Fontan Procedure for Tricuspid Atresia

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SUMMARY A Fontan procedure has been performed on 29 patients for tricuspid valvular atresia. The age range was 8 months to 33 years (median 10 years), the pulmonary vascular resistance ranged from 1.8–6.1 units \( \cdot \) m\(^2\) (mean 3.3 units \( \cdot \) m\(^2\)), and the mean pulmonary arterial pressure ranged from 13–45 mm Hg (mean 21 mm Hg). Twenty-nine previous operations had been performed in 23 patients. Fourteen other associated cardiopulmonary anomalies were present in 12 patients. There were four hospital deaths (13.8%), all in patients with complicating features. Among the last 22 consecutive patients who have undergone operation, one died (4.5%). Complete atrioventricular block necessitated pacemaker implantation in one patient. No late deaths occurred. Of the 19 patients followed 3 months or more from the time of operation, eight have no restriction of exercise capacity, nine have only mild restriction, and two have a poor result. The Fontan approach to tricuspid atresia has several theoretical advantages over previously used shunts or pulmonary artery banding, the operative mortality in patients who have suitable anatomy and hemodynamics is low, and the results have been good.

TRICUSPID VALVULAR ATRESIA, a congenital cardiac defect often occurring in association with other complex cardiovascular malformations, carries a very poor prognosis, although certain types of tricuspid atresia have benefited from various shunting and banding procedures. Recently, a more physiologic repair — anastomosis of the right atrium to either the rudimentary right ventricle or the pulmonary arteries (Fontan procedure) — has been reported from various centers. This report describes the entire Mayo Clinic experience with the Fontan procedure for tricuspid atresia, involving 29 patients. The term “tricuspid atresia” can be confusing unless it is further defined, because the term has been used to describe hearts in which either the right or the left atrioventricular valve may be atretic. In this report, therefore, the term is used to describe the congenital cardiac malformation in which there is situs solitus of the atria and absence of the right atrioventricular connection, with a main ventricle of left ventricular type and a rudimentary ventricle of right ventricular type (outlet chamber). The ventriculoarterial connections may be of any type. We agree with the observation that most cases of tricuspid atresia are examples of univentricular heart with outlet chamber and atresia of the right atrioventricular valve. In this manuscript, the term “rudimentary right ventricle” will be used to describe this outlet chamber.

Patient Profile

The first Fontan procedure for tricuspid atresia at our institution was performed in October 1973, and this consecutive series of 29 patients extends from then through March 1979. The patients were classified ac-
According to a modification of Keith et al.\textsuperscript{11} (table 1). In type I the great arteries are normally related, in type II the aorta is anterior and to the right of the pulmonary artery, and in type III the aorta is anterior and to the left of the pulmonary artery. Each type is further subdivided according to the adequacy of the flow pathway via the pulmonary artery: type a, pulmonary valvular or subvalvular atresia, or a closed ventricular septal defect; type b, decreased pulmonary blood flow due to relative stenosis at the level of the valve, subvalvular area, or ventricular septal defect; and type c, no severe obstruction between the left ventricle and the pulmonary artery.

The largest group was type I and the largest subgroup had pulmonary stenosis with reduced pulmonary blood flow (type Ib). Of the seven patients with type c anatomy (six type Ic and one IIc), five had been previously banded. Of the 17 patients who had type b anatomy with reduced pulmonary blood flow, two had valvular pulmonary stenosis, two had subvalvular pulmonary stenosis and 13 had a small ventricular septal defect.

Operation was indicated in each patient because of worsening cyanosis or progressive exercise intolerance or both. Nine patients were taking diuretics. One patient had classic angina. There were 15 males and 14 females; the age range was 8 months to 33 years (median 10 years). The mean hemoglobin concentration was 18.5 g/dl (range 12.7–22.8 g/dl). The total pulmonary vascular resistance ranged from 1.8–6.1 units \cdot m\textsuperscript{2} (mean 3.3 units \cdot m\textsuperscript{2}). The mean pulmonary arterial pressure ranged from 13–45 mm Hg (mean 21 mm Hg). The left ventricular end-diastolic pressure was elevated above 15 mm Hg in six patients, the highest being 23 mm Hg. All patients had normal sinus rhythm and left-axis deviation preoperatively. One patient had first-degree atrioven-

