Baffle Fenestration With Subsequent Transcatheter Closure

Modification of the Fontan Operation for Patients at Increased Risk

Nancy D. Bridges, MD, James E. Lock, MD, and Aldo R. Castaneda, MD

Ventricular dysfunction, elevated pulmonary vascular resistance, and residual distal pulmonary artery distortion contribute to early mortality after a Fontan operation; they may be transient or reversible. A baffle fenestration, allowing right-to-left shunting, maintains cardiac output and limits right atrial pressure. A baffle fenestration was surgically created at the time of a modified Fontan repair in 20 consecutive patients. Risk factors included pulmonary artery pressure of 18 mm Hg or more, end-diastolic pressure of 12 mm Hg or more, valvar regurgitation, pulmonary artery distortion, pulmonary vascular resistance of 2 Woods' units or more, ventricular outflow obstruction, and complex anatomy. Nineteen of 20 patients survived. After the operation, mean arterial oxygen saturation was 86%, mean right atrial pressure was 15 mm Hg, and mean duration of pleural effusions was 6 days. Twelve of 19 survivors tolerated early test occlusion and had permanent transcatheter umbrella closure. Four patients failed early test occlusion, with a significant decrease in venous O₂ saturation and a rise in central venous pressure, due to ventricular dysfunction, pulmonary artery distortion, or aortopulmonary collaterals. Three of four had successful late closure of the fenestration after correction of these abnormalities. (Circulation 1990;82:1681–1689)

The Fontan repair and its modifications can be performed in select patients with a reported survival of 83–100% and with a relatively low morbidity. However, in “higher risk” patients, reported survival decreases to 25–83%. The risk of death is greatest in the immediate postoperative period; death commonly occurs in the clinical setting of a low output state. Elevated right atrial pressure in the postoperative period is a predictor of mortality and morbidity.

Some of the phenomena that contribute to this early mortality may be transient; for example, elevated pulmonary vascular resistance or ventricular dysfunction may be related to cardiopulmonary bypass or to the hemodynamic consequences of the Fontan operation itself. Others may be reversible, such as ventricular dysfunction in a patient with chronic volume overload, or treatable, as in the case of residual distal pulmonary artery distortion. Elevated right atrial pressure reflects these conditions and in itself may contribute to postoperative effusions.

Based on these observations, we reasoned that a surgically created, temporary communication between the right and left atria should decrease operative mortality and morbidity in the presence of these conditions. Right-to-left shunting through this baffle fenestration would allow cardiac output to be maintained (at the expense of oxygenation) in the presence of conditions that hinder pulmonary blood flow and would also limit elevation of right atrial pressure. Our previous experience with transcatheter closure of postoperative atrial leaks and of native atrial septal defects using a double umbrella “clamshell” device indicated that transcatheter closure of a surgically created baffle fenestration should be accomplished without difficulty.

Methods

Patient Population

A modified Fontan repair with fenestration of the intra-atrial baffle was performed in 20 patients, who ranged in age from 9 months to 22 years (mean, 6.8±6.2 years). The anatomic diagnoses and previous surgeries are listed in Table 1. Most of these patients...
TABLE I. Patient Diagnoses and Previous Surgeries

