postoperative bradyarrhythmia was seen more often after the ECC than the ILT. Therefore, the decision on which type of Fontan to perform on a specific patient should be based on individual, patient-related hemodynamic factors and surgical center preferences, and not on a presumed reduction in arrhythmias.

The authors thank the Department of Pediatrics, Children's Hospital of Philadelphia, for providing funding for this study; Jan Weber, RN, for nurse coordination support; Xuemei Zhang, MS, for statistical support; and the Pediatric and Congenital Electrophysiology Society and its membership for their support.

References

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax*. 1971;26: 240-8.
2. de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience. *J Thorac Cardiovasc Surg*. 1988;96:682-95.
3. Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit. A new form of right heart bypass. *J Thorac Cardiovasc Surg*. 1990;100:228-32.
4. Balaji S, Gewillig M, Bull C, de Leval MR, Deanfield JE. Arrhythmias after the Fontan procedure. Comparison of total cavopulmonary connection and atriopulmonary connection. *Circulation*. 1991;84(5 Suppl):III162-7.
5. Cecchin F, Johnsrude CL, Perry JC, Friedman RA. Effect of age and surgical technique on symptomatic arrhythmias after the Fontan procedure. *Am J Cardiol*. 1995;76:386-91.
6. Stephenson EA, Lu M, Berul CI, Etheridge SP, Idriss SF, Margossian R, et al. Arrhythmias in a contemporary Fontan cohort: prevalence and clinical associations in a multicenter cross-sectional study. *J Am Coll Cardiol*. 2010; 56:890-6.
7. Giannico S, Corno A, Marino B, Cicini MP, Gagliardi MG, Amodeo A, et al. Total extracardiac right heart bypass. *Circulation*. 1992;86(5 Suppl):II110-7.
8. Azakie A, McCrindle BW, Van Arsdell G, Benson LN, Coles J, Hamilton RET, et al. Extracardiac conduit versus lateral tunnel cavopulmonary connections at a single institution: impact on outcomes. *J Thorac Cardiovasc Surg*. 2001;122: 1219-28.
9. Nurnberg JH, Ovroutski S, Alexi-Meskishvili V, Ewert P, Hetzer R, Lange PE. New onset arrhythmias after the extracardiac conduit Fontan operation compared with the intraatrial lateral tunnel procedure: early and midterm results. *Ann Thorac Surg*. 2004;78:1979-88.
10. Bae EJ, Lee JY, Noh CI, Kim WH, Kim YJ. Sinus node dysfunction after Fontan modifications–influence of surgical method. *Int J Cardiol*. 2003;88:285-91.
11. Lee JR, Kwak J, Kim KC, Min SK, Kim WH, Kim YJ, et al. Comparison of lateral tunnel and extracardiac conduit Fontan procedure. *Interact Cardiovasc Thorac Surg*. 2007;6:328-30.
12. Sarkis V, Sreeram N, Trieschmann U, Ben Mime L, Bennink G. Comparison of arrhythmia incidence after the extracardiac conduit versus the intracardiac lateral tunnel Fontan completion. *Int J Cardiol*. 2011;146:258-9.
13. Ovroutski S, Dahnert I, Alexi-Meskishvili V, Nurnberg JH, Hetzer R, Lange PE. Preliminary analysis of arrhythmias after the Fontan operation with extracardiac conduit compared with intra-atrial lateral tunnel. *Thorac Cardiovasc Surg*. 2001; 49:334-7.
14. Dilawar M, Bradley SM, Saul JP, Stroud MR, Balaji S. Sinus node dysfunction after intraatrial lateral tunnel and extracardiac conduit Fontan procedures. *Pediatr Cardiol*. 2003;24:284-8.
15. Kumar SP, Rubinstein CS, Simsic JM, Taylor AB, Saul JP, Bradley SM. Lateral tunnel versus extracardiac conduit Fontan procedure: a concurrent comparison. *Ann Thorac Surg*. 2003;76:1389-96.
16. Hakacova N, Lakomy M, Kovacikova L. Arrhythmias after Fontan operation: comparison of lateral tunnel and extracardiac conduit. *J Electrocardiol*. 2008; 41:173-7.
17. Cohen MI, Bridges ND, Gaynor JW, Hoffman TM, Wernovsky G, Vetter VL, et al. Modifications to the cavopulmonary anastomosis do not eliminate early sinus node dysfunction. *J Thorac Cardiovasc Surg*. 2000;120:891-900.
18. Blaufox AD, Sleeper LA, Bradley DJ, Breitbart RE, Hordof A, Kanter RJ, et al. Functional status, heart rate, and rhythm abnormalities in 521 Fontan patients 6 to 18 years of age. *J Thorac Cardiovasc Surg*. 2008;136:100-7.

