SURGERY FOR CONGENITAL HEART DISEASE

STAGED OPERATION TO FONTAN INCREASES THE INCIDENCE OF SINODIAL NODE DYSFUNCTION

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Morbidity and mortality of total cavopulmonary connection (modified Fontan procedure) may be decreased in many patients with single ventricle in whom the risk of surgery is high by performing the operations in a staged fashion. Each operative intervention, however, exposes the sinoatrial node region to risk of injury, and a multistaged approach may increase the risk of altered sinoatrial node function in these patients. The purpose of this study was to compare the prevalence of perioperative arrhythmias in patients undergoing either a primary or staged approach to the Fontan operation. Records were retrospectively reviewed for all patients having a Fontan procedure between January 1988 and December 1992. Of 324 patients undergoing a Fontan operation, 227 had a Fontan operation without a prior cavopulmonary shunt (group 1) and 97 had a cavopulmonary shunt before a Fontan operation (group 2). Arrhythmias were classified as altered sinoatrial node function, supraventricular tachycardia, or atrioventricular block. The prevalence of both transient (resolving before hospital discharge) and fixed (persisting until hospital discharge) altered sinoatrial node function was similar for the two groups after cavopulmonary shunt or primary Fontan despite a heterogeneous patient population (group 1: 10.6%/4.4%; group 2: 10.3%/3.1%; $p = 0.28$). Conversion from cavopulmonary shunt to Fontan in group 2 resulted in a higher prevalence of altered sinoatrial node function in the early postoperative period (transient: 23.7%; fixed: 23.7%; $p < 0.001$) and on follow-up (group 1: 7.7%; group 2: 16.7%; $p < 0.02$). In group 2, 40 of 82 patients without arrhythmia after first intervention (cavopulmonary shunt) had an arrhythmia after the second intervention (Fontan) (49%); of 14 with an arrhythmia after the first operation, 10 (71%) had one at the second intervention ($p < 0.01$). In conclusion, a multistaged operative pathway to Fontan reconstruction is associated with a higher early risk of altered sinoatrial node function. The occurrence of altered sinoatrial node function after cavopulmonary shunt is itself a risk factor for arrhythmia after the Fontan operation. Longer follow-up is needed to assess the full impact of this finding. (J THORAC CARDIOVASC SURG 1996;111:833-40)

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Over the past 25 years modifications of the Fontan procedure have become the primary method of permanent palliation for cardiac defects with a functional single ventricle. The successful application of this principle to a broad range of patients with cardiac defects of varied anatomic and physio-

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Table I. Series and group data on patients completing Fontan operation, 1988-1992

	All patients	Group 1	Group 2
No.	324	227	97
Male/female	190:134 (1.4)	136:91 (1.5)	54:43 (1.3)
Age at Fontan (range) (yr)	5.14 yr (0.6-36)	5.36 yr (.6-36)	4.63 yr (1.3-18.7)
Age at cavopulmonary shunt (range) (yr)			3.02 yr (.25-16)
Ventricular morphology			
Right ventricle	102 (31%)	61 (27%)	41 (42%)
Left ventricle	183 (57%)	133 (59%)	50 (52%)
Both	39 (12%)	33 (14%)	6 (6%)
Hospital deaths	25 (7.7%)	17 (7.5%)	8 (8.2%)

Table II. Palliative procedures done before cavopulmonary shunt or Fontan procedure

Prior operation	Group 1 (n = 227)	Group 2 (n = 97)
Systemic-pulmonary shunt	151	76
PA banding	58	19
Atrial septectomy	25	9
Norwood procedure	12	21
Repair PA stenosis	5	9
Coarctation	7	5
Damus-Kaye-Stansel	5	3
RVOT enlargement	2	2
Repair TAPVC	1	2
Pacemaker	4	1
Other	2	0

PA, Pulmonary artery; RVOT, right ventricular outflow tract; TAPVC, total anomalous pulmonary venous connection.

logic complexity has involved a number of modifications to Fontan's original techniques and selection criteria.¹⁻⁵ Morbidity and mortality may be decreased in many patients with single ventricle by performing the repair in a staged fashion.^{6,7} For this reason the trend has been toward routine use of an intermediate cavopulmonary shunt procedure before a Fontan procedure in patients with single ventricle physiology.

