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The Fontan Procedure for Tricuspid Atresia: Early and Late Results of a 25-Year Experience With 216 Patients

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OBJECTIVES We assessed the operative and late mortality and the present clinical status of 216 patients with tricuspid atresia who had a nonfenestrated Fontan procedure performed at the Mayo Clinic in the 25-year period 1973 to 1998.

BACKGROUND The Fontan operation eliminates the systemic hypoxemia and ventricular volume overload characteristic of prior forms of palliation. However, it originally did so at the cost of systemic venous and right atrial hypertension, and the long-term effects of this "price" were unknown when the procedure was initially proposed.

METHODS We reviewed the clinical records of the 216 patients retrospectively. These were arbitrarily grouped into early (1973 through 1980), middle (1981 through 1987) and late (1988 through 1997) surgical eras. Patient outcome was also analyzed according to age at surgery. Operative and late mortality rates were determined and present clinical status was ascertained in 167 of 171 surviving patients.

RESULTS Overall survival was 79%. Operative mortality steadily declined and was 2% (one of 58 patients) during the most recent decade. Late survival also continues to improve. Age at operation had no effect on operative mortality, and late mortality was significantly increased only in patients who were operated on at age 18 years or older. Eighty-nine percent of surviving patients are currently in New York Heart Association class I or II.

CONCLUSIONS The initial 25-year experience with the nonfenestrated Fontan procedure for tricuspid atresia has been gratifying, with most survivors now leading lives of good quality into adulthood. These results justify continued application of this procedure for children born with tricuspid atresia. (J Am Coll Cardiol 2001;37:933–9) © 2001 by the American College of Cardiology

The first nonfenestrated Fontan operation at the Mayo Clinic was performed in March 1973 on an adult with tricuspid atresia, and through January 1, 1998, a total of 216 nonfenestrated Fontan procedures had been carried out for patients with tricuspid atresia; 182 of these patients had normally related great arteries and 34 had associated transposition of the great arteries.

Early and late results from this series of 216 patients with tricuspid atresia were reviewed. The current status of surviving patients was ascertained by direct examination, chart review, questionnaire or telephone contact. Of the 171 patients still alive, present clinical status could not be accurately determined in only four, all of whom live outside the U.S. However, all four of these patients were known to be alive no more than 12 months previously. Follow-up of survivors now ranges from 1 to 24 years, with a median of 13 years.

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METHODS

Era of operation. We arbitrarily divided the 25-year experience into an early period, 1973 through 1980; a middle period, 1981 through 1987; and the most recent 10-year experience, 1988 through 1997. These groups comprised 54, 104 and 58 patients, respectively.

Age at operation and prior palliation. The patients' ages at which the Fontan procedure was performed are listed in Table 1. Because the vast majority of patients were beyond early childhood, all but 17 had undergone at least one prior palliative cardiovascular operation (Table 1). The frequency and type of previous palliative operations are listed in Table 2.

Modifications of surgical technique. Modifications in surgical technique for the Fontan operation have evolved with experience. In the early cases, the atrial septal defect was closed by direct suture or patch. Five patients, all operated on before 1978, had a porcine glutaraldehyde-preserved heterograft valve placed at the inferior vena cava-right atrial junction, a practice that was abandoned when it was learned that these valves degenerate and become nonfunctional and often obstructed. The type of connection used to direct systemic venous blood to the pulmonary arteries (PAs) in these patients is listed in Table 2. The use of valved conduits was also characteristic of the early experience; all four of the right atrium-to-PA valved

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Abbreviations and Acronyms

LV = left ventricular
NYHA = New York Heart Association
PA = pulmonary artery
RA = right atrium
RV = right ventricle, right ventricular

conduits and 16 of the 27 right atrium-to-hypoplastic right ventricle valved conduits were placed before 1981. In the middle period, the method of choice for directing systemic venous blood to the PAs was direct anastomosis of the right atrium or right atrial appendage to the right or main PA. Beginning in early 1989 and continuing to the present, alternative approaches have included a lateral tunnel combined with either an atriopulmonary or a bidirectional cavopulmonary anastomosis, or an extracardiac nonvalved conduit combined with a bidirectional cavopulmonary anastomosis.

