UNIVENTRICULAR REPAIR

Early and midterm results

A total of 202 patients (62 with tricuspid atresia and 140 without tricuspid atresia) underwent univentricular repair at our unit from January 1990 to September 1994. Of these patients, 182 had nonfenestrated and 20 had fenestrated interatrial baffles. Early mortality was 15.9% (29/182) in the group with nonfenestrated baffles and 5% (1/20) in the group with fenestrated baffles. The follow-up period ranged from 2 to 58 months. Seven late deaths occurred, and five patients were lost to follow-up. Of 160 patients who have been evaluated in the outpatient department in the past 3 months, 142 (88.75%) required no cardiac medicines and were in functional class I. Risk factors analyzed for early mortality and significant effusion were age, preoperative diagnosis, type of Fontan modification, cardiopulmonary bypass time, aortic crossclamp time, pulmonary artery size, associated pulmonary arterioplasty, takedown of systemic-pulmonary artery shunt, and pulmonary artery debanding, along with the Fontan operation. Bypass time exceeding 120 minutes was associated with a higher early mortality (12/47 vs 18/155; $p = 0.0187$). Bypass time exceeding 120 minutes ($p = 0.0456$) and aortic crossclamp time exceeding 60 minutes ($p = 0.0278$) were associated with significant postoperative effusion. Other factors were not associated with any significantly increased risk for early mortality or postoperative effusions. Fenestration of the interatrial baffle appeared to decrease early mortality, although the numbers are too small to be statistically significant. The prevalence of effusions did not differ significantly between the group with fenestrated baffles and the group without fenestrated baffles. (J THORAC CARDIOVASC SURG 1995;110:1692-701)

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Since the first univentricular repair described by Fontan and Baudet¹ in 1971, the basic concept of systemic vein-pulmonary artery connection has been modified many times.²⁻⁸ In the early era operative mortality was high, ranging from 17% to 21%.^{9,10} Recent results are more encouraging be-

cause of the introduction of total cavopulmonary connection,¹¹ an interim bidirectional connection between the superior vena cava and pulmonary artery as staged palliation,^{12,13} and, finally, baffle fenestration.¹⁴⁻¹⁶

This study was designed to analyze our experience with univentricular repair and especially to review the effects of baffle fenestration on outcome.

Patients and methods

A total of 202 patients who underwent univentricular repair from January 1990 to September 1994 form the subject of this study. The group included 149 male and 53 female patients. Mean age was 7.32 years (standard deviation 6.13 years, range 0.6 to 52 years). Age distribution of these patients is shown in Fig. 1. Sixty-six patients had palliation before the Fontan operation: Blalock-Taussig shunt, $n = 58$; Waterston shunt, $n = 1$; pulmonary artery banding, $n = 3$; and bidirectional Glenn shunt,

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Fig. 1. Age distribution and Fontan failure.

$n = 4$. Underlying diagnoses were tricuspid atresia in 62 patients, complex congenital heart disease with functional single ventricle in 139 patients, and right ventricular tumor in 1 patient (Table I).

Selection criteria used for the Fontan operation included functional single ventricle in combination with the following:

1. Satisfactory pulmonary artery size
2. Reparable localized pulmonary artery stenosis with good distal branch pulmonary arteries
3. Pulmonary artery pressure less than 18 mm Hg or less than 20 mm Hg with net left-to-right shunt
4. Satisfactory ventricular function (end-diastolic pressure <12 mm Hg and/or angiographically good ventricular function) with no significant ventricular hypertrophy
5. No left ventricular outflow tract obstruction
6. No more than mild atrioventricular valve regurgitation

Preoperative McGoon ratio ranged from 1.2 to 4.5 (mean 2.46, standard deviation 0.56), mean pulmonary artery pressure ranged from 5 to 20 mm Hg (mean 11.05 mm Hg, standard deviation 5.29 mm Hg), and left ventricular end-diastolic pressure ranged from 3 to 15 mm Hg (mean 10.72 mm Hg, standard deviation 3.94 mm Hg). Fifteen patients had stenosis of the pulmonary artery bifurcation and 1 patient had atrioventricular valve regurgitation.