<table>
<thead>
<tr>
<th>TABLE 1. Fontan Operation for Tricuspid Atresia: Anatomic Types*</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I Normally related great arteries</td>
<td>25</td>
</tr>
<tr>
<td>Ia</td>
<td>4</td>
</tr>
<tr>
<td>Ib</td>
<td>15</td>
</tr>
<tr>
<td>Ic</td>
<td>6</td>
</tr>
<tr>
<td>Type II Aorta anterior and to the right</td>
<td>2</td>
</tr>
<tr>
<td>IIa (DORV)\textsuperscript{†}</td>
<td>1</td>
</tr>
<tr>
<td>IIc</td>
<td>1</td>
</tr>
<tr>
<td>Type III Aorta anterior and to the left</td>
<td>2</td>
</tr>
<tr>
<td>IIIb</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>29</td>
</tr>
</tbody>
</table>

*Modified from Keith et al.\textsuperscript{11} Type a = absence of direct connection from ventricle to pulmonary artery; type b = decreased pulmonary blood flow; type c = no obstruction from ventricle to pulmonary artery. Abbreviation: DORV = double outlet right ventricle.

Tricuspid block. The cardiothoracic ratio was greater than 0.5 in 16 patients, and five had radiographic evidence of increased pulmonary blood flow.

Twenty-nine previous operations had been performed in 23 patients (table 2). Twenty-two of these were procedures designed to increase pulmonary blood flow, either with systemic arterial-to-pulmonary arterial or superior vena caval-to-pulmonary arterial shunts.

All but two patients had a ventricular septal defect at the time of operation. These two patients both had closure of an earlier ventricular septal defect documented by cardiac catheterization. Fourteen other associated cardiopulmonary anomalies were present in 12 patients (table 3).

**Operative Technique**

After median sternotomy and lysis of pericardial adhesions, cardiopulmonary bypass was established with selective cannulation of both cavae. In patients with a prior Glenn shunt, venous return from the superior vena cava was retrieved via a left atrial vent placed through the right superior pulmonary vein. When bilateral superior vena cavae were present, the left cava was temporarily snared; if the pressure in it rose above 30 mm Hg, it was also cannulated.\textsuperscript{12} Prior systemic-to-pulmonary arterial shunts were controlled by standard techniques. In patients with a previous Glenn anastomosis, this arrangement was not dis-

<table>
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<tr>
<th>TABLE 2. Fontan Operation for Tricuspid Atresia: Prior Surgical Procedures (83 Patients)</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blalock-Taussig shunt</td>
<td>8</td>
</tr>
<tr>
<td>Glenn shunt</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary artery banding</td>
<td>5</td>
</tr>
<tr>
<td>Waterston shunt</td>
<td>3</td>
</tr>
<tr>
<td>Aortopulmonary graft</td>
<td>3</td>
</tr>
<tr>
<td>Potts shunt</td>
<td>1</td>
</tr>
<tr>
<td>Ligation of patent ductus arteriosus</td>
<td>1</td>
</tr>
<tr>
<td>Exploratory cardiotomy</td>
<td>1</td>
</tr>
<tr>
<td>Total procedures</td>
<td>29</td>
</tr>
</tbody>
</table>

TABLE 3. Fontan Operation for Tricuspid Atresia: Associated Anomalies (12 Patients)

<table>
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<th>No. of patients</th>
</tr>
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<tbody>
<tr>
<td>Bilateral superior vena cavae</td>
<td>7</td>
</tr>
<tr>
<td>Mitral valve incompetence</td>
<td>2</td>
</tr>
<tr>
<td>Anomalous pulmonary venous drainage</td>
<td>2</td>
</tr>
<tr>
<td>Common atrium</td>
<td>1</td>
</tr>
<tr>
<td>Coronary arteriovenous fistulas</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary arteriovenous fistulas</td>
<td>1</td>
</tr>
<tr>
<td>Total procedures</td>
<td>14</td>
</tr>
</tbody>
</table>
turbed and the repair was made to the left pulmonary artery only. Pulmonary artery bands were excised, and the pulmonary arteries were repaired by end-to-end anastomosis.\textsuperscript{13}