Concomitant surgical procedures. Additional procedures carried out at the time of the Fontan procedure included mitral valve repair in six patients and mitral valve replacement in six additional patients, all of whom had moderate or severe mitral valve incompetence. In three of the valve replacements, a heterograft valve (Hancock) was inserted; in the others a mechanical valve was used. Twenty-six patients underwent varying degrees of PA reconstruction, usually to alleviate narrowing and distortion caused by a prior shunt. Two patients, one with a prior Waterston shunt and one with a prior Potts shunt performed during early infancy, had iatrogenic complete occlusion of the left PA with some secondary hyperplasia of the right PA. In these two patients, the Fontan procedure was carried out to the single right PA.

The cumulative probability of patient survival was estimated as a function of time since surgery by the Kaplan-Meier method. Ninety-five percent confidence levels were calculated for patient survival at five-year intervals and plotted on the survival curves. The effects of risk factors, such as era of surgery and patient age group, on cumulative patient survival were assessed with log-rank tests.

Table 1. Age at Operation and Previous Palliative Operations

Age at Operation (yr)	No. of Patients	No. of Previous Cardiovascular Surgical Procedures	No. of Patients
<2	9	0	17
2–3	27	1	118
4–6	53	2	64
7–10	44	3	16
11–15	37	4	1
16–19	21	Total	216
20–30	18		
31–40	6		
>40	1		
Total	216*		

*Range 7 months to 42 years; median 8 years.

RESULTS

Operative and late mortality. Figure 1 is a Kaplan-Meier survival curve for the entire group of 216 patients. Operative, late and overall mortality rates for the patients operated on during the early, middle and most recent periods are shown in Table 3. There has been a steady, significant reduction in operative mortality, from 17% in the early period to 9% in the middle period, and 2% during the most recent decade (p value <0.01, exact Wilcoxon rank sum test). Late survival also continues to improve significantly as demonstrated in Figure 2, where Kaplan-Meier survival curves demonstrate continued survival rates for the 197 operative survivors during the three periods.

Causes of late death. Twenty-six late deaths occurred, ranging from four months to 18 years after operation (median 8 years). Seven deaths were sudden and unexpected, presumably a result of dysrhythmia, inasmuch as postmortem examination showed no other cause of death. Three of these patients had a history of post-Fontan rhythm problems and were receiving antidysrhythmic medication at the time of death. The four other patients did not have postoperative disturbances in rhythm. Four late deaths were attributed to progressive myocardial deterioration and failure. Two of these four patients were adults, ages 25 and 34 years at the time of their Fontan operation, who died 18 and 12 years, respectively, after operation, at ages 43 and 46 years. Three patients died at the time of reoperation: one for mitral valve replacement, one for resection of subaortic obstruction, and one at the time of attempted cardiac transplantation at another institution. Three late deaths were believed to be thromboembolic in nature. The deaths in these three patients, all of whom had postmortem examinations, were attributed to a large pulmonary embolus in one, thrombosis of a right atrial to right ventricular (RV) Hancock conduit in another and Budd-Chiari syndrome caused by hepatic vein thrombosis in the third. Three other late deaths were due to the debilitating effects of chronic protein-losing enteropathy. These three deaths occurred six, 13 and 15 years after operation, and the time from recognition of the protein-losing enteropathy until death was two, five, and six years, respectively.

Of the six remaining late deaths, two were accidental (a motorcycle and an automobile accident). Two relatively early late deaths, four and six months postoperatively, were attributed to persistent pleural effusions in one case and to chronic respiratory insufficiency in the second. The causes of the remaining two late deaths, in which no postmortem examination was performed, could not be conclusively determined.