Atrial arrhythmias are seen in the early and late postoperative periods after the Fontan procedure and may be poorly tolerated by the patient with single ventricle physiology.⁸⁻¹² With a staged approach, each operative intervention can expose the sinoatrial (SA) node area to the risk of injury, thus potentially increasing the risk of atrial arrhythmia with such a management scheme. The purpose of this study was to attempt to determine if a staged approach including an intermediate cavopulmonary shunt resulted in a higher incidence of abnormal SA node function than did performance of primary modified Fontan procedure.

Patients and methods

A total of 324 patients underwent a modified Fontan procedure at the Children's Hospital in Boston between January 1988 and December 1992 (Table I). In 227 patients a primary Fontan operation was done in a single stage (group 1). In 97 patients a preliminary cavopulmonary shunt was performed before the completion Fontan operation (group 2). Seventy-nine percent of group 1 patients and 98% of group 2 patients had undergone palliative cardiac procedures before the Fontan operation or the cavopulmonary shunt (Table II). References to "first" or "second" operative intervention in this report refer to the cavopulmonary shunt or Fontan procedures only, not to prior palliative procedures. The distribution of patients between groups by year of Fontan completion is shown in Fig. 1. The average age at the Fontan procedure was not different between the groups (5.4 ± 0.3 vs 4.6 ± 0.5 years, $p = 0.21$). Patients in group 2 underwent their cavopulmonary shunt procedure at an average age of 3.0 ± 0.5 years (range 3 months to 16 years). Cardiac anatomy was classified into six groups used in prior analyses of single ventricle from this institution.⁴ The one- and two-stage groups were both heterogeneous, with some notable differences between groups (Fig. 2). The proportion of patients with single left ventricle and transposed great arteries was higher in group 1, and the proportion of patients with hypoplastic left heart syndrome and heterotaxy was higher in group 2. In more general anatomic terms, patients in group 2 more frequently had a ventricle of right ventricular morphology (42% vs 27%), whereas more group 1 patients had functional single ventricle anatomy though with two ventricular chambers (15% vs 6%).

Throughout the study period the operative techniques were relatively constant, although with a number of surgeons performing the operations over the years and with the anatomic variety of the anomalies, some variations in technique were unavoidable. Total cavopulmonary connection (rather than atriopulmonary connection) was accomplished by anastomoses between the superior vena cava and the right pulmonary artery. In general, the superior vena cava was transected at the level at which it crossed the pulmonary artery, typically 5 to 10 mm above the cavopulmonary junction. In some cases the right pulmonary artery was transected and sewn end to side to the superior vena cava. An intraatrial lateral tunnel

Fig. 1. Number of patients having the completion Fontan procedure by year.

directing the inferior caval flow to the cavopulmonary connection was created with an intraatrial polytetrafluoroethylene baffle. The baffle was fenestrated with a 4 mm punch in 77% of patients. In group 2, 70 patients underwent a bidirectional cavopulmonary anastomosis between the divided superior vena cava and the right pulmonary artery, and the cardiac end of the vena cava was oversewn. In 16 of the 97 patients a classic Glenn procedure had been performed, and in nine a hemi-Fontan procedure was done, which included anastomosis of the cardiac end of the superior vena cava to the underside of the right pulmonary artery with placement of an intraatrial patch at the superior vena cava–right atrial junction.

All patients in this review underwent routine 12-lead electrocardiograms (ECGs), while awake but resting, before the operation, immediately after the operation, and at the time of hospital discharge. Additional studies were performed as clinically indicated. In addition to these, hospital progress notes and nursing records were reviewed for other documentation of arrhythmias.

ECGs were routinely obtained during follow-up visits for those patients followed up at our institution. Further evaluations of rhythm disturbances (Holter monitoring, electrophysiologic studies) were obtained when clinically indicated.