Mean bypass time for the series was 125 minutes (range 66–190 minutes). Myocardial protection was achieved with moderate hypothermia (24–28°C) and multiple limited periods of ischemic arrest (19 patients) or with cold potassium cardioplegic arrest (8–12°C) (10 patients). A right atriotomy was performed and the atrial septal defect was closed with a patch in 21 patients, including the one with common atrium and the two in whom partial anomalous pulmonary venous drainage required redirection of pulmonary venous flow to the left atrium. In eight patients the atrial septal defect was small and was closed by direct suture. In five early patients a glutaraldehyde-preserved porcine xenograft was placed in or over the orifice of the inferior vena cava.

The rudimentary right ventricle or pulmonary artery was opened and the pulmonary valve was inspected. Two patients had a hypoplastic pulmonary valve; in one the valve was excised and in the other it was sutured closed before insertion of a right atrial-to-pulmonary arterial conduit. Resection of subpulmonary stenosis was required in two patients. A ventricular septal defect was closed either by direct suture (13 patients) or with a patch (nine patients). The ventricular septal defect was not closed in patients with pulmonary atresia or those in whom the proximal main pulmonary artery was sutured or ligated before the establishment of a communication between the right atrium and the main pulmonary artery.

In 20 patients a conduit was used to establish continuity between the right atrium and the rudimentary right ventricle, and in three patients conduits were placed from the right atrium to pulmonary artery. Of these 23 conduits, 19 contained a porcine xenograft valve, which ranged from 20–30 mm in diameter. In four patients, a Dacron-tube graft without a valve was used (18–25 mm in diameter). Direct anastomosis of the right atrium to either the rudimentary right ventricle or the pulmonary arteries was accomplished in six recent patients. In three the anastomosis was made to the rudimentary right ventricle, each requiring pericardial patch augmentation, and in three the anastomosis was made to a pulmonary artery, one of which required pericardial patch augmentation.\textsuperscript{14, 15} These anastomoses, and the proximal anastomoses when conduits were used, were usually accomplished with the heart beating during rewarming.

In four patients additional surgical procedures were required:

Mitr al valve replacement with a porcine xenograft was done in a 33-year-old man who had type Ib anatomy, a previous Potts anastomosis (at age 3 years), severe mitral regurgitation, poor left ventricular function (left ventricular end-diastolic pressure 18 mm Hg), a total pulmonary vascular resistance of 6.1 units \cdot m\textsuperscript{2} (pulmonary arteriolar resistance 2.1 units \cdot m\textsuperscript{2}), and a pulmonary artery pressure of 41/28 mm Hg (mean 34 mm Hg). Mitr al annuloplasty was performed in a 3-year-old child with poor left ventricular function (left ventricular end-diastolic pressure 22 mm Hg) and type IIC anatomy who had previously undergone pulmonary artery banding at the age of 6 weeks.

Transplantation of the atrial septum was done in a 9-year-old patient with pulmonary arteriovenous fistulas in the right lung subsequent to a Glenn procedure, in order to direct unsaturated blood from the right lower lobe to the right atrium.

Suture ligation of multiple coronary artery fistulas from the right coronary artery to the right atrium was done in a 31-year-old patient with type Ib anatomy.

Results

Hospital Mortality

Four hospital deaths (13.8\%) occurred, all in patients with complicating features. Since June 1976, 22 consecutive patients have undergone operation and there was one death (4.5\%). One 31-year-old woman (type Ib anatomy) had a stenotic right pulmonary artery secondary to a previously performed Blalock shunt. This patient also required suture ligation of multiple coronary artery fistulas. A permanent pacemaker was required for sick sinus syndrome on the fiftieth postoperative day; she died of uremia on the ninety-sixth postoperative day despite dialysis. The genesis of the uremia was obscure, but poor renal function was present preoperatively, as evidenced by an elevation of serum creatinine to 3.5 mg/dl after preoperative cardiac angiography.

The 9-year-old patient (type Ib anatomy) with pulmonary arteriovenous fistulas in the right lung died in combined respiratory and renal failure on the twelfth postoperative day despite tracheostomy and dialysis.