Effect of operative age on survival. The effect of age at the time of the Fontan operation on early and late survival was also analyzed (Fig. 3). There was no statistically significant difference in operative mortality. Although there is a trend toward improved survival in patients who had their Fontan operation at younger ages, it does not reach statistical

Table 2. Type of Previous Palliation and Systemic Venous-to-Pulmonary Connection

Type of Previous Palliative Procedure	No. of Patients	Type of Connection*	No. of Patients
Blalock-Taussig (classic or modified)	141	RA to RV valved conduit	27
Waterston	44	RA to PA valved conduit	4
Classic Glenn	39	RA to RV direct anastomosis	35
Pulmonary artery banding	30	RA to PA direct anastomosis (with or without lateral tunnel)	146
Potts	15	Total cavopulmonary connection	2
Blalock-Hanlon	7	Extracardiac nonvalved conduit	2
Central shunt	9	Total	216
Bidirectional cavopulmonary anastomosis	6		
Other	7		
Total	298		

*RA = right atrium; RV = right ventricle; PA = pulmonary artery.

significance. Only in patients who had their Fontan procedure as adults age 18 or older is a clear pattern of significantly decreased late survival beginning to emerge. This pattern does not reach statistical significance until more than 15 years after the operation.

Incidence of reoperation in current survivors. One hundred seventy-one patients survived. In this group, 24 patients have had 26 reoperations, two patients having had two separate reoperations. Six reoperations involved replacement of the valved conduit with a new valved conduit; these occurred in the 1970s and early 1980s before the use of such conduits was abandoned. Nine reoperations were performed for removal of a valved conduit and conversion to an atriopulmonary anastomosis. Two reoperations, both in the 1970s, were for closure of a recurrent atrial septal defect. Three operations were pericardiectomy procedures carried out because of recurrent pericardial effusion. Two reoperations were successful cardiac transplantations in patients who were doing poorly because of gradual myocardial deterioration. Two patients had conversion of their original atriopulmonary anastomosis to a lateral tunnel type of Fontan procedure. The remaining two reoperations were: 1) a repeat pulmonary artery angioplasty in a patient who experienced recurrent left PA stenosis in the era before balloon angioplasty and stenting were available, and 2) resection of subaortic stenosis in a patient originally having had tricuspid atresia and associated transposition of the great arteries.

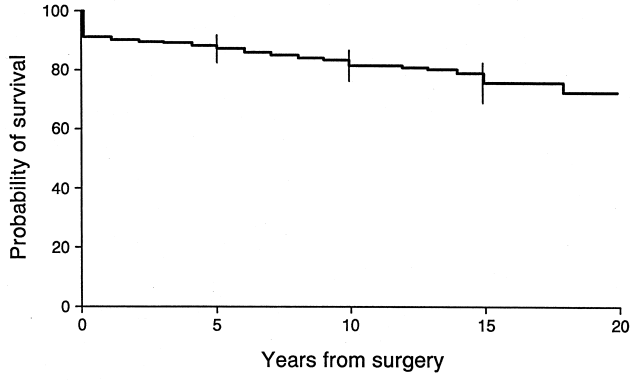

Figure 1. Kaplan-Meier curve for cumulative survival of the total 216 patients. The median postoperative follow-up period is 13 years and 79% of the patients survive.

Additional procedures for the treatment of dysrhythmias. Among the 171 surviving patients, 11 required additional procedures to treat symptomatic dysrhythmias. Eight had insertion of a permanent pacemaker, four because of bradycardia and four because of paroxysmal tachycardia. Three patients had catheter ablation of an accessory pathway or ectopic focus to eliminate paroxysmal atrial tachycardia.