For purposes of this study, an arrhythmia was considered as any variance from a normal sinus mechanism with antegrade atrioventricular (AV) conduction. Normal sinus mechanism for patients with situs solitus atria was accepted as a P-wave axis between 0 and 90 degrees in the frontal plane and a rate appropriate for age. For heterotaxy or atrial situs inversus, the P-wave axis on the earliest available preoperative ECG was accepted as “normal sinus” for that patient. Arrhythmias were subclassified as (1) abnormal SA node function, including sinus bradycardia, ectopic atrial pacemaker, or junctional bradycardia; (2) supraventricular tachycardia, including atrial fibrillation, atrial flutter, ectopic atrial tachycardia, junctional ectopic tachycardia, and AV nodal reentrant tachycardia; and (3) AV block. An ectopic atrial focus was designated as a persistent shift in the P-wave axis greater than 45 degrees as compared with the patient’s preoperative ECG. Ectopic atrial foci tended to retain a nearly normal heart rate for the patient’s age; junctional rhythms typically were 50 to 90 beats/min, commonly prompting use of temporary atrial pacing. Ten patients in the study had

Fig. 2. Distribution of patients by anatomic diagnosis. NRGA, Normally related great arteries; TGA, transposed great arteries; LV, left ventricle; RV, right ventricle; ?V, indeterminate ventricular morphology; HLHS, hypoplastic left heart syndrome.

complete AV block, of which nine cases were evident before the cavopulmonary shunt or Fontan operation. Atrial activity was able to be analyzed in all patients because of the presence of DDD pacing systems, which in all patients were atrial sensing and ventricular pacing.

Rhythm disturbances isolated to the first 24 hours after the operation were not considered for this evaluation because they were not consistently documented by 12-lead ECG, and exogenous catecholamines were frequently used. Transient arrhythmias were defined as those that persisted beyond the first 24 postoperative hours but resolved by hospital discharge. Fixed arrhythmias were defined as those that persisted beyond the first 24 postoperative hours and were still present at hospital discharge (although some were noted to resolve on later follow-up).

Results

A total of 25 hospital deaths occurred in the total population (7.7%), 17 (7.5%) in group 1 and eight (8.2%) in group 2. In group 1, seven of the 17 patients who died had an arrhythmia: six had a form of supraventricular tachycardia, typically junctional ectopic tachycardia, and one patient had ventricular tachycardia. Only in the patient with ventricular tachycardia was the arrhythmia believed to be the primary cause of death. In group 2, two of the eight patients who died had an arrhythmia, atrial flutter in both patients, and in neither was the arrhythmia judged to be the primary cause of death. Three late deaths have occurred in group 1 and one in group 2, none known to be due to rhythm disturbances.

The prevalence of transient and fixed arrhythmia, predominantly abnormal SA node function, was similar for the two groups after the first operative

Table III. Arrhythmias after cavopulmonary shunt or Fontan operation

	Altered SA node function	Supraventricular tachycardia
Group 1 (n = 227)		
After Fontan		
Total	34 (15%)	15 (6.6%)
Transient	24 (10.6)	9 (3.9)
Fixed	10 (4.4)	6 (2.6)
Group 2 (n = 97)		
After cavopulmonary shunt		
Total	13 (13.4%)	1 (1%)
Transient	10 (10.3)	1 (1)
Fixed	3 (3.1)	0
After Fontan		
Total	46 (47.4%)	5 (5%)
Transient	23 (23.7)	5 (5)
Fixed	23 (23.7)	0

intervention (i.e., cavopulmonary shunt or Fontan). Transient arrhythmias developed in 33 patients in group 1 (14.5%) and fixed arrhythmias in 16 (7.0%); by comparison, transient arrhythmias developed in 11 patients in group 2 (11.3%) and fixed arrhythmias in three (3%) ($p = 0.28$ vs group 1). Heterotaxy was the only anatomic type that did not follow this pattern. Arrhythmia was more prevalent after the first intervention in patients with heterotaxy if that procedure was a Fontan operation rather than a cavopulmonary shunt (55% vs 7%, $p < 0.01$). The distribution of arrhythmia type is listed in Table III. Approximately 80% of all *transient* abnormal SA node function was junctional bradycardia, whereas nearly 70% of *fixed* abnormal SA node function was ectopic atrial foci. The use of a temporary pacemaker (nearly always atrial pacing for bradycardia) was also similar for the two groups after the first operative intervention (group 1, 11.9%; group 2, 7.2%; $p = 0.14$). Postoperative tachycardia (junctional ectopic) was seen in only a single group 2 patient after cavopulmonary shunt and in a similar but low incidence in each group after the Fontan procedure (group 1, 6.6%; group 2, 5%).

