Two-dimensional and pulsed Doppler echocardiography in the postoperative evaluation of total anomalous pulmonary venous connection

JEFFREY F. SMALLHORN, M.D., P. BURROWS, M.D., G. WILSON, M.D., J. COLES, M.D., D. L. GILDAY, M.D., AND R. M. FREEDOM, M.D.

ABSTRACT The role of combined two-dimensional and pulsed Doppler echocardiography in the postoperative assessment of patients with total anomalous pulmonary venous connection was evaluated. Twenty-two cases with a median age of 9.5 weeks at the initial examination were evaluated. Serial ultrasonic examinations were performed throughout the study period. The ultrasound results were compared with chest radiographs obtained during the same period. Of the 22 patients, 16 had normal pulmonary venous flow profiles characterized by low-velocity laminar flow. Of this group 12 had persistent radiographic postoperative pulmonary edema that cleared in all by 4 months. Six patients with pulmonary venous obstruction were identified, the diagnosis being confirmed at catheterization or autopsy. The venous flow pattern in this group was uniformly high velocity and turbulent. It was possible to localize the site of obstruction in each case. Although pulmonary edema was present in each patient, the chest radiograph did not provide reliable information as to the exact site of obstruction. Combined two-dimensional and Doppler echocardiography is a useful adjunct in the postoperative evaluation of patients with total anomalous pulmonary venous connection.


TWO-DIMENSIONAL echocardiography is routinely used in the preoperative evaluation of patients with total anomalous pulmonary venous connection. The addition of pulsed Doppler echocardiography has provided further important preoperative information. Thus far, postoperative evaluation has depended on radiologic assessment and angiography in those with suspected residual venous obstruction.

More recently pulsed Doppler echocardiography has been used in the evaluation of normal pulmonary venous flow and in a selected group of patients with either congenital pulmonary vein stenosis or obstruction secondary to a Mustard or Senning procedure.

This study evaluates the role of combined two-dimensional and pulsed Doppler echocardiography in a group of patients after repair of total anomalous pulmonary venous connection.

From the Department of Pediatrics, Divisions of Cardiology, Radiology, Thoracic Surgery, and Pathology, The Hospital for Sick Children, Toronto, Ontario, Canada.

Address for correspondence: Jeffrey F. Smallhorn, Division of Cardiology, The Hospital for Sick Children, 555 University Ave., Toronto, Ontario, Canada M5G 1X8.

Received April 9, 1986; revision accepted April 2, 1987.

Materials and methods

The patients were drawn from the inpatient department at The Hospital for Sick Children, Toronto, between January 1983 and February 1986. A total of 22 were included, all of whom met the following criteria. All had undergone a preoperative study, serial postoperative evaluations during the study period, and serial chest radiographic examinations. Another seven patients seen during this period were excluded because they did not meet the above criteria; in particular, they were not serially reviewed at our institution. The serial studies were performed within 24 hr of surgery, at the time of discharge, and then within the first 6 months in all but one patient who was assessed at 8 months after surgery. The patients had an age range of 1 day to 2.5 years (median 9.5 weeks) at the time of their initial examination. The modes of drainage and associated lesions present are listed in table 1.

Echocardiographic evaluation. All patients were evaluated with either an Advanced Technology Laboratory Mechanical Sector Scanner or a Hewlett-Packard phased-array unit. Imaging was performed at a frequency of 7.5, 5, or 3 MHz, depending on the patient’s size. The Doppler interrogation was performed with the use of a transmitted frequency of 5 or 3 MHz, with a sample volume size of 2 mm.

The pulmonary veins, which usually enter the right atrium via a single orifice after surgical correction, were best assessed in an apical or subcostal four-chamber view. In the subcostal view the right-sided veins could be seen entering in close proximity to the atrial septum, while the left veins are situated directly opposite them (figure 1). A similar appearance was observed in the standard apical four-chamber view. These
TABLE 1
Preoperative diagnosis

<table>
<thead>
<tr>
<th>Total anomalous pulmonary venous connection</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intracardiac</td>
<td>9</td>
</tr>
<tr>
<td>Supracardiac</td>
<td>3</td>
</tr>
<tr>
<td>Intracardiac</td>
<td>4</td>
</tr>
<tr>
<td>Mixed</td>
<td></td>
</tr>
<tr>
<td>LUPV LVV</td>
<td>1</td>
</tr>
<tr>
<td>LLLPV + RPV CS</td>
<td>2</td>
</tr>
<tr>
<td>Complex lesions (asplenia and reduced pulmonary blood flow)</td>
<td>3</td>
</tr>
</tbody>
</table>