Current clinical status of survivors. The length of postoperative follow-up and the present ages of surviving patients are shown in Table 4. We could ascertain the current clinical status in 167 patients. One hundred forty-nine patients, 89% of those responding, were in New York Heart Association (NYHA) class I or II and were either full-time students or were employed full time or capable of full-time work. Eighteen patients (11%) were judged to be in NYHA class III or IV, with significant symptoms or disability. The problems that these 18 patients were experiencing included recurrent and refractory atrial dysrhythmias, protein-losing enteropathy, congestive heart failure due to deterioration of left ventricular (LV) function and, in one patient, sequelae from a post-Fontan stroke.

DISCUSSION

Fontan performed the first successful right heart bypass on a patient with tricuspid atresia in 1969 (1). It was designed to completely separate the systemic and pulmonary circulations and thereby to eliminate the systemic arterial hypoxemia and LV volume overload that are the hallmarks of this condition. It was recognized at the onset, however, that the

Table 3. Operative and Late Mortality by Era of Operation

Years of Operation	No. of Patients	Deaths (no.)		Deaths (%)	
		Operative	Late	Operative	Overall
1973–1980	54	9	14	17	43
1981–1987	104	9	10	9	18
1988–1997	58	1	2	2	5
Total	216	19	26	9	21

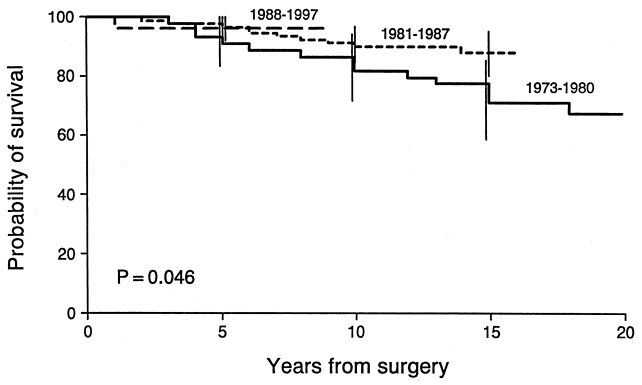

Figure 2. Kaplan-Meier curves for cumulative continued survival of the 197 operative survivors divided into era of surgery: 1973 through 1980 (**solid line**), 1981 through 1987 (**dotted line**), 1988 through 1997 (**dashed line**). There is a statistically significant improvement in continued survival among patients operated on more recently.

elevated systemic venous pressures inevitably present after the Fontan procedure—necessary to propel the blood through the lungs in the absence of a "booster pump" on the right side of the circulation—might lead to late complications in surgical survivors. Also, functional limitations would likely still exist in these patients, particularly for physical activities that required a substantial increase in cardiac output.

More than 30 years have passed since Fontan's original surgical procedure, and it has undergone many technical modifications, an evolution that still continues. This Mayo Clinic series encompasses 25 years and many of the surgical survivors have reached adulthood. It has been 10 years since we last reported our early and late results with the Fontan procedure for tricuspid atresia (2). We are therefore now in a better position to assess what has been accomplished by this novel surgical approach and to make recommendations concerning its continued application.

Table 4. Length of Follow-up and Present Age of 171 Surviving Patients

Length of Follow-up (yr)	No. of Patients	Present Age (yr)	No. of Patients
0–5	5	0–10	17
6–10	50	11–20	57
11–15	54	21–30	60
16–20	52	31–40	30
21–25	10	41–50	6
		>50	1
Total	171*	Total	171†

*Range, 1–24 years; median, 13 years. †Range, 6–58 years; median, 22 years.

Selection criteria for the Fontan procedure. Choussat and Fontan and other colleagues (3) originally listed "ten anatomic and physiologic commandments" that they believed must be met before this new operative approach could be successfully applied. Over time, it has become clear that the most important criteria involve the size of the PAs, the level of pulmonary arteriolar resistance and the status of systolic and diastolic function of the patient's single functional ventricle. As experience with the procedure increased, institutions with a large and ongoing patient population have established and published modifications of the original criteria in these areas, and well-recognized guidelines for patient selection now exist (2,4).