The surgical approach consisted of cardiopulmonary bypass (CPB) with direct caval cannulation, core cooling, and myocardial protection with cold blood cardioplegia. The following types of operations were performed:

1. Atriopulmonary connection was performed in 64 patients with atrial septal defect closure (in patients with tricuspid atresia) or with atrial neoseptation. A polytetrafluoroethylene (PTFE) patch was used to close the defect.
2. Total cavopulmonary connection was performed in 138 patients. Three variations of the operation were used:

Table I. Underlying diagnosis in the patients undergoing Fontan procedure

Diagnosis	No.
Tricuspid atresia	62
DORV, VSD, PS with small LV or RV	55
D-TGA, VSD, PS with small LV or RV	15
L-TGA, VSD, PS with small LV or RV	20
Single ventricle with or without PS	37
Tetralogy of Fallot with small LV	4
Tumor of RV	1
Pulmonary atresia with intact ventricular septum	1
Hypoplastic right heart syndrome	5
Total anomalous pulmonary venous drainage with functional single ventricle	2
Total	202

DORV, Double-outlet right ventricle; VSD, ventricular septal defect; PS, pulmonary stenosis; LV, left ventricle; RV, right ventricle; D-TGA, Transposition of great arteries; L-TGA, congenitally corrected transposition of great arteries.

- type A—total cavopulmonary connection with a lateral atrial tunnel from the inferior vena cava to the superior vena cava using a PTFE patch ($n = 108$); type B—total cavopulmonary connection with a lateral tunnel created with interatrial septum ($n = 25$); type C—bilateral bidirectional superior cavopulmonary connection with diversion of the inferior vena cava to the left pulmonary artery via the coronary sinus ($n = 5$).
3. A fenestrated Fontan operation was performed with a 4 or 5 mm coronary punch in the last 20 patients: atriopulmonary connection, $n = 8$; total cavopulmonary connection type A, $n = 11$; and total cavopulmonary connection type B, $n = 1$.

Table II. Associated procedures

	No.
Takedown of Blalock-Taussig shunt	58
Takedown of Waterston shunt	1
Pulmonary artery debanding	3
Atrioventricular valve repair	1
Pulmonary arterioplasty	15
Total anomalous pulmonary venous drainage repair	2
Subaortic resection	1

Associated procedures are listed in Table II. Four patients also had preexisting bidirectional superior cavopulmonary connection and underwent Fontan completion.

Results

All patients who died within 30 days of the operation and all who survived Fontan takedown were included in the category *Fontan failure*. This designation may not be strictly true inasmuch as obvious *hemodynamic* Fontan failure was present in only 19 patients. *Hemodynamic Fontan failure* was defined as low cardiac output after a Fontan-type repair, with persistent need to maintain central venous pressure higher than 18 mm Hg and/or inordinately high fluid requirement to maintain acceptable systemic perfusion. The immediate causes of death are included in Table III. Some of the deaths ($n = 15$) not caused by hemodynamic Fontan failure (causes 2 to 7) may have been the result of chronic low cardiac output with elevated systemic venous pressure. Others were relatively sudden terminal events in seemingly satisfactory postoperative recoveries. To eliminate any bias in including patients in or excluding them from the category of Fontan failure, we included all deaths within 30 days of operation in the broad category of Fontan failure.

Any pleural effusion that lasted for more than 10 days after the operation was called *significant*. Any pleural effusion that recurred after chest tube removal was also included in the list of significant effusions.

Early results. Thirty-four patients were included in the category Fontan failure: four who survived Fontan takedown and 30 who died within 30 days. The four patients who survived Fontan failure had conversion to bidirectional superior cavopulmonary connection immediately ($n = 3$) or within 24 hours ($n = 1$). Six patients had conversion to bidirectional superior cavopulmonary connection more than 48 hours after Fontan repair and one patient was treated by fenestration of the atrial baffle within 24

Table III. Causes of early death

	No.
Hemodynamic Fontan failure	15
Sepsis	6
Pulmonary embolism	2
Intractable supraventricular tachycardia	2
Respiratory failure	2
Unexplained sudden death	1
Neurologic	2
Total	30

hours. None of these patients survived. In one patient with a fenestrated Fontan the repair was converted to a bidirectional Glenn shunt immediately, but the patient later died of low cardiac output. The other seven patients with hemodynamic Fontan failure died without any additional procedure (takedown/fenestration) being performed.