CS = coronary sinus; LLLPV = left lower lobe pulmonary vein; LUPV = left upper pulmonary vein; LVV = left vertical vein; RPV = right pulmonary vein.

views, along with a high precordial short-axis view, provided the optimal assessment of the anastomotic site. In the neonate and infant the individual pulmonary veins could also be visualized from the suprasternal position with the ultrasound beam directed inferoposteriorly parallel to the frontal plane such that the upper part of the ascending aorta was transected in its short axis and the right pulmonary artery in its long axis.

The echocardiographic findings were compared with normal pulmonary venous flow patterns obtained in our laboratory and also with previously published data. Hemodynamic confirmation of the presence or absence of obstruction was available in 11 patients, while autopsy information was obtained in three. The serial chest x-rays films of all patients were reviewed by the cardiac radiologist (P. B.) without prior knowledge of the presence or absence of pulmonary venous obstruction.

Definition of normal vs obstructed pulmonary venous flow. Normal pulmonary venous flow is characterized by a pattern that is low velocity, laminar, and biphasic or triphasic. Obstructed pulmonary venous flow characteristically produces a turbulent continuous pattern.

Results

Unobstructed pulmonary venous pathway. The flow pattern in the pulmonary veins and through the pulmonary venous confluence into the left or common atrium was triphasic or biphasic and laminar in 16 patients with no clinical or radiologic evidence of residual obstruction after surgery (figure 1). The mean peak velocity across the anastomosis site was 71 cm/sec (SD 19.6 cm/sec). Of this group, three patients had angiographic and one autopsy confirmation of unobstructed flow.

FIGURE 1. Normal flow proximal and distal to an anastomotic site. Top left, The sample volume is placed proximal to the site of anastomosis (indicated by the white arrows). Top right, The spectral trace revealing biphasic laminar flow. Bottom right, The sample of volume is placed just distal to the anastomotic site. Bottom left, The spectral trace with a slight increase in velocity, but with preservation of normal laminar phasic flow. LA = left atrium; PVC = pulmonary venous confluence.
## TABLE 2
Postoperative venous obstruction

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Mode of drainage</th>
<th>Obstruction at anastomosis*</th>
<th>Pulmonary vein obstruction*</th>
<th>Hemodynamics</th>
<th>Chest x-ray</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Infra cardiac</td>
<td>Yes</td>
<td>No</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Infra cardiac</td>
<td>Yes</td>
<td>Left</td>
<td>50/12(26)</td>
<td>6</td>
</tr>
<tr>
<td>3</td>
<td>Infra cardiac</td>
<td>No</td>
<td>Right and left mildly narrowed</td>
<td>30/18(20)</td>
<td>13</td>
</tr>
<tr>
<td>4</td>
<td>Infra cardiac</td>
<td>Yes</td>
<td>Right and left mildly narrowed</td>
<td>45/10(25)</td>
<td>9</td>
</tr>
<tr>
<td>5</td>
<td>Coronary sinus + LUL to LVV</td>
<td>Yes</td>
<td>RLL, LLL, RUL</td>
<td>100/50(65)</td>
<td>20</td>
</tr>
<tr>
<td>6</td>
<td>Right veins to LVV left veins infra cardiac</td>
<td>Yes (small confluence)</td>
<td>Left and right</td>
<td>ND</td>
<td>ND</td>
</tr>
</tbody>
</table>

LUL = left upper lobe; LLL = left lower lobe; LVV = left vertical vein; ND = not done; RUL = right upper lobe; RLL = right lower lobe.

*By angiography or discovered at autopsy.

**Values in parentheses are means.

**Associated pulmonary venous obstruction.** Pulmonary venous obstruction at the anastomotic site, the individual pulmonary veins, or both was identified by pulsed Doppler echocardiography in six patients (tables 2 and 3). In each of these six patients a typical high-velocity turbulent pattern was identified irrespective of the site of obstruction (figures 2A and 2B). Angiographic and/or autopsy confirmation of the presence and site of obstruction was available in all cases (table 2; figures 3A and 3B).