Evolution of surgical techniques. Fontan advocated placing caval valves and using a valved right atrium-to-hypoplastic RV conduit in his surgical patients with tricuspid atresia. It subsequently became clear that the caval valves do not function well and often deteriorate rapidly and that a hypoplastic RV was not necessary to propel the systemic venous blood into the lungs. An experimental model in the late 1980s (5) demonstrated that flow turbulence and kinetic energy losses are reduced when pulsation in a valveless chamber is eliminated and flow pathways are streamlined

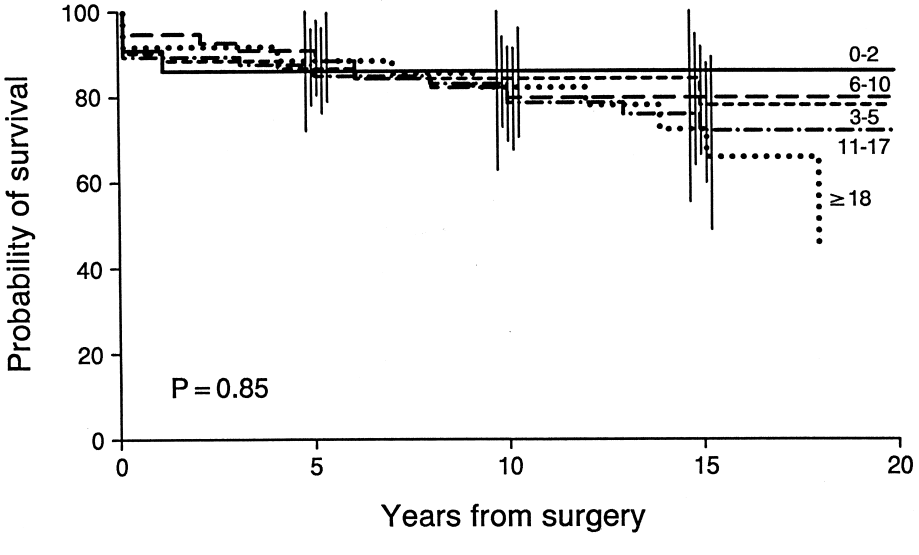

Figure 3. Kaplan-Meier curves for cumulative survival of the 216 patients according to age at the Fontan procedure. There is no significant difference in operative mortality. There is a trend toward improved continued survival in patients operated on at younger ages, but it does not reach statistical significance in patients operated on before age 18 years.

maximally with the elimination of corners. This study proposed total cavopulmonary connections with exclusion of most of the right atrium (RA), a concept that, it was thought, might reduce the incidence of postoperative atrial dysrhythmias as well as improve hemodynamics. In recent years, this concept has gained wide favor, and today in most major centers the inferior vena caval flow is routed by a lateral tunnel baffle within the right atrium to either a bidirectional cavopulmonary anastomosis (6) or an atriopulmonary connection, or via an entirely extracardiac pathway (7), with the medial aspect of this pathway located along the lateral border of the right atrium. There are still two controversial questions. One is whether to "fenestrate" (8) the inferior vena cava pathway. The other is whether it is necessary to "stage" the Fontan procedure by performing a bidirectional superior vena cava-to-PA anastomosis as a preliminary operation before completing the Fontan circulation by directing inferior vena caval blood directly to the PAs as a "second stage" (9). Some institutions (10) have favored these approaches in an attempt to decrease operative mortality. At the Mayo Clinic, however, we continue to favor the nonfenestrated, nonstaged approach in low-risk surgical candidates. Our series had 39 patients who had received a prior classic Glenn anastomosis and therefore could be considered staged. The early and late mortality in this subgroup did not differ significantly from the results in the remaining 177 nonstaged patients. We reserve fenestration and/or staging for patients whose operative risk is somewhat increased as a result of compromised ventricular function, borderline-sized PAs or mildly elevated pulmonary arteriolar resistance, or for patients who may require concomitant atrioventricular valve repair or replacement or subaortic resection at the time of their Fontan procedure.