In group 2, both transient and fixed arrhythmias (transient 28.9%; fixed 23.7%) were more prevalent after the second operative intervention (completion Fontan) than after the first intervention (cavopulmonary shunt) ($p < 0.001$ vs group 1). This pattern was seen in all anatomic groups, including those with heterotaxy. This increase was accounted for predominantly by abnormal SA node function. The prevalence of arrhythmias for the different types of cavopulmonary shunt procedures is shown in Table

IV. Use of temporary atrial pacing was also more frequent after the Fontan operation than cavopulmonary shunt in group 2 (27.8% after Fontan, 7.2% after cavopulmonary shunt; $p < 0.001$). The occurrence of an arrhythmia after the first intervention (cavopulmonary shunt) was a significant risk factor for arrhythmia after the completion Fontan operation. Forty of 82 patients with no arrhythmia after the first operation had one after the second (49%), but the arrhythmia rate was 10 of 14 (71%) after the second procedure for those having arrhythmia after the first operation ($p < 0.01$), with six of these patients exhibiting the same rhythm disturbance after each intervention.

Reliable follow-up information on cardiac rhythm was available for 130 group 1 patients (57%, mean duration 1.8 years) and for 54 group 2 patients (56%, mean duration 1.6 years). At the time of most recent follow-up, 10% of these 130 group 1 patients were noted to have a nonsinus rhythm (7.7% abnormal SA node function, 2.3% supraventricular tachycardia) compared with 24.1% of the 54 group 2 patients (16.7% abnormal SA node function, 7.4% supraventricular tachycardia) ($p < 0.02$). Permanent pacemaker insertion for management of atrial arrhythmias was infrequent. Four patients in each group required such a procedure (group 1, 3%; group 2, 7%; $p = 0.18$). Indications for permanent pacing were sick sinus syndrome in three, junctional bradycardia in three, and supraventricular tachycardia in two.

Neither year of operation nor age of the patient had any bearing on risk of arrhythmia. Arrhythmia (transient or fixed) was more prevalent after the first intervention in patients with a morphologically right ventricle as the single ventricle if that intervention was a complete Fontan operation, that is, a one-stage approach (31% vs 14%, $p < 0.05$); however, when managed with a two-staged plan, they demonstrated the additive risk of arrhythmia (53%) after the second intervention, similar to that seen with other ventricular morphologies.

Discussion

Atrial arrhythmias have been reported to occur in 15% to 50% of patients after modifications of the Fontan procedure both in the early and late postoperative periods.^{12, 13} Management of these arrhythmias is often challenging.¹⁴ Surgical techniques have been modified significantly over the past 20 years, including direct cavopulmonary connections, non-distensible intraatrial baffles, fenestrations or adjust-

Table IV. Group 2 arrhythmias subgrouped by type of first-stage (cavopulmonary shunt) procedure

Type of cavopulmonary shunt	Arrhythmia after cavopulmonary shunt			Arrhythmia after Fontan procedure		
	None	Transient	Fixed	None	Transient	Fixed
Bidirectional Glenn	59 (84%)	8 (11%)	3 (4%)	31 (44%)	23 (33%)	16 (23%)
Hemi-Fontan	6 (67)	3 (33)	0 (0)	7 (78)	1 (11)	1 (11)
Classic Glenn	16 (100)	0 (0)	0 (0)	9 (56)	2 (13)	5 (31)

able atrial septal defects,¹⁵ and preliminary cavopulmonary shunt procedures. These modifications, along with better patient selection, have been associated with marked improvements in survival, especially in patients with high-risk anomalies. The effect of these modifications on the prevalence of atrial arrhythmia is not yet clearly demonstrated. Our results indicate that a staged approach is associated with a higher incidence of atrial arrhythmia, predominantly alterations in sinus node function.

Some authors have reported a reduction in early¹⁶ and late^{17, 18} postoperative atrial tachyarrhythmias with the cavopulmonary connection technique as compared with use of an atriopulmonary connection, although follow-up is shorter for patients who have had cavopulmonary connection. Early postoperative arrhythmias and length of follow-up have both been reported to be significant risk factors for late rhythm problems.^{13, 18} Atrial tachyarrhythmias have been the predominant type of arrhythmia reported in other studies, although in the current series we observed more problems with SA node function. Comprehensive surveillance methods for arrhythmias were not used during the period of this study; thus we cannot make any statement regarding the true prevalence of arrhythmias after the Fontan operation. Our current follow-up is too brief to make any comment regarding long-term risk of these arrhythmias.