In only three patients in whom the obstruction was situated distally at the anastomotic site did two-dimen-

![FIGURE 2A. Series of pictures from a patient with early mild postoperative obstruction, as shown in the bottom left. Also note phasic flow within the pulmonary veins (bottom right). Some 2 weeks later (top left), the velocity has increased with evidence of turbulence. Top right, Nonphasic flow in the pulmonary veins. This corresponded with clinical deterioration.](http://circ.ahajournals.org/attachments/300/300CIRCULATION.pdf)
TABLE 3
Postoperative venous obstruction

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Two-dimensional Doppler</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Right</td>
</tr>
<tr>
<td>1</td>
<td>Stenosis at anastomosis</td>
</tr>
<tr>
<td>2</td>
<td>Normal</td>
</tr>
<tr>
<td>(also stenosis of anastomosis site)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Normal</td>
</tr>
<tr>
<td>4</td>
<td>↑ Velocity across LPV and RPV</td>
</tr>
<tr>
<td>5</td>
<td>Turbulent</td>
</tr>
<tr>
<td>6</td>
<td>Turbulence at site of anastomosis</td>
</tr>
</tbody>
</table>

LPV = left pulmonary vein; RPV = right pulmonary vein.

sional echocardiography alone provide a clue to the diagnosis (figure 3B). It was not possible to identify isolated vein obstruction by imaging alone in any patient. The mean peak velocity across the distal stenotic anastomatic site in those with obstruction at that level was 198 cm/s (figure 2A). In other patients in whom the anastomosis was anatomically large with the obstruction situated at the orifice of the pulmonary veins, calculation of a peak gradient was not possible due to the inability to image enough of the vein to ensure parallel Doppler interrogation.

Progressive pulmonary venous obstruction was identified in five patients, while in the sixth the stenosis was evident within the first 24 hr. In one patient (No. 4) (figures 2A and 2B) obstruction was noted 1 week after surgery and was then observed to progress during the second week, following which further surgery was undertaken. The obstruction in the remainder was variable, with detection at 1 month (patients 3 and 5), 5 months (patient 2), and 8 months (patient 1; figures 3A, 3B, 4, and 5).

Elevation of mean pulmonary arterial pressure was observed in each patient with postoperative obstruction, although significant elevation was only noted in patient 5. Since the studies were not simultaneous it was not possible to relate the wedge pressures to the Doppler gradients in those with obstruction at the anastomotic site. However, patients with higher wedge pressures had more severe obstruction noted angiographically. Chest radiographs did not allow differentiation of those with higher wedge pressures from those with lower values (compare patients 4 and 5). Similarly, unilateral changes in the chest radiographs were noted with bilateral elevated wedge pressures (patient 2).

Comparison of the chest x-ray and two-dimensional echocardiography/Doppler

Patients without postoperative obstruction. Ten of 13 patients with an isolated total anomalous pulmonary venous connection had radiologic pulmonary edema 1
FIGURE 3B. This montage shows the changes in pulmonary vein flow profile 5 months after discharge (patient 2, table 2). Obstruction occurred at the anastomotic site and left pulmonary vein. The upper right-hand panel is a left pulmonary artery wedge angiogram demonstrating significant obstruction of the left pulmonary veins. The upper-hand panel on the left is a main pulmonary artery angiogram from the same patient demonstrating dilated right pulmonary veins with no visible left pulmonary veins on the levophase. It was not possible to visualize the anastomotic site in this view. The middle left-hand panel is with the sample volume in the right pulmonary veins, while the right-hand image demonstrates sampling in the vicinity of the left-sided veins. The lower left-hand panel shows phasic right vein flow, while the right-hand panel reveals turbulence from the left-sided veins. Ao = aorta; D = diastole; ES = early systole; LA = left atrium; LS = last systole; MPA = main pulmonary artery; PVC = pulmonary venous confluence; RPV = right pulmonary vein.

FIGURE 4. This montage demonstrates obstructive right pulmonary vein flow from the patient in figure 3 some 2 months after the second surgical procedure. Autopsy confirmation is shown in figure 5. LA = left atrium; LV = left ventricle; RA = right atrium; RPV = right pulmonary vein; RV = right ventricle; SV = sample volume.

Stenosis at the anastomosis in patient 1 did not produce bilateral edema, while reduced flow to the affected lung was observed in a patient with an isolated left-sided obstruction (patient 2, table 2).

One patient with a complex intracardiac anatomy (table 1) had no postoperative pulmonary edema, while both of the others had edema that persisted for several months, despite a normal Doppler venous profile. Confirmation of the absence of obstruction at autopsy was available in one patient.