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Diagnosis</th>
<th>Previous surgery</th>
<th>Size of hole (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9.5</td>
<td>DORV (S,D,D), inlet VSD,PS</td>
<td>None</td>
<td>9</td>
</tr>
<tr>
<td>2</td>
<td>7.8</td>
<td>TGA (S,D,L), superoinferior ventricles, VSD, PS</td>
<td>APS (2)</td>
<td>5</td>
</tr>
<tr>
<td>3</td>
<td>12.8</td>
<td>TGA (S,D,L), superoinferior ventricles, VSD, PS</td>
<td>APS</td>
<td>8</td>
</tr>
<tr>
<td>4</td>
<td>8.3</td>
<td>TGA (S,L,L), DILV</td>
<td>PAB</td>
<td>7</td>
</tr>
<tr>
<td>5</td>
<td>4.8</td>
<td>PA/IVS</td>
<td>RVOT patch, APS (2), R-CPA</td>
<td>8</td>
</tr>
<tr>
<td>6*</td>
<td>1.8</td>
<td>TGA (S,L,L), DILV</td>
<td>PAB, AscAo-MPA anastomosis, APS</td>
<td>6</td>
</tr>
<tr>
<td>7</td>
<td>4.9</td>
<td>TA, PA, PAPVC</td>
<td>APS (2)</td>
<td>6</td>
</tr>
<tr>
<td>8</td>
<td>9.8</td>
<td>DORV (S,D,D), PS, MS, hypoplastic LV</td>
<td>Atrial septectomy (2), APS</td>
<td>6</td>
</tr>
<tr>
<td>9</td>
<td>1.3</td>
<td>TA, PS</td>
<td>APS</td>
<td>6</td>
</tr>
<tr>
<td>10</td>
<td>2.7</td>
<td>TGA (S,D,A), DILV, coarctation</td>
<td>Atrial septectomy, AscAo-MPA anastomosis, B-CPA</td>
<td>9</td>
</tr>
<tr>
<td>11</td>
<td>2.9</td>
<td>HLHS, PAPVC</td>
<td>Stage I palliation, B-CPA</td>
<td>4.5</td>
</tr>
<tr>
<td>12</td>
<td>3.9</td>
<td>DORV (S,D,D), MS, PS, hypoplastic LV</td>
<td>None</td>
<td>5</td>
</tr>
<tr>
<td>13</td>
<td>11.1</td>
<td>TA, PA</td>
<td>APS</td>
<td>7</td>
</tr>
<tr>
<td>14</td>
<td>0.8</td>
<td>Ebstein's hypoplastic RV, PA</td>
<td>APS</td>
<td>4</td>
</tr>
<tr>
<td>15</td>
<td>2.4</td>
<td>TA, PS</td>
<td>APS</td>
<td>4</td>
</tr>
<tr>
<td>16</td>
<td>4.1</td>
<td>TGA (S,D,L), superoinferior ventricles, PS</td>
<td>APS, R-CPA</td>
<td>4</td>
</tr>
<tr>
<td>17</td>
<td>22.5</td>
<td>DORV (S,D,L), multiple VSDs, PS</td>
<td>None</td>
<td>4</td>
</tr>
<tr>
<td>18</td>
<td>0.7</td>
<td>DORV/DIRV (S,X,L), PS</td>
<td>APS</td>
<td>4</td>
</tr>
<tr>
<td>19</td>
<td>21.2</td>
<td>DORV (S,D,D), CCACV malaligned to the right</td>
<td>PAB</td>
<td>5</td>
</tr>
<tr>
<td>20</td>
<td>2.9</td>
<td>DORV (S,D,D), superoinferior ventricles, VSD, PS</td>
<td>APS</td>
<td>4</td>
</tr>
</tbody>
</table>

Letters in parentheses refer to the segmental anatomy of the heart—visceroatrial situs: S, solitus; I, inversus; A, ambiguous; ventricular loop: D, normal; L, inverted; X, undetermined; aortic-to-pulmonary relation: D, rightward; L, leftward; A, directly anterior; DORV, double-outlet right ventricle; VSD, ventricular septal defect; PS, pulmonary stenosis; APS, aortopulmonary shunt; TGA, transposition of the great arteries; DILV, double-inlet left ventricle; PAB, pulmonary artery band; PA, pulmonary atresia; IVS, intact ventricular septum; R-CPA, right cavopulmonary anastomosis; TA, tricuspid atresia; PAPVC, partial anomalous pulmonary venous connection; MS, mitral stenosis; LV, left ventricle; B-CPA, bidirectional cavopulmonary anastomosis; HLHS, hypoplastic left heart syndrome; DIRV, double-inlet right ventricle; CCACV, complete common atroventricular canal defect.

*Died on ninth postoperative day.

(16 of 20) had diagnoses other than tricuspid atresia and were considered to be at increased risk for a Fontan type of repair because of the presence of one or more of the following: pulmonary vascular resistance greater than 2 Woods' units (three of 14 patients in whom this value could be calculated), pulmonary artery pressure of 18 mm Hg or more (six patients), ventricular end-diastolic pressure of 12 mm Hg or more (six patients), pulmonary artery distortion (five patients, one with discontinuous pulmonary arteries), mild to moderate atroventricular valve regurgitation (five patients), systemic ventricular outflow obstruction (two patients), anomalous pulmonary venous connection requiring concurrent surgical correction (two patients), and qualitatively diminished systolic ventricular function (three patients). One of the 20 patients had none of these risk factors; however, he lives at an altitude of 4,500 feet, which we felt might place him at increased risk for chronic effusions.

