total superior vena caval obstruction, the contrast echoes filled the systemic venous atrium entirely from below by way of azygos-inferior vena cava collateral vessels.
After the saline injection into an arm or scalp vein in nine patients, contrast echoes were seen immediately in the SVA (fig. 2). No contrast echoes were seen in the lower IVC. However, reflux of contrast echoes into the upper IVC and hepatic veins was seen during vigorous atrial contraction. At cardiac catheterization, these patients were found to have no SVC obstruction and minor SVC-SVA mean pressure differences of 0–4 mm Hg.

After the saline injection in five patients, contrast echoes were seen first in the systemic venous atrium (fig. 3). During subsequent cardiac cycles, contrast echoes were then seen in the lower IVC flowing toward the SVA. These contrast echoes reached the lower IVC by way of azygos-IVC collateral vessels. At cardiac catheterization, these five patients had partial SVC obstruction and SVC-SVA mean pressure differences of 7–13 mm Hg.

After the contrast injection in four patients, contrast echoes were seen first in the lower IVC flowing toward the SVA. In this group of patients, there was no opacification of the SVA before the arrival of contrast material in the lower IVC (fig. 4). In these patients, contrast echoes reached the inferior vena cava by way of azygos-IVC collateral vessels. At cardiac catheterization, these patients had complete SVC obstruction and SVC-SVA mean pressure differences ranging from 14–20 mm Hg. In patient 15, the SVC-SVA mean pressure difference was not obtained because of an additional total IVC obstruction. Therefore, right-heart catheterization was not performed in this patient and SVC obstruction was diagnosed by an angiogram in the superior vena cava.

Because of symptomatic SVC obstruction and increasing head circumference above the ninety-fifth percentile for age, surgery was performed to relieve SVC obstruction in patients 14, 17 and 18. After baffle revision, patient 17 underwent repeat 2-D contrast echo and angiography. There was no evidence of SVC obstruction by either test.

**Discussion**

The significance of SVC obstruction after Mustard's operation has not been fully determined. It is an indication for catheterization of the asymptomatic patient after the Mustard or Senning repair. Most patients with SVC obstruction remain asymptomatic, but some develop facial edema, dilated upper body veins, increasing head circumference or pleural effusions. Although SVC obstruction may occur in the immediate postoperative period,1–7 contraction of the intra-atrial baffle with time may result in late SVC obstruction.

**Figure 2.** Two-dimensional contrast echocardiogram in the subcostal long-axis plane from a patient after the Mustard operation. A = anterior; I = inferior; IVC = inferior vena cava; PVA = pulmonary venous atrium; RV = right ventricle; SVA = systemic venous atrium. In the second frame, the contrast echoes fill the SVA from above. In the third frame, the contrast echoes are refluxing into the upper IVC and hepatic veins. There are no contrast echoes in the IVC below the hepatic veins. In the bottom frame, contrast echoes are in the PVA due to a right-to-left baffle leak.
Two-dimensional contrast echocardiography is a noninvasive test that can be repeated serially to identify SVC obstruction in the early and late postoperative periods. It can also be used after baffle revision to determine if the obstruction has been relieved. In addition, right-to-left baffle leaks, which have occurred in over 90% of all of our patients who underwent the Mustard procedure, can be detected during 2-D contrast echo for SVC obstruction. In the postoperative evaluation of patients after the Mustard procedure, the detection of SVC obstruction before
cardiac catheterization may be important, especially if iliofemoral vein thrombosis precludes venous catheterization from the groin.

Because contrast refluxes from the SVA into the upper IVC during atrial contraction, contrast echoes within the upper IVC are not sufficient evidence of SVC obstruction. It is important to image the IVC below the hepatic veins so as not to mistakenly identify reflux into the upper IVC as azygos-IVC collateral flow.

Two-dimensional contrast echocardiography accurately distinguished between patients with partial SVC obstruction and those with complete SVC obstruction. In this regard, the technique offers an advantage over the range-gated Doppler ultrasound studies, which have not differentiated partial from complete SVC obstruction. We did not attempt to estimate the severity of partial SVC obstruction from the intensity of the IVC microcavitations for several reasons. The intensity of the contrast echoes is related to factors such as the adequacy of the contrast bolus and the volume of blood diluting the contrast passing into the azygous system. These factors may so disturb the contrast bolus effect that only the crudest estimate of the degree of obstruction is possible.

The SVC-SVA baffle junction may be imaged from the suprasternal notch and subcostal planes; however, our success rate for deriving diagnostic images of the SVC in these views is not adequate. The subcostal approach provides satisfactory images of the IVC in all patients and, we believe, with almost any type of two-dimensional echocardiographic system.

Two-dimensional contrast echocardiography provides an accurate and rapid method for the detection of SVC obstruction after Mustard’s procedure. It can also be used serially in the early and late postoperative periods to detect SVC obstruction and after baffle revision to detect the relief of obstruction.

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References

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