Durations of CPB and aortic crossclamping as risk factors for Fontan failure and significant effusion were analyzed initially by considering both of the independent variables to be continuous variables using the Wilcoxon rank-sum test for two groups. Then distribution of both CPB and aortic crossclamp time were analyzed to find a clinically useful cutoff point at which the incidence of Fontan failure or effusion started to rise significantly. For the CPB time the cutoff point was found to be 120 minutes and for aortic crossclamp time it was 60 minutes.

Multivariate analysis of all the risk factors showed results similar to those derived by univariate analysis; that is, CPB time greater than 120 minutes is a significant risk factor for both Fontan failure and effusion, and aortic crossclamp time greater than 60 minutes is a significant risk factor for significant effusion only. All other risk factors were found to be insignificant.

Table IV shows the risk factors for Fontan failure and related p values. There is no statistically significant difference in early mortality in patients of different age groups ($p > 0.3$). Age distribution and Fontan failure in different age groups are shown in Fig. 1.

Fontan failure and early postoperative mortality were more prevalent in patients having a CPB time of more than 120 minutes than a shorter CPB time ($p < 0.02$), whereas long aortic crossclamp time ($p > 0.7$) did not affect the early postoperative outcome. The incidence of Fontan failure was similar in patients with atriopulmonary connection (12/64) and in those with total cavopulmonary connection (22/138) ($p > 0.5$).

Associated procedures like takedown of an arterial shunt ($p > 0.4$), pulmonary artery debanding ($p > 0.4$), or pulmonary arterioplasty ($p > 0.3$) did not affect early mortality (see Table IV).

Four patients had a bidirectional Glenn shunt as interim palliation. This small group of patients was potentially predisposed to Fontan-related complications because of greater number of risk factors (average number of Choussat criteria violated 2.25) than were those patients undergoing Fontan repair without staging (average number of Choussat criteria violated 0.59). No mortality or significant effusion occurred in this group of patients with non-fenestrated Fontan repairs. This difference did not achieve statistical significance ($p > 0.3$).

Fontan failure occurred in nine of 62 patients with tricuspid atresia and in 25 of 140 patients without tricuspid atresia. This difference is statistically insignificant ($p > 0.6$).

This retrospective study may be divided into two study periods: 1990 to 1993, when all patients had nonfenestrated interatrial baffles, and 1994, when all patients underwent a fenestrated Fontan procedure. Analysis of patients in both study periods reveals no significant preoperative difference in patient characteristics (Table V).

Fontan failure was encountered in 33 of 182 patients having nonfenestrated Fontan repairs (29 early deaths and four takedown survivors). One Fontan failure occurred in the 20 consecutive patients having fenestrated Fontan repair. This patient had a concomitant repair of total anomalous pulmonary venous connection and died of respiratory infection and disseminated intravascular coagulation after takedown. The small number of patients in the fenestration group prevents the difference from becoming statistically significant ($p > 0.1$) (Fig. 4, Table VI).

Seventy patients (34.7%) had persistent pleural effusion for more than 10 days. Incidence of effusion was similar in different age groups, in patients with different types of Fontan operations, and in patients with or without the diagnosis of tricuspid atresia, with or without baffle fenestration (Table VII). However effusion was significantly more prevalent when aortic crossclamp time exceeded 60 minutes ($p < 0.03$) or CPB time exceeded 120 minutes ($p < 0.05$) (Tables VIII and IX).