Staging surgical correction of single ventricle anomalies with the addition of an intermediate bidirectional Glenn procedure exposes the SA node region to dissection twice, thus potentially risking dysfunction caused by direct injury to the SA node or injury to the SA node blood supply. Despite the significant heterogeneity of the two patient groups from the standpoint of anatomic type, both had a similar risk for abnormal SA node function after their first interventions whether this was a cavopulmonary shunt or a Fontan procedure. Our data suggest an additive risk of abnormal SA node function with repeated dissection in the region of the SA

node. Only patients with heterotaxy anatomy demonstrated an equally high risk of arrhythmia with single or multistaged Fontan reconstruction. The clinical significance of the observed arrhythmias is less clear. Although an ectopic atrial pacemaker rhythm was observed in many cases, this rhythm did not disrupt AV synchrony, and heart rates were not sufficiently slow to cause symptoms. The long-term stability of this rhythm, though, cannot be predicted from the current study. Kürer, Tanner, and Vetter¹⁹ performed electrophysiologic studies on 30 patients in the early postoperative period after primary modified Fontan operations. They demonstrated abnormal SA node function, prolonged atrial refractoriness, delayed intraatrial conduction, and inducibility of atrial tachyarrhythmias. Clinical problems related to SA node function were uncommonly seen in their small cohort of patients, but we believe that there will be a need for careful, lifelong follow-up.

SA node function is best evaluated by 24-hour Holter monitoring, which was done only selectively during the period of this study. Although this patient selection factor is a potential source of error in this retrospective study, the bias is more likely toward the underdetection of arrhythmias rather than an overestimation.

The routine use of temporary atrial and ventricular pacing leads has been our practice in this group of patients. Their utility from a therapeutic standpoint was seen in a number of cases of postoperative abnormal SA node function. Moreover, we have found the atrial leads particularly useful in the diagnosis and evaluation of atrial arrhythmias in these and other patients, and we strongly encourage their routine use in this population at risk for such rhythm disturbances.

Most important, this study has focused our attention on the fact that the additive risk of arrhythmia from multiple interventions delineates a patient population requiring careful, long-term postoperative follow-up. The risk of atrial arrhythmia increases with duration of follow-up.²⁰

Further modification of surgical technique to avoid repeated dissection around the SA node may diminish the additive risk of arrhythmia from a multistaged approach to the patient with single ventricle while preserving the overall morbidity and mortality advantage. The "hemi-Fontan" procedure, as we and others have done it,²¹ may achieve this result. With this technique both cavopulmonary anastomoses are done during the initial procedure and an intraatrial patch blocks flow from the pulmonary circuit to the right atrium. Only superior vena caval flow is committed to the pulmonary arteries, as with a standard bidirectional Glenn procedure, but the subsequent intervention may be done completely intraatrially, avoiding dissection in the region of the SA node or its blood supply. This technique differs from that described by Douville, Sade, and Fyfe,²² in which an atriopulmonary anastomosis was used. Balaji and associates²³ examined 21 patients for atrial arrhythmias after undergoing a hemi-Fontan operation. One patient had atrial flutter in the postoperative period, but none had junctional or ectopic atrial rhythm, suggesting no SA node damage. However, no follow-up through completion of the Fontan procedure was reported. In the current series, three of nine patients undergoing a hemi-Fontan operation had transient arrhythmias after this first-stage procedure (two junctional, one supraventricular tachycardia). At completion Fontan, occlusion of the segment of superior vena cava between the patch and the pulmonary artery anastomosis was noted in two of the nine patients, thus necessitating dissection again in the SA node region, and both had transient abnormal SA node function. Thus arrhythmia was less prevalent after a second intervention with the hemi-Fontan as the initial stage, although the number of cases in our series in which this technique was used was too small to allow meaningful statistical comparison.