Steady significant decrease in operative mortality. With this approach, the operative mortality for the nonfenestrated Fontan procedure for tricuspid atresia over the past decade has been 2% (one death in 58 patients). The steadily decreasing surgical mortality exhibited over the initial 25-year experience has resulted from better patient selection, improved surgical techniques and advances in immediate postoperative intensive care management. In reviewing surgical deaths from patients operated on during the 1970s and early 1980s, it is clear that a number of these patients—nearly all of whom had significant symptoms and for whom there were no other therapeutic options in an era before cardiac transplantation was well established—would not be accepted as Fontan candidates today.

Age at operation today. Now that the Fontan procedure is being performed electively in early childhood, usually at ages 2 to 4 years, the group reported here is clearly unique and will not be duplicated, because many of these patients were adolescents or even adults when they had their Fontan procedure. Follow-up to date indicates that good early and late results can be achieved in properly selected patients up through at least the adolescent years. However, one could reasonably expect that the operative and long-term late results obtained in the preschool-aged children now undergoing the procedure will continue to improve. Although the results obtained in this unique older group are generally good, many of these patients endured years of systemic hypoxemia and volume overload, with their potential detrimental long-term effects on ventricular function, before their Fontan procedure.

Postoperative cardiac dysrhythmia. Because the Fontan procedure as originally performed involved multiple suture lines within the atrium and exposed the entire RA to increased pressure and wall tension after completion, it was recognized that postoperative atrial dysrhythmias might be a concern. Follow-up studies (11–13) have established that as many as 10% to 20% of Fontan patients may have paroxysmal atrial tachycardia or atrial flutter-fibrillation. At the recent follow-up of this series, eight patients were receiving antidysrhythmic medication. Five patients were very well controlled on their medical program whereas three continued to have periodic episodes of symptomatic atrial dysrhythmias. Although the rhythm disturbances are usually amenable to medical therapy, in occasional instances they may be refractory to drug therapy and require more aggressive treatment, such as overdrive pacing or a surgical intervention like the maze procedure (14). However, the evolution of newer surgical techniques for the Fontan operation such as the "extracardiac Fontan," in which the right atrium does not have multiple suture lines and also does not have large portions of its wall exposed to increased wall tension, offers promise for a reduced incidence of troublesome atrial dysrhythmias in future operative survivors (15).

Potential for late thromboembolic complications. The potential for right heart thromboembolism in the postoperative Fontan patient is of concern because of the nonpulsatile nature of the circulation and the decreased cardiac output these patients may manifest. As previously outlined, three patients in this series died of thromboembolic complications. One presently surviving patient is moderately disabled as a result of a post-Fontan stroke, presumably caused by a cerebral embolus originating in the left heart. In our 167 surviving patients on whom follow-up was obtained, only eight were receiving any type of anticoagulation therapy. These patients were all receiving warfarin, three because of a mechanical prosthetic mitral valve and five, with a history of atrial dysrhythmias, as thrombogenic prophylaxis. The decision whether to routinely give the post-Fontan patient long-term warfarin anticoagulant therapy because of the potential for thromboembolism remains controversial, but it has not been our policy to do so. Again, with the new surgical techniques that "streamline" the flow of blood through the right atrium and decrease the right atrial volume, it is hoped that the potential for thrombus formation may be reduced.