Midterm results of nonfenestrated Fontan repair. The follow-up period for nonfenestrated Fontan repair ranged from 12 months to 54 months. There were six late deaths among these patients; two

Table IV. Risk factors for early mortality and Fontan failure

Risk factor	p Value	
	Early mortality	Fontan failure
Age	0.3974	0.1010
Type of operation	0.5249	0.6197
Absence of fenestration	0.1918	0.1362
Takedown of Blalock-Taussig shunt	0.4432	0.7003
CPB time >120 min*	0.0187	0.0129
Aortic crossclamp time >60 min	0.7215	0.8580
Pulmonary artery size	0.4393	0.6481
Other than tricuspid atresia	0.6043	0.5583
Pulmonary arterioplasty	0.3542	0.7066
Previous pulmonary artery band	0.4661	0.4324
One-stage Fontan procedure	0.3989	0.3635

*Significant risk factor for early mortality and Fontan failure.

patients died of pneumonia, one of protein-losing enteropathy, and one during reoperation for baffle leak and residual pulmonary artery stricture. One patient died of refractory ventricular fibrillation during surgical pleurodesis for recurrent pleural effusion. One patient had acute pancreatitis and died after exploratory laparotomy.

Five patients of the nonfenestrated group were lost to follow-up. A total of 142 patients with a nonfenestrated atrial baffle have been followed up in the past 6 months; 124 were receiving no cardiac medicines and were in New York Heart Association functional class I. The remaining 18 patients were receiving diuretics with or without vasodilators and were in New York Heart Association functional class II (15 patients) or III (3 patients) (Fig. 5).

Midterm results of fenestrated Fontan repair. One late death (cause unknown) occurred in the group of patients receiving fenestrated intraatrial baffles. All 18 surviving patients with fenestrations have been followed up for 2 to 11 months. All of them are free of symptoms (class I) and require no cardiac medications (Fig. 5).

Oxygen saturation ranged from 85% to 94% in patients having a 4 mm fenestration ($n = 14$) and from 80% to 85% in patients having a 5 mm fenestration ($n = 4$) at a mean follow-up period of 5 months. Four of these patients had spontaneous closure of their fenestration as demonstrated by lack of any right-to-left shunt by contrast echocardiography. None of the patients with fenestrations have been taken up for closure of the fenestration.

Fig. 2. Duration of aortic crossclamp and incidence of effusion and Fontan failure.

Fig. 3. Duration of CPB and incidence of effusion and Fontan failure.

Discussion

Because of the difficulty in separating the non-Fontan failure deaths from the Fontan failure deaths without bias, all deaths within 30 days of repair were included in the heading *Fontan failure*. Some of the deaths would not fall strictly under the hemodynamic definition of Fontan failure but for the reason mentioned earlier are included in this category.

We have followed a strict policy of selecting patients for the Fontan procedure¹⁷⁻¹⁹ with reference to pulmonary artery size, pulmonary artery distortion, and ventricular function. However, inasmuch as the majority of patients had severe pulmonary stenosis, were severely cyanotic, and underwent the Fontan procedure as their first intervention, pulmonary artery entry was the exception at preoperative cardiac catheterization. A preoperative pul-

monary artery pressure reading was therefore not available, although it could be estimated echocardiographically or directly recorded during the operation. Hence calculations of pulmonary vascular resistance were not commonly available.

Possibly because of the unavailability of preoperative data, a Fontan failure rate of 18.1% (33/182) was encountered in the first part of the experience when only nonfenestrated univentricular repairs were performed. Fenestration of the interatrial baffle probably acts as a safety valve for such patients so that cardiac output is preserved at the cost of systemic arterial desaturation even in patients with an elevated pulmonary vascular resistance index.

Unlike the usual patient population referred for Fontan repair in the Western hemisphere, most of our patient population had not undergone palliation and was severely cyanosed. For those two reasons, only infrequently were pulmonary artery pressure and pulmonary vascular resistance index determined in the preoperative period. This article highlights the role of routine fenestration in providing a vent to combat undiagnosed elevations of pulmonary vascular resistance index that were probably resulting in high Fontan failure rates in nonfenestrated repair.

Oxygen saturation ranged between 85% and 94% in patients with 4 mm fenestration and 80% and 85% in those with 5 mm fenestrations. Four patients had no evidence of right-to-left shunting through the baffle. We have not made any attempts at closing fenestrations because of the excellent symptomatic status of patients and reluctance of the children's parents to subject them to further intervention.