Discussion

Two-dimensional echocardiography alone provides
important anatomic information in the preoperative evaluation of total anomalous pulmonary venous connection.\(^1\) Doppler echocardiography has been used preoperatively,\(^4\) but has not been applied in a postoperative population. Direct evaluation of the anastomotic site alone may be difficult postoperatively because the connection is frequently elliptical in shape. Therefore, a dimension in one plane cannot be used to assess the absolute size. Similarly, the direct identification of venous stenosis is very difficult.

More recently pulsed Doppler echocardiography has been used to evaluate normal pulmonary venous flow, which is of low velocity, laminar, and mirrors left atrial pressure events, with peaks occurring during the X and Y descent.\(^8\) Although this type of flow was observed in 16 of our patients, absolute documentation of absent obstruction was only available in three because repeat angiocardiology is not routinely performed on asymptomatic cases. We can therefore only speculate that there was absent obstruction based on the Doppler studies. This method has now been extended to patients with abnormalities of venous flow, notably those with congenital pulmonary vein stenosis\(^9\) or who have undergone a Mustard or Senning procedure for complete transposition of the great arteries.\(^10\)

The pattern in these patients appears constant, and is one of turbulent high-velocity flow.

While improved preoperative and surgical techniques have reduced surgical mortality,\(^7\) stenosis at the anastomotic site still remains a major postoperative complication. While this usually occurs as a late complication, it may account for morbidity in the early postoperative period.\(^14\) Pulsed Doppler echocardiography provides the optimal noninvasive tool for its detection, since the anastomotic site may be sampled repeatedly with relative ease. Radiologic evidence of pulmonary edema may be misleading, since it can be observed in the unobstructed patient in the early postoperative period. Although correlation between hemodynamic and Doppler-derived gradients was not possible in this study, recent data from an animal preparation indicates that a close relationship between the two exists.\(^15\)

Associated pulmonary vein stenosis is rarely recog-
nized preoperatively, but may be seen postoperatively in isolation, or in association with stenosis at the anastomotic site. Pulmonary vein stenosis may be related to trauma at the time of surgery, but in patients with a wide anastomosis situated well away from venous orifices it is more difficult to explain. As observed in this series, progressive obstruction of the pulmonary veins may occur. If the majority of the veins are involved, this usually proves fatal. Since the veins from either lung usually enter each side of the left atrium via a single orifice after surgery, this is an optimal situation for Doppler interrogation (figure 6). One must recognize that one limitation is that turbulence within the orifice may arise from any lobe on that side. The other limitation is that complete obstruction of one segment cannot be detected. In this case the lung perfusion scan may be valuable because it outlines the segments of the lung involved. While this may be of value in the child with unobstructed pulmonary blood flow, caution must be observed in the patient with obstructed flow invariably seen in the setting of situs ambiguous. In this situation Doppler interrogation of the anastomotic site provides important information regarding the presence of venous obstruction. It is our experience thus far that even when pulmonary blood flow is significantly reduced, obstruction can be detected.6 Another problem encountered with the lung perfusion scan is the detection of obstruction at two levels: that at the individual veins and at the anastomotic site. In this situation a combination of two-dimensional and pulsed Doppler echocardiography with a lung perfusion scan can play an important role in the detection of obstruction at more than one level. How then should this invasive technique be included in the postoperative management of total anomalous pulmonary venous connection? Unobstructed Doppler-determined flow in the immediate postoperative period is reassuring in those cases with early pulmonary edema. A second study is of value before discharge, with another one at some point during the first 6 months, since most cases of obstruction develop early rather than late.

If high-velocity turbulent flow at the anastomotic site is detected then further surgery is indicated. At present, we perform an angiographic evaluation before surgery to provide more information regarding possi-
ble involvement of the individual veins. Thus far the indications for surgery in the presence of milder obstruction are less clear and will only be elucidated with further experience.

Combined two-dimensional and pulsed Doppler echocardiography is a valuable, simple technique for the postoperative evaluation of the patient undergoing surgery for total anomalous pulmonary venous connection. While chest radiographs provide some clues to the presence of obstruction, limitation with regard to site and severity must be appreciated.

References
Two-dimensional and pulsed Doppler echocardiography in the postoperative evaluation of total anomalous pulmonary venous connection.
J F Smallhorn, P Burrows, G Wilson, J Coles, D L Gilday and R M Freedom

Circulation. 1987;76:298-305
doi: 10.1161/01.CIR.76.2.298

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/76/2/298