A third patient, age 22 years (type Ia), had previous bilateral Blalock procedures that had resulted in moderate pulmonary vascular obstructive disease, which caused high right atrial pressures and low cardiac output postoperatively. She required prolonged ventilation and a tracheostomy and died on the fifty-first postoperative day. Autopsy revealed grade 2 plexo-ogenic pulmonary arterioopathy (Heath and Edwards).

The pulmonary artery was not entered on preoperative cardiac catheterization in any of the above three patients, but each had a measured mean pulmonary artery pressure of less than 15 mm Hg at the time of operation.

The fourth patient, age 8 months (type Ic), who had a pulmonary blood flow of 8.9 l/min/m\textsuperscript{2} and a pulmonary artery pressure of 64/30 mm Hg (mean 45 mm Hg), died of low cardiac output on the second postoperative day despite inotropic support and infusion of tolazoline (Priscoline) into the pulmonary artery. This was the only death in the last 22 operations.

Three of the four hospital deaths occurred in the group of five patients who received inferior vena caval valves.
Early Complications

Maintenance of adequate cardiac output required right atrial filling pressures above a mean value of 12 mm Hg in all patients and above a mean of 20 mm Hg in seven patients; of these seven, four died and one suffered low cardiac output syndrome for 4 days but survived. Among these seven patients were the two who required additional mitral valve repair or replacement; both survived. Pleural effusions, more often on the right, were seen in most patients, but only eight required aspiration or the insertion of drainage tubes. Most patients required diuretic therapy and strict salt restriction during their hospital convalescence, and this was routinely advised for 6 weeks after hospital dismissal.

Nine patients experienced renal failure (defined as an elevation of serum creatinine level to twice the preoperative value) in association with low cardiac output and elevated mean right atrial pressures greater than 20 mm Hg. Dialysis was necessary in three patients, two of whom died; one additional patient died without dialysis.

Three patients required reexploration for control of postoperative bleeding. Another patient required reoperation on the third postoperative day when the atrial septal defect, which had been sutured, reopened and allowed significant right-to-left shunting with cyanosis.

Three patients had serious wound or bloodstream infections; two survived. All three suffered low cardiac output and renal failure before the development of their infections.

One of the patients in whom subvalvular pulmonary stenosis required resection experienced complete atrioventricular block, necessitating the insertion of a permanent pacemaker. The patient who required mitral annuloplasty experienced transient atrioventricular block for 24 hours. The duration of hospital stay for the operative survivors varied from 8–50 days (median 13 days).

Late Results

The clinical status of all operative survivors was documented, with a mean follow-up of 15 months (range 1–55 months). There were no late deaths. In four patients signs of right-heart failure secondary to conduit stenosis or occlusion have developed; one of these patients had a concomitant recurrent atrial septal defect. Three patients have undergone reoperation and replacement of the conduit, all successfully; one of these had an episode of severe gastroenteritis and dehydration 10 months after operation and suffered occlusion of the conduit, but pulmonary blood flow by way of a prior Glenn anastomosis maintained life until reoperation. The remaining two patients had slow progression of their symptoms, and at reoperation a thick peel was observed in each conduit which significantly narrowed the lumen. Two of the three explanted conduits contained porcine valves, and the third was a nonvalved tube graft. Two of the three patients reoperated on for conduit stenosis had an inferior vena caval valve placed at the initial procedure; in both cases the valve was found to be nonfunctional and degenerated. The valves were removed and not replaced, and the patients have subsequently done well.

The fourth patient, in whom signs of right-heart failure secondary to conduit obstruction developed, was restudied 16 months after operation for symptoms of decreasing exercise tolerance and supraventricular tachycardia. He was found to have an occluded conduit, but a large fistulous communication between the right atrium and the outlet chamber allowed systemic venous return to reach the lungs. His clinical status is now stable, and he has not yet required reoperation.

Six patients are convalescing within 3 months of operation and have been excluded from the following analysis of late results. Eight patients have no restriction of exercise capacity. Five of these are taking no medications and the remainder continue to take digitalis glycosides or diuretic agents or both. Nine additional patients have only mildly restricted exercise capacity; most continue to require salt restriction or diuretic therapy with or without concomitant digitalis glycosides. Two patients have a poor result, continue to retain fluid despite the use of potent diuretics (including aldosterone antagonists) and digitalis glycoside therapy, and are severely restricted in activity. Both had type Ib anatomy and a prior Glenn shunt.