Our overall early mortality rate of 14.9% compares favorably with those of other series.^{13, 20} More liberal use of the fenestrated Fontan approach has reduced the mortality to 5%. The only patient who died (among 20 having the fenestrated Fontan procedure) had conversion to a bidirectional Glenn shunt on the operating table because of difficulty in weaning from CPB after fenestrated atriopulmonary connection and total anomalous pulmonary venous drainage repair. Improvement in early results with the fenestrated Fontan approach had been reported previously.^{19, 21}

Fontan and associates²² (1983) reported that age at operation (<4 years or >15 years) was a significant risk factor ($p < 0.001$) for early mortality. Table IV shows the various risk factors for early mortality in our series. There was no statistically significant difference in mortality among patients of different

Table V. Comparison of preoperative status of patients undergoing fenestrated and nonfenestrated Fontan operation

	Fenestrated (n = 20)	Nonfenestrated (n = 182)
Mean age (yr)	7.68	7.29
Standard deviation	3.95	6.33
Mean McGoon Ratio	2.50	2.46
Standard deviation	0.48	0.57
Mean PA pressure (mm Hg)	11.50	10.96
Standard deviation	6.97	5.03
LVEDP (mm Hg)	11.00	10.68
Standard deviation	0.82	4.24
Percent with PA bifurcation stenosis	25	5.5
Percent with Choussat criteria violations		
None	50	47.8
One	40	44.0
Two	10	8.2

PA, Pulmonary artery; LVEDP, left ventricular end-diastolic pressure.

age groups ($p > 0.3$). Other workers have also noted that age does not exert significant influence on survival.^{17, 23} Possibly use of a strict selection criteria has eliminated this risk factor.

One category of patients, albeit small, was devoid of any significant effusion or Fontan failure. Patients who had had an interim palliation by means of a bidirectional Glenn shunt had neither Fontan failure nor effusions on Fontan completion. This possible advantage has been mentioned before.¹² This finding attains greater significance because these patients had a higher number of Fontan risk factors (average number of Choussat criteria violated 2.25) than those coming directly to Fontan repair (average number of Choussat criteria violated 0.59).

The superiority of one modification of the Fontan operation over the other cannot be substantiated in our series, because there is no significant difference in early mortality among the patients receiving different types of total cavopulmonary or atriopulmonary connection ($p > 0.5$). The choice of one procedure over the other was dictated by the surgical anatomy in a particular case, and this fact highlights the importance of performing a nonrestrictive anastomosis between the systemic veins and the pulmonary artery rather than preferring a particular modification of the Fontan procedure.

In our series, early mortality was positively correlated with CPB time longer than 120 minutes ($p < 0.02$). Longer bypass time was often associated with complex repair procedures.

Fig. 4. Effects of baffle fenestration on outcome.

Table VI. Incidence of early mortality, Fontan failure, and effusion in patients with fenestrated and nonfenestrated baffle

Intraatrial baffle	Total No.	Early mortality	Fontan failure	Significant effusion
Fenestrated	20	1	1	6
Nonfenestrated	182	29	33	64
<i>p</i> Value		0.1918	0.1362	0.6450

The damaging effects of CPB are amplified as total CPB time increases.²⁴ Although we do not have evidence of whole body inflammation (e.g., complement levels), analysis of CPB time as a continuous variable highlights the progressively increasing risk with longer duration of CPB, with values reaching statistical significance at a CPB time exceeding 120 minutes.

Interference with integrity of the capillary endothelium resulting in "capillary leak syndrome"²⁴ may contribute to the prevalence of effusion after longer periods of CPB. This would be further compounded by elevated systemic venous pressure that results after univentricular repair. Postoperative persistent effusions into body cavities had been traditionally explained by increased venous pressure. Our findings suggest that increased duration of CPB may be an etiologic factor too, along with increased venous pressure.

Longer aortic crossclamp time (despite cardioplegic protection), with attendant greater myocardial cell damage and myocardial dysfunction, would also

Table VII. Risk factors for effusion

Risk factor	<i>p</i> Value
Age	0.8577
Type of operation	0.3125
Absence of Fenestration	0.6450
Takedown of BT shunt	0.8848
CPB time >120 min*	0.0456
Aortic crossclamp time >60 min*	0.0278
Pulmonary artery size	0.8644
Patients other than tricuspid atresia	0.8689
Pulmonary arterioplasty	0.9111

*Significant risk factor for effusion.

result in a higher central venous pressure, thereby compounding the damaging effects of CPB.