Surgical Technique

An intra-atrial baffle (using polytetrafluoroethylene in 19 patients and pericardium in one) was constructed, as has been previously described, in conjunction with end-to-side anastomosis of the divided superior vena cava to the pulmonary artery or of the divided pulmonary artery to the superior vena cava.24 Once the baffle was sutured in place, a central hole was made, assuring that there would be room on all sides for the 8.5-mm arms of the 17-mm umbrella device that would be used for closure. Fenestration size was determined with consideration given to the size of the desired shunt, the patient's weight, and the technical requirements of transcatheter closure. We reasoned that the magnitude of the shunt would be determined by both the postoperative resistance in the pulmonary capillary bed and by the size of the fenestration. The umbrella device is delivered by means of an 11F sheath with an outer diameter of 4.0 mm, which must cross through the fenestration. The size of the hole ranged from 4 to 9 mm; in 16 of the 20 patients, the hole was between 4 and 6 mm in diameter. Table 1 lists the fenestration size for each patient.

Additional surgical procedures performed concurrently with the Fontan repair included the following: pulmonary artery reconstruction in five patients, with continuity of the pulmonary arteries restored in one; repair of partial anomalous pulmonary venous connection in two patients; and end-to-side pulmonary artery to ascending aorta anastomosis in one patient. Two patients required bilateral cavopulmonary anas-
tomoses because of absence of a connecting vein between the cavae.

Catheterization

Cardiac catheterization was performed after the operation, when patients no longer required drainage of pleural effusions. After hemodynamic and angiographic assessment, the fenestration was occluded with a balloon for 10 minutes. If this did not result in a marked increase in right atrial pressure or decrease in right atrial saturation, we permanently closed the fenestration with a 17-mm “clamshell” double-umbrella device. An angiogram was performed in the right atrium to assess adequacy of closure (Figure 1).

Results

Numerical results are given as mean±SD. The Wilcoxon rank-sum test is used as the test of significance.

Surgical Survival and Postoperative Course

Nineteen of the 20 patients (95%) survived. The single death was attributed to severe postoperative ventricular dysfunction, in a patient infected with human immunodeficiency virus. The modified Fontan repair was taken down to a bidirectional cavopulmonary shunt on the seventh postoperative day; the patient died 2 days later. In the 19 survivors, inotropic support was required for 2±1 days, and intubation and ventilation was provided for 2±1 days; patients remained in the cardiac intensive care unit for 4±2 days. The mean right atrial pressure on the first postoperative day was 15±4 mm Hg. One patient required a second operation for bleeding from chest wall vessels. None of the 19 survivors had low cardiac output as manifested by poor perfusion or renal or hepatic dysfunction.

Arterial oxygen saturations at extubation, in room air, ranged from 70% to 96% (mean, 86±7%). One patient with a 4-mm fenestration had no right-to-left shunt detectable by oximetry or by echocardiography. Two patients had marked cyanosis in the early postoperative period. In one, this was attributed to severe ventricular dysfunction; test occlusion of the fenestration resulted in increased arterial oxygen saturation, decreased mixed venous oxygen saturation, and an increase in right atrial pressure, which were felt to be incompatible with survival (Table 2). The other was found to have a systemic vein draining to the pulmonary venous atrium (see below).

Another patient developed episodic marked cyanosis associated with crying, starting on the fourth postoperative day. This child, who weighed 10 kg, had an unintentionally large (9-mm) fenestration. It was closed at catheterization on postoperative day five, with resolution of the cyanotic episodes.

In 17 of the 19 survivors, pleural effusions were of brief duration (5±3 days). Two of 19 patients had prolonged pleural effusions (18 and 31 days, respectively). One of these was the patient just described, who had early closure of the fenestration because of episodic cyanosis; this was the only patient in whom the fenestration was closed before resolution of pleural effusions. The other was a child with hypoplastic left heart syndrome, who had moderate tricuspid regurgitation and right ventricular dysfunction.

Results of Catheterization

Seventeen of 19 survivors have undergone postoperative cardiac catheterization. One patient, aged 23 years, sustained a transient brachial plexus injury at cardiac catheterization, presumably related to positioning during the procedure; there were no other catheterization-related complications.

Five patients were found to have sites of right-to-left shunting separate from and in addition to the baffle fenestration. These included leaks at the baffle margin in two patients (superior edge of the baffle, near the mouth of the right atrial appendage, in both patients, plus inferior margin in one), a vascular communication between the systemic and pulmonary venous atria in one patient, and systemic veins to the pulmonary venous atrium in two patients. The average arterial oxygen saturation in these patients at the time of cardiac catheterization was significantly lower than in the other survivors who were catheterized (72±5% versus 84±6%; p<0.05). Three patients had these leaks closed with umbrellas or coils at cardiac catheterization. One of the marginal baffle leaks required surgical closure, and in that patient, the baffle fenestration was closed at the same operation.