Discussion

Since the first report of successful creation of right atrial-to-pulmonary arterial communication for the treatment of tricuspid atresia,4 this procedure has gained increasing application for alleviating cyanosis and improving the clinical status of selected patients with this anomaly. Our surgical techniques have steadily evolved as a result of our experience with these 29 patients and with 17 other "modified Fontan" operations performed for univentricular heart and other complicated congenital lesions.15 We currently prefer to cannulate each cava directly with a right-angle cannula in order to limit incisions in the right atrium that might be detrimental to subsequent right atrial function. Similarly, the right atriotomy is made chiefly in the right atrial appendage and is extended only as far into the right atrial free wall as is required to avoid any constriction of the atrial anastomosis. Since the appendage is mobile, adequate exposure for closure of the atrial septal defect is afforded by this approach.

We currently protect the myocardium by aortic root injection of cold, potassium-containing solution, which additionally improves surgical exposure by relaxing the heart. The atrial septal defect is closed with a patch, for direct suture may result in recurrence of the defect with its associated right-to-left shunting,15,16 as in two of our patients. The right atrial-to-pulmonary arterial or rudimentary right ventricular connection is often made with the aortic clamp removed and the heart beating.

In all patients in whom a valved or nonvalved con-
duit was used, a size was chosen as large as was deemed technically possible. In spite of this policy, four grafts or conduits in this series became stenotic or occluded 10 months to 4.7 years after their insertion. In two of three reoperated cases there was a thick, white, fibrotic peel within the graft, which narrowed the lumen. This peel formation has been more florid in this setting than in similar conduits inserted between the right ventricle and the pulmonary artery or between the left ventricle and the aorta. For this reason and because of uncertainty as to the value or necessity for a valved communication, we now prefer, whenever possible, to perform a direct anastomosis. If the right atrial appendage cannot easily reach the rudimentary right ventricle or the main pulmonary artery (or proximal right or left pulmonary artery), it may be extended and enlarged with a pericardial patch, as was done in four recent patients in the present series. A widely open, obstruction-free communication must be established.

In our early experience with the Fontan procedure, glutaraldehyde-preserved porcine heterografts (Hancock) were used in or over the orifice of the inferior vena cava. Three of the five patients who received such valves died in the hospital. Both of the survivors came to reoperation for other reasons, and in both, the valves were found to be nonfunctional and partially stenotic. Because of accelerated xenograft valve deterioration in this position, such valves are not used. We have not detected any deleterious early or long-term effects from omitting caval valves. It is not yet known whether the valves in conduits inserted in the atrio pulmonary position will develop a similar problem of accelerated deterioration.

In contrast to a significant incidence of atrioventricular block (five of 16 patients) after the modified Fontan procedure for univentricular heart, atrioventricular block is uncommon after the Fontan operation for tricuspid atresia. In this series, the only instance of permanent complete atrioventricular block occurred in a patient who had concomitant resection of infundibular pulmonary stenosis. Temporary block has been noted in our experience from distortion of the ventricular septal defect when it has been closed by direct sutures. Moreover, although the conduction bundle courses along the posterior, inferior border of the defect in type I tricuspid atresia, its course in type II and type III is variable and often anomalous. We therefore prefer to have the heart beating when the ventricular septal defect is closed, preferably with a patch, to ensure that sinus rhythm is preserved.

The criteria for selection of the ideal candidate for atrio pulmonary anastomosis have been well defined by Drs. A. Choussat and F. Fontan and their associates. Their 10 criteria include age of 4–15 years, sinus rhythm, normal drainage of venae cavae, right atrium of normal volume, mean pulmonary artery pressure 15 mm Hg or less, pulmonary resistance less than 4 units m–2, pulmonary artery–aortic diameter ratio 0.75 or more, ejection fraction of ventricle 0.60 or more, no mitral incompetence and no impairing effects of a previous shunt. Operation for patients who meet these criteria can be accomplished at low risk and with good late results. Eleven of our patients met these criteria and all survived; 10 are doing well on follow-up.