It can therefore be inferred that a complete Fontan procedure is best avoided when complex repair is to be undertaken. However, associated procedures like takedown of classic or modified Blalock-Taussig shunts ($p > 0.4$), pulmonary arterioplasty ($p > 0.3$), or pulmonary artery debanding ($p > 0.9$) did not increase early mortality (see Table IV). Mayer and associates¹⁸ had noted that pulmonary artery distortion from a previous palliative procedure increased the risk of early death. However, in our series previous palliative procedures were not associated with increased risk of either early death or postoperative effusion. In fact, the early mortality among 15 patients who underwent pulmonary arterioplasty was 6.7% (1/15) compared with an overall mortality of 14.9% ($p > 0.3$). This difference can be explained by wider use of the bidirectional Glenn shunt in patients in whom a less

Fig. 5. Freedom from early death and takedown after fenestrated and nonfenestrated Fontan repair.

Table VIII. Incidence of early mortality, Fontan failure, and effusion in patients with different durations of CPB

CPB time	Total No.	Early mortality	Fontan failure	Significant effusion
<120 min	155	18	20	48
>120 min	47	12	14	22
<i>p</i> Value		0.0187	0.0129	0.0456

than satisfactory pulmonary arterioplasty was achieved.

Debate continues regarding the suitability of the one-stage complete Fontan procedure in patients with borderline pulmonary artery size. The successful outcome of the Fontan procedure is mainly dependent on low pulmonary vascular resistance. Senzaki and coworkers²⁵ concluded that smaller pulmonary artery size was hemodynamically disadvantageous after the Fontan procedure because it increased peak central venous pressure. They have not shown the effects of smaller pulmonary artery on ultimate outcome. We calculated the McGoon ratio in our cases and found no significant difference in incidence of either early mortality ($p > 0.4$) or effusion ($p > 0.8$) in patients with different pulmonary artery sizes.

Table IX. Incidence of early mortality, Fontan failure, and effusion in patients with different durations of crossclamping

Crossclamp time	Total No.	Early mortality	Fontan failure	Significant effusion
<60 min	122	19	21	35
>60 min	80	11	13	35
<i>p</i> Value		0.7215	0.8580	0.0278

The prevalence of effusion (persisting more than 10 days) in our series is positively correlated with the duration of aortic crossclamping exceeding 60 minutes ($p < 0.03$) and the duration of CPB exceeding 120 minutes ($p < 0.05$).

Experience has shown that Fontan failure should be treated aggressively. When takedown to bidirectional superior cavopulmonary connection was performed within 24 hours of the Fontan operation, the outcome was successful, whereas a delay of more than 48 hours resulted in multiorgan failure, leading to death.

Conclusion

Early mortality is higher if CPB time exceeds 120 minutes, suggesting that complicated reconstructive procedure should be avoided with a complete Fon-

tan procedure. The incidence of effusion is related to the duration of aortic crossclamping and to the duration of CPB (see Figs. 2 and 3). In the past year, by adopting the fenestrated Fontan approach for all cases, we have been able to reduce mortality after the Fontan operation. Whether small (4 mm) fenestrations should be closed at all is a question that still needs to be addressed.

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Discussion

Dr. John E. Mayer (Boston, Mass.). We agree with a number of the conclusions you have reached, for example, that a fenestrated Fontan seems to be the preferable technique and that if a patient with a Fontan repair is failing, then the decision to take it down to a bidirectional cavopulmonary shunt should be made early.

I would like to clarify one aspect regarding some of our prior publications, and, in particular, the issue of pulmonary artery distortion as a risk factor. In our previously reported series, pulmonary artery distortion that could not be repaired, in other words, unrepaired pulmonary artery distortion, was almost universally associated with failure,

whereas a repairable pulmonary artery distortion was a less powerful risk factor.