Patients Tolerating Test Occlusion

Eleven patients had transcatheter closure of the fenestration performed within 20 days of surgery (mean, 10±4 days). In each case, balloon occlusion of the baffle fenestration resulted in little if any increase in central venous pressure (mean increase, 1.6±3.4 mm Hg). The arterial oxygen saturation increased (mean increase, 8%), and the mixed venous oxygen saturation increased or was unchanged (mean increase, 2%). There was thus an increase in the arteriovenous difference in oxygen content, corresponding to a mean decrease in cardiac output of 21%. The fenestrations were permanently closed with a 17-mm clamshell device. One additional child tolerated test occlusion after the operation; however, because of his small size (he was 9 months old and weighed 7 kg at the time of surgery), he was sent home to grow with the fenestration open. He returned and had the fenestration closed 3 months later, having gained 1 kg.

Patients Failing Test Occlusion

Four patients failed test occlusion, due to a marked fall in cardiac output (31–56% decrease) or a marked rise in right atrial pressure (increase of 4–6 mm Hg) (see Table 3). The important finding in these patients was the fall in right atrial saturation with test occlusion. They were discharged without closure. Failure to tolerate test occlusion was attributed to residual distal pulmonary artery stenosis.
FIGURE 1. Upper panel: Angiogram showing contrast injection into the systemic venous atrium after a modified Fontan operation with fenestration of the atrial baffle. The wire and catheter pass through the fenestration, and dye crosses from the baffle space into the pulmonary venous atrium. Lower panel: Angiogram after transcatheter closure with a 17-mm "clamshell" double umbrella.
ventricular dysfunction, and aortopulmonary collaterals. Three of these four patients have returned for successful closure of the fenestrations after transcatheter pulmonary artery dilation (Figures 2 and 3), embolization of aortopulmonary collaterals, and improvement in ventricular function with time and medical management.

All of the 15 patients who have undergone transcatheter closure had appropriate placement of the device by angiography. Twelve had postclosure echocardiographic studies with color flow mapping, which were considered adequate for assessment of residual flow; of these, 10 had complete closure, and two patients had trivial residual leaks that were not felt to be clinically significant. Fourteen of the 15 had complete clinical closure, with saturations of 92–93%; one child with a small leak at the inferior baffle margin had a saturation of 89%.

**Discussion**

The definition and quantification of risk factors for patients undergoing a Fontan repair remains elusive. Most studies, but not all,23 report a better outcome in patients with tricuspid atresia than in those with other anatomic diagnoses. Adequate pulmonary artery dimension remains controversial,17,26–28 and methods of assessing ventricular function, especially in patients who do not have morphological left ventricles, remain qualitative for the most part. Thus, the design and assignment of strategies for reduction of risk is difficult. Nevertheless, there is experiential and theoretical support for the position that uncorrected pulmonary artery distortion or ventricular dysfunction contribute to early mortality after a Fontan repair.

In patients reviewed at our institution, no patient with uncorrected significant pulmonary artery distortion survived a Fontan repair.15 Surgical repair of distorted or discontinuous central pulmonary arteries may be performed in preparation for or at the time of a Fontan repair.15,17,29 In the former case, the patient is subjected to an additional thoracotomy and cardiopulmonary bypass, and adhesions may increase the difficulty of the subsequent Fontan repair. In the latter, inadequate relief of distortion may be fatal, and the additional bypass time required for pulmonary artery reconstruction may contribute to a transient but significant postoperative elevation in pulmonary vascular resistance.30–32 Stenoses that are beyond the hilum of the lung are not accessible to the surgeon. Residual pulmonary artery stenosis can be corrected by transcatheter balloon angioplasty after a Fontan procedure, although the procedure may be hazardous in the first 2 months after surgery.33

Ventricular hypertrophy is thought to increase operative mortality after a Fontan repair.4 Decreased ventricular contractility and increased ventricular muscle mass and dimension34,35 have been found in unrepaird patients with single left ventricles. Gewellig et al36 reported a significant increase in ventricular wall thickness, particularly during diastole, immediately after a Fontan procedure. These patients demonstrated normalization of left ventricular dimension, contractility, and mass-volume ratios over a period of months. These findings suggest that ventricular dysfunction is likely to be greatest in the early postoperative period. This early postoperative dysfunction may be exacerbated by the reversible diminution in ventricular function that follows cardiopulmonary bypass.32,37