The retrospective nature of this study imposed a number of limitations that prevented an estimate of the true prevalence of arrhythmia after these procedures. We did not have data on catecholamine dosages or atrial filling pressures, and precise details of the surgical technique were not always included in the operative reports. Holter monitor recordings to determine the true prevalence of arrhythmias were available in only a fraction of the patients. Despite these limitations, our results do suggest that a staged approach to the Fontan operation, including an intermediate cavopulmonary shunt, is associ-

ated with an increased risk of abnormal SA node function after the Fontan operation.

These results suggest that the trend toward more routine application of a staged approach must be approached with caution. The risk of early postoperative atrial arrhythmia, with its possible implications regarding risk of late arrhythmia, must be considered along with other widely recognized risk factors, such as ventricular function, AV valve competence, pulmonary vascular resistance, and pulmonary artery distortion in the selection of a surgical management strategy for the patient with a single ventricle anomaly. In addition, close attention to SA node function during follow-up with periodic Holter monitoring is recommended in all patients who have had the Fontan operation, especially those undergoing a staged correction.

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Discussion

Dr. Gordon K. Danielson (*Rochester, Minn.*). Dr. Manning and coauthors have shown convincingly that the more one operates near the SA node, the more one can expect to see SA node dysfunction. We first observed this relationship many years ago from the technique used then for cannulation of the superior vena cava for the Mustard procedure.

The cannula was initially inserted through the right atrial appendage and an incision was made from the appendage to the inferior vena cava. The incision was then extended superiorly across the cavoatrial junction, the cannula was moved to the superior end of the incision, and the atrium was closed caudal to the cannula. This provided excellent exposure and was an innovative way to position the cannula away from the baffle suture line in this era before direct cannulation of the cava, but it effectively divided the SA node or its blood supply. Only 53% of the patients having the Mustard procedure had sinus rhythm on follow-up a mean of 13 years later.

There is still some controversy about the significance of interruption of the SA node artery, because controlled

studies are lacking. However, we and others have noted similar observations of SA node dysfunction when incisions are carried across the cavoatrial junction during repair of anomalous pulmonary venous connection from the right lung to the superior vena cava and after earlier versions of the maze procedure in which the SA node artery was interrupted.

The London anatomist Dr. Robert H. Anderson has shown that in one third of the patients the SA node artery ascends on the medial surface of the superior vena cava, in one third it ascends laterally, and in one third it ascends both medially and laterally. Thus incisions, either medial or lateral, put the SA node in some jeopardy.

With regard to the modified Fontan procedure, it is important to avoid enlarging the proximal cavopulmonary anastomosis with an incision made either medially or laterally across the cavoatrial junction. The authors did not describe their technique for the completion portion of the Fontan, and my question to them is this: Where were the incisions placed and how was the proximal cavopulmonary connection established?

The authors have suggested that the hemi-Fontan may reduce the prevalence of SA node dysfunction at the completion operation. I would like to add another option that we have used—a lateral tunnel combined with an atriopulmonary connection at the superior end of the lateral tunnel. This can be constructed with a flap of atrial appendage or anterior atrial wall. It can be constructed to avoid the SA node and artery, and it can be used to create a large anastomosis for both a one-stage or two-stage modified Fontan procedure.

Dr. Manning. Thank you, Dr. Danielson. Part of the problem in trying to group together patients with such a variety of anatomic types is that it is difficult to generalize specifics about operative technique. In attempting to do that, many times on going back to do a completion Fontan procedure we continued the longitudinal atriotomy up through the superior caval orifice, generally neither laterally nor medially. Then we constructed the baffle and closed the incision. But that was not by any means a uniform technique.

Frank L. Hanley (*San Francisco, Calif.*). I think it is a tribute to the field that this entire session on the Fontan operation is devoted to subtleties of fluid dynamics and concern over the SA node function rather than mortality and stroke. The field has come a long way.

Regarding these topics of fluid dynamics and rhythm, I would like to comment about the total extracardiac Fontan, which is the modification that we have been using. We, of course, are not the first to use this technique routinely. Dr. Marcelletti has a longer and, I believe, a larger series than we do. This technique addresses all of the issues being discussed today. The total extracardiac Fontan creates a completely tubular conduit from the inferior vena cava on up. The upper connection tapers onto the peripheral aspect of the right pulmonary artery and, in fact, onto the lower lobe branch of the right pulmonary artery.