I have some questions about some of the other conclusions that you have reached. You had little preoperative data on pulmonary vascular resistance and only inferential data from echocardiograms on pulmonary artery pressure. Inasmuch as the pulmonary artery pressure measured in an open chest, anesthetized patient may not be closely related to that measured in a catheterization laboratory, it would be difficult to conclude that pulmonary artery pressure is not a risk factor for Fontan operations. Alternatively, if pulmonary artery pressure was low in all patients, then it would be unlikely to appear as a risk factor either. Can you tell us the number patients in whom pulmonary artery pressure was directly measured and how many really had a significant elevation in the pulmonary arteries?

Second, you have concluded that age was not a risk factor, but you have not told us how this variable was evaluated. Was it analyzed as a continuous variable or was it considered only as age outside the classic 4- to 15-year age range?

Finally, you have mentioned that approximately 50% of the patients exceeded one or more of the Choussat-Fontan limits. Can you tell us which limits were exceeded and with what frequency? There is some danger in simply counting the number of deviations from those classic criteria, because that assumes that all of the criteria have an equally important impact on outcome. Our prior studies and those of others would suggest that some of the criteria either are not important or are of much greater impact than others of the original Fontan criteria.

Dr. Sharma. Thank you, Dr. Mayer. You are correct in observing that we had little information on pulmonary vascular resistance and that a majority of our pulmonary artery pressure measurements were done in the operating room. In the risk factors that we analyzed, we did not include pulmonary artery pressure because we performed a Fontan repair only if the pulmonary artery pressure was less than a mean of 18 mm Hg. I also agree with your observation that the pulmonary artery pressure measurements in the operating room may not accurately represent the true state of the existing circulation. This is probably the reason that the Fontan failure rate was high before we started using fenestrations; that is, a failure rate of about 18% when we were really adhering closely to the other prescribed guidelines for Fontan selection. Also, since we started using fenestrated baffles in the last 20 patients (and there have been another 10 to date), mortality has decreased substantially. Our impression is that borderline pulmonary vascular resistance and pulmonary artery pres-

sure elevations (undetected in the open chest, anesthetized patient) probably contributed to the excessive mortality in the group with nonfenestrated baffles. Whatever marginal elevation of pulmonary vascular resistance or pulmonary artery pressure was present caused the fenestrations to decompress from right to left without compromising cardiac output (albeit with some amount of desaturation, which improved on follow-up).

We have analyzed age both as to whether it fell outside the classic Fontan criteria of 4 to 15 years and as a continuous variable, and we did not find it to be a significant risk factor. Of course, only five of our patients were younger than 1 year of age, and only 22 were younger than 2 years.

Fontan criteria violations included age, pulmonary artery size, pulmonary artery distortion, and atrioventricular valve regurgitation. Pulmonary artery pressures were all less than 18 mm Hg (mean). Not having any information on pulmonary vascular resistance index, we did not include it in our analysis. No Fontan criteria were violated in 50% of the cases, one criterion in 40%, and two or more criteria were violated in 10% of the patients. No statistically significant difference in Fontan failure or effusion rate was detected between any of these patients groups.

Dr. Frank L. Hanley (*San Francisco, Calif.*). Before I make my comment, I would like to emphasize that it is not meant to be critical of the fenestrated Fontan itself. In a very large series of patients with univentricular heart disease such as this, in which institutional learning curves are extremely important and many parameters are rapidly changing, it is important to be careful about the way the conclusions are drawn. In a series of over 200 cases, to examine just the last 20 cases (which were fenestrated) and compare them to the previous 180 or so cases and conclude that all of the differences between the two groups were due to the fenestration may not be valid. A more sophisticated analysis that takes into account a number of other potentially important variables would be necessary before one can conclude that the benefits are all due to introduction of the fenestration.

Dr. Sharma. Thank you, Dr. Hanley. The distribution of cases between the two groups is definitely unequal and the time periods are different. However, since we began doing univentricular repairs in our center, we had a consistent Fontan failure rate of more than 15% until we switched over to fenestrating all our intraatrial baffles. The mortality figures then dropped markedly and abruptly. Nevertheless, your objection to our conclusion is still valid. The only way we could prove whether a fenestration really helps would be to perform a randomized trial between two comparable patient groups.