Patients who do not meet these criteria, and for whom no other palliative procedure can be expected to offer significant improvement, offer a dilemma. We have therefore accepted for operation selected patients who have significant, progressive symptoms but who do not meet these criteria, if there is a reasonable expectation that they will survive operation and experience a good result. Eighteen patients did not meet these criteria, and 14 of them survived (78%). Of this group, 10 had one contraindication to operation, six had two contraindications, one had three and one had four. This last patient survived and enjoys an excellent late result. He was 33 years old and had a mean pulmonary artery pressure of 34 mm Hg, a pulmonary resistance of 6.1 units m–2, and severe mitral insufficiency. His repair included mitral valve replacement.

An elevated pulmonary arterial pressure does not necessarily indicate inoperability, as long as the pulmonary vascular resistance is acceptable. Nine of the patients in this series had mean pulmonary arterial pressures of more than 15 mm Hg (range 16–45 mm Hg); eight survived operation, and seven are doing well on follow-up. In two patients, ages 8 months and 2 years, who had type c anatomy with increased pulmonary blood flow and without prior banding of the pulmonary artery, the mean pulmonary arterial pressure was 45 and 39 mm Hg, and the pulmonary resistance was 4.2 and 2.7 units m–2, respectively. The younger patient died in low cardiac output, but the older survived and is doing well 2 years later.

Clearly, a fixed pulmonary resistance (or more accurately, pulmonary arteriolar resistance) significantly above normal levels is an absolute contraindication to operation. Exactly what this level is, however, remains uncertain. Sometimes it is not possible to calculate pulmonary resistance preoperatively because the pulmonary artery cannot be entered during catheterization. Such patients have either severe valvular or subvalvular stenosis or a restrictive ventricular septal defect, and hence their chance of having significant pulmonary arteriolar obstructive disease is minimal. Also, in circumstances of decreased pulmonary blood flow, particularly in poly cicthemic patients, calculation of pulmonary resistance is misleading, and an elevated value may be obtained in a patient who actually has normal pulmonary arterioles and would, therefore, be a good candidate for the Fontan procedure.

A normal left ventricular end-diastolic pressure is preferable for patients undergoing the Fontan operation, for this decreases the driving pressure (right atrial pressure) needed to propel venous return through the lungs. An elevated end-diastolic pressure, however, does not necessarily preclude survival or a good result. Five of our six patients who had end-diastolic pressures above normal (range 16–23 mm Hg) survived operation and four have a good late
result. Separation of the circulatory pathways by this procedure, as well as closure of systemic-pulmonary shunts, reduces the previously increased volume load on the compromised left ventricle and may thereby decrease its filling pressure and improve its function.

Mitrval insufficiency also need not be a contraindication to operation if the insufficiency is repaired at the same time. The two patients with severe mitral insufficiency in this series are both doing well on follow-up; one had mitral valve replacement with a gluteraldehyde-preserved porcine prosthesis and the other underwent a mitral annuloplasty.

Previous pulmonary arterial banding was not associated with increased surgical mortality; all five such patients survived operation.

Age has been a factor in operative mortality. The overall mortality was four of 29 patients (14%). There was one death (age 8 months) in four patients (25%) younger than age 4 years. Among seven patients age 16 years or older, two died (29%). Only one of 18 patients (6%) in the ideal age range of 4-15 years died at operation.

Few patients have been followed up long enough after the Fontan procedure to permit evaluation of their long-term results, and the late effects of chronic elevation of venous pressure are not known. The operative mortality in patients with tricuspid atresia who have suitable anatomy and hemodynamics is low, and the early results are good. The value of this procedure compared with the previously used shunts or pulmonary artery banding will depend primarily on the quality and duration of the patient's improvement compared with that after other palliative procedures. The Fontan approach has the theoretical advantage, however, of providing a normal systemic arterial oxygen tension while also maintaining a normal left ventricular volume load — a favorable dual effect not offered by any previous palliative procedure. Additionally, it reduces the risk of systemic embolization which is present with other palliative operations that allow continued right-to-left shunting. The Fontan procedure is also effective in alleviating cyanosis and in improving the clinical well-being of selected patients who have had previous shunts but whose condition is again worsening because of increasing hypoxemia or deteriorating left ventricular function, or both, and for whom no other palliative procedure can be expected to offer significant improvement.

References
Fontan procedure for tricuspid atresia.
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