One of its most promising potential benefits of this technique relates to rhythm. The less trauma the right atrium sustains the better, whether it be in the form of suture load, manipulation, incisions, or digging out the SA

node at the time of the conversion from a bidirectional cavopulmonary shunt or hemi-Fontan to a lateral tunnel Fontan. With the extracardiac conduit technique, to borrow a phrase from our general surgery colleagues, one can literally do a "no touch" technique regarding atrial tissue in a fair number of the cases. There are, of course, cases in which it is necessary to do an atrial septectomy or a valve repair. As a result, some atrial trauma is inevitable in these cases. But, no matter which morphologic category is examined, the manipulation of the atrium is markedly reduced when comparing the extracardiac conduit to the lateral tunnel.

It is a positive sign that many groups continue to explore different Fontan techniques. There is, at present, no answer as to which is the best Fontan technique. Only long-term follow-up will answer this question.

Dr. Manning. Thank you for those comments. We did not use the extracardiac tunnel technique in this series of patients.

Dr. Richard A. Jonas (Boston, Mass.). In the analysis of results of the Fontan operation from Children's Hospital in Boston, which was presented at a recent meeting of the Castaneda Society, one of the negative risk factors for Fontan survival was higher pulmonary blood flow before the Fontan operation. It seems that if the ventricle was handling a greater volume load, so long as the end-diastolic pressure and pulmonary vascular resistance were not elevated, it was an advantage to have more pulmonary blood flow before the Fontan operation. I think many of us have seen this. The patient who is very cyanotic and has very low pulmonary blood flow before a Fontan procedure should arouse more concern than the patient who is very pink and is handling a big volume load.

With that in mind, Dr. Manning, would you please comment on your statement that there is a hemodynamic benefit to the staged Fontan? Is it really hemodynamically beneficial to split this operation into two stages? Aren't we reducing pulmonary blood flow with the preliminary bidirectional Glenn shunt? In these days of the fenestrated Fontan operation, I wonder if we should not be bypassing this step in the majority of patients.

Dr. Manning. That is a provocative question. The initial benefit in this staged procedure may not have been hemodynamic as much as an overall survival benefit, especially demonstrated in the population with hypoplastic left heart syndrome. The advantage was probably due to removing the volume load from the single ventricle,

particularly the morphologically right ventricle. People are just starting to realize that other things are being changed, and the pulmonary blood flow may be negatively affected.

Because of the good success with hypoplastic left heart syndrome, recent trends have been to increasingly use a staged approach to avoid any risk factor, such as pulmonary artery distortion or elevated pulmonary artery pressures. However, using the fenestration as an alternative stage or avoiding the stage altogether may enhance overall survival and still improve ventricular function, preserving pulmonary blood flow and avoiding complications.

Mr. Marc Roger de Leval (London, England). I want to follow on Dr. Jonas's comments. I do agree that patients with a high pulmonary blood flow, low pulmonary vascular resistance, and normal ventricular functions are the best candidates for the Fontan operation. With this in mind, I have continued to use the systemic-pulmonary artery shunt as a first-stage palliation, rather than the bidirectional cavopulmonary anastomosis.

Dr. Jonas. A little bit of extra volume load but not too much, right? Because if you overload the ventricle, then you have lost any advantage.

Dr. Hanley. To continue the discussion, there are other explanations for the observation that unpredictable Fontan outcomes can occur in patients with low preoperative pulmonary blood flow. Fontan candidates who have low preoperative pulmonary blood flow present certain difficulties in the calculations used to determine pulmonary vascular resistance. Under low flow conditions, small differences in the estimates of certain pieces of physiologic data necessary for these calculations can result in extremely wide variability in the calculated pulmonary vascular resistance. It may well be that the patients with low pulmonary blood flow preoperatively who are struggling postoperatively are struggling not because of a change in physiology but because they were inappropriately selected on the basis of inaccurate preoperative pulmonary vascular resistance calculations.

Dr. Jonas. I think it is both. It is certainly difficult to calculate pulmonary resistance when pulmonary blood flow is low, but also it is difficult to know what the ventricular compliance is going to be. Dr. Manning, can you comment on what the reserve compliance is under those circumstances?

Dr. Manning. No.