Protein-losing enteropathy as a late complication. With the elevation of systemic venous pressure as an inevitable consequence of the Fontan procedure, it was realized that

protein-losing enteropathy might develop, a complication seen previously in patients with constrictive pericarditis and secondarily elevated systemic venous pressures. Three of the late deaths in this series were clearly a result of this complication. Only two of the survivors in our series are presently requiring therapy for symptomatic protein-losing enteropathy. One has become asymptomatic, with normalization of serum albumin, on daily subcutaneous heparin therapy; the second has had symptomatic improvement on heparin therapy but continues to have slightly subnormal serum albumin levels. A recent review from our institution (16) revealed that the risk of development of protein-losing enteropathy was 13.4% after 10 years. Although in an appreciable number of patients the enteropathy may be mild and remain asymptomatic, in others it may produce varying degrees of hypoalbuminemia, with secondary pleural effusions, ascites and so on. Medical therapy with anticongestive measures and albumin infusion may benefit some symptomatic patients. In our experience, corticosteroids, as advocated by some (17), have not been beneficial. Fenestration of the atrial baffle, either surgically or in the catheterization laboratory—which reduces RA pressure and increases cardiac output, although at the expense of some arterial hypoxemia—has reportedly (18) improved or eliminated protein-losing enteropathy. Recently, the use of daily subcutaneous heparin has been advocated (19,20). On occasion, when the patient's disorder is refractory to all other measures and is continuing to deteriorate, cardiac transplantation is considered and has been successful in reversing this debilitating and potentially fatal complication in some but not all patients (21).

Advisability of pregnancy in adult women. As many female Fontan patients reach adulthood, a frequent question is the advisability of pregnancy. Four of the adult female survivors in this series have had one successful full-term pregnancy each. In each instance, the pregnancy and delivery were uncomplicated, the child had no congenital anomalies and the mothers felt their postpartum state of health was unchanged from their status before pregnancy. There were four known spontaneous miscarriages in our follow-up group, three in one woman with no successful pregnancies and one in one of the women who also had a successful pregnancy. We have surveyed the adult female post-Fontan population in an attempt to determine pregnancy outcome (22). Although these patients did seem to have an increased incidence (33%) of spontaneous first-trimester miscarriage, in those who did not miscarry there were 15 live births from 14 different mothers. Only one child was born considerably prematurely, and all 15 infants, including the premature baby, are doing well. The only congenital heart disease in the offspring was a case of secundum atrial septal defect. In addition, all of the 14 mothers, including the one who had two children, seemed to tolerate their pregnancies well, had no special problems at the time of delivery and believe that their postpartum state of health was not compromised in comparison with their antepartum condition. Thus, although this series is small, we concluded that women with a good clinical result who want to have a child have a good chance of a successful pregnancy without putting themselves at substantial risk. Clearly, however, these women should be made aware of the potential hazards of pregnancy, and if they do become pregnant the pregnancy should be managed in a facility with a "high-risk" obstetric unit.

Conclusions. Before the Fontan approach was devised, patients with tricuspid atresia had a natural history characterized by gradually increasing disability due to progressive hypoxemia and ventricular myocardial deterioration. In patients who did survive the childhood years, their quality of life was usually substantially impaired by the time of adolescence, and survival beyond early adulthood was uncommon (23). The initial 25-year experience with the Fontan procedure at this institution has generally been gratifying. Well-established selection criteria have evolved, and for such properly selected patients the operative risk is now 2%. Although follow-up reveals a significant late mortality and morbidity, surgical advances and the current tendency to perform the operation earlier in life (24), before serious ventricular impairment has occurred, allow us to be optimistic that these problems will become less prevalent. Of the surviving 167 patients in this series on whom follow-up information was available, most of whom are adults, 89% were in NYHA class I or II, capable of full-time school or employment and leading lives of good quality. Clearly, additional time must pass before firm conclusions can be drawn regarding the benefits of this palliative approach for a congenital heart lesion for which correction is not possible. However, this initial 25-year experience suggests that the Fontan procedure has been a major step forward in providing help and hope to patients with tricuspid atresia, and that the children currently undergoing the procedure have an excellent chance of surviving the operation and leading good-quality lives well into their adult years.

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