Thus, a strategy that allows an adequate cardiac output in the presence of these conditions and that will not be detrimental to the patient over the long term once these conditions have resolved appears advantageous. The concept of allowing a right-to-left “popoff” at the atrial level during palliative or definitive surgery for congenital heart disease is not new. Edwards and Bargeron,38 in 1968, recommended delayed ligation of theazygous vein when performing a Glenn shunt in very young infants, in whom physiologically elevated pulmonary vascular resistance limited pulmonary blood flow after cavopulmonary anastomosis. Billingsley et al39 described creation of an “adjustable atrial septal defect” in seven patients undergoing biventricular repair of pulmonary atresia with intact ventricular septum; this approach has

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**Table 2. Result of Test Occlusion of Fenestration in Patient 6**

<table>
<thead>
<tr>
<th>Arterial saturation</th>
<th>Mixed venous saturation</th>
<th>Right atrial pressure (mm Hg)</th>
<th>Arteriovenous O₂ difference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Open</td>
<td>Closed</td>
<td>Open</td>
<td>Closed</td>
</tr>
<tr>
<td>76%</td>
<td>95%</td>
<td>29%</td>
<td>25%</td>
</tr>
</tbody>
</table>

**Table 3. Patients Who Failed Balloon Occlusion of Fenestration**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Venous pressure (mm Hg)</th>
<th>Venous saturation (%)</th>
<th>Arterial saturation (%)</th>
<th>Decrease in cardiac index (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Open</td>
<td>Closed</td>
<td>Open</td>
<td>Closed</td>
</tr>
<tr>
<td>5</td>
<td>16</td>
<td>17</td>
<td>52</td>
<td>29</td>
</tr>
<tr>
<td>9</td>
<td>14</td>
<td>14</td>
<td>72</td>
<td>60</td>
</tr>
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<td>36</td>
<td>27</td>
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<tr>
<td>19</td>
<td>14</td>
<td>20</td>
<td>57</td>
<td>NR</td>
</tr>
</tbody>
</table>

NR, not recorded.
FIGURE 2. Pulmonary angiogram after a modified Fontan operation with fenestration of the atrial baffle. There is left pulmonary artery stenosis just beyond the hilum. The fenestration is open, and some dye crosses into the pulmonary venous atrium. Test occlusion of the fenestration resulted in a 31% decrease in cardiac output. Upper panel: Anteroposterior projection. Lower panel: Lateral projection.
FIGURE 3. Pulmonary angiogram (same patient as in Figure 2) after balloon angioplasty of the left pulmonary artery stenosis. Occlusion of the fenestration after pulmonary artery dilation resulted in a decrease in cardiac output of 14%. The fenestration was permanently closed at the same catheterization. Upper panel: Anteroposterior projection. Lower panel: Lateral projection.
since been used in patients undergoing a Fontan type of repair.

Transcatheter closure of the surgically created fenestration offers several advantages. The most important of these is that it allows test closure (by means of balloon occlusion) of the fenestration in the cardiac catheterization laboratory before permanent (umbrella) closure. Patients in whom test occlusion is not tolerated as a result of a correctable or reversible problem (such as residual pulmonary artery stenosis, aortopulmonary collaterals, or ventricular dysfunction) can be sent home to recover from surgery with the fenestration open and may return electively for surgical or transcatheter treatment. The fenestration diminishes the risk of a low output state and its consequences in such patients. Patients who undergo a Fontan operation at a young age can be allowed to grow before transcatheter closure.

The number of patients in this series, and the manner of their selection, precludes any generalizations about the effectiveness of this procedure; however, we are encouraged by our early experience. We believe that use of a fenestrated atrial baffle with subsequent transcatheter closure may be advantageous in patients at increased risk for a Fontan repair, particularly those with distal pulmonary artery stenosis, ventricular dysfunction, or elevated pulmonary vascular resistance. Systematic use of this procedure in selected patients continues.

Acknowledgments

The authors would like to express their appreciation to Dr. John E. Mayer, who operated on one of the patients in this series; to Dr. John F. Keane and Dr. Stanton B. Perry, who performed a number of the transcatheter closures; and to Dr. Michael D. Freed, who was the cardiologist for many of these patients.

References

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