Journal of the American College of Cardiology
© 2010 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Vol. 56, No. 11, 2010
ISSN 0735-1097/$36.00
doi:10.1016/j.jacc.2010.03.079

Arrhythmias in a Contemporary Fontan Cohort

Prevalence and Clinical Associations in a Multicenter Cross-Sectional Study

Elizabeth A. Stephenson, MD, MSc,* Minmin Lu, MS,† Charles I. Berul, MD,‡ Susan P. Etheridge, MD,§ Salim F. Idriss, MD, PhD,‖ Renee Margossian, MD,‡ John H. Reed, MD,¶ Ashwin Prakash, MD,#‡ Lynn A. Sleeper, ScD,† Victoria L. Vetter, MD,†† Andrew D. Blaufox, MD,** for the Pediatric Heart Network Investigators

Toronto, Ontario, Canada; Watertown and Boston, Massachusetts; Salt Lake City, Utah; Durham, North Carolina; Charleston, South Carolina; New York and New Hyde Park, New York; and Philadelphia, Pennsylvania

Objectives Our aim was to examine the prevalence of arrhythmias and identify independent associations of time to arrhythmia development.

Background Since introduction of the Fontan operation in 1971, long-term results have steadily improved with newer modifications. However, atrial arrhythmias are frequent and contribute to ongoing morbidity and mortality. Data are lacking regarding the prevalence of arrhythmias and risk factors for their development in the current era.

Methods The Pediatric Heart Network Fontan Cross-Sectional study evaluated data from 7 centers, with 520 patients age 6 to 18 years (mean 8.6 ± 3.4 years after the Fontan operation), including echocardiograms, electrocardiograms, exercise testing, parent-reported Child Health Questionnaire (CHQ) results, and medical history.

Results Supraventricular tachycardias were present in 9.4% of patients. Intra-atrial re-entrant tachycardia (IART) was present in 7.3% (32 of 520). The hazard of IART decreased until 4 to 6 years post-Fontan, and then increased with age thereafter. Cardiac anatomy and resting heart rate (including marked bradycardia) were not associated with IART. We identified 3 independent associations of time to occurrence of IART: lower CHQ physical summary score ($p < 0.001$); predominant rhythm ($p = 0.002$; highest risk with paced rhythm), and type of Fontan operation ($p = 0.037$; highest risk with atriopulmonary connection). Time to IART did not differ between patients with lateral tunnel and extracardiac conduit types of Fontan repair. Ventricular tachycardia was noted in 3.5% of patients.

Conclusions Overall prevalence of IART was lower in this cohort (7.3%) than previously reported. Lower functional status, an atriopulmonary connection, and paced rhythm were determined to be independently associated with development of IART after Fontan. (Relationship Between Functional Health Status and Ventricular Performance After Fontan–Pediatric Heart Network; NCT00132782) (J Am Coll Cardiol 2010;56:890–6) © 2010 by the American College of Cardiology Foundation

Since its introduction in 1971, the Fontan operation has consistently been the primary surgical technique used for palliation of patients with single-ventricle physiology (1). Morbidity and mortality in Fontan patients have decreased dramatically, but rhythm abnormalities remain a significant problem (2–6). Previous studies of cohorts having undergone the Fontan operation have recognized that arrhythmias are an important contributor to morbidity. The most frequent of these is intra-atrial re-entrant tachycardia (IART), seen in 16% to 22% of patients at 5-year follow-up (2,7,8). Detailed information regarding factors that influence the development of arrhythmias in this population is limited. Importantly, controversy exists in the pediatric cardiology and cardiovascular surgery community as to possible differences in

See page 897

From *The Hospital for Sick Children, Toronto, Ontario, Canada; †New England Research Institutes, Watertown, Massachusetts; ‡Children's Hospital Boston, Boston, Massachusetts; §Primary Children's Hospital, Salt Lake City, Utah; ‖Duke University Medical Center, Durham, North Carolina; ¶Medical University of South Carolina Children's Hospital, Charleston, South Carolina; #Morgan Stanley Children's Hospital of New York-Presbyterian, New York, New York; **Schneider Children's Hospital, New Hyde Park, New York; and the ††Children's Hospital of Philadelphia, Philadelphia, Pennsylvania. Supported by U01 grants from the National Heart, Lung, and Blood Institute (HL068269, HL068270, HL068279, HL068281, HL068285, HL068292, HL068290, HL068288, HL085057). All authors have reported that they have no relationships to disclose.

Manuscript received December 15, 2009; revised manuscript received February 17, 2010, accepted March 18, 2010.

arrhythmia burden between the lateral tunnel Fontan and the extracardiac conduit Fontan procedures.

Using data from the Pediatric Heart Network's (PHN) Fontan Cross-Sectional study, we examined the prevalence of arrhythmias in the current generation of Fontan survivors, as well as features of anatomy and surgical repair that may be associated with arrhythmia prevalence. We sought to identify any factors associated with development of IART, and any difference in atrial arrhythmia prevalence between the 2 most commonly performed Fontan procedures in the current era.

Methods

Funded by the National Heart, Lung, and Blood Institute, the PHN is a collective of pediatric cardiac centers in the U.S. and Canada, supported by a data coordinating center at the New England Research Institutes. The Fontan Cross-Sectional study gathered data from 7 centers in 2003 and 2004, and examined multiple clinical aspects of Fontan survivors, with a primary aim of exploring the correlations between functional outcomes and ventricular performance measures (9–11). The study was approved by an institutional review board or research ethics board at each participating institution. Written informed consent was obtained from a parent or guardian, and assent obtained where applicable.

Fontan Cross-Sectional study. The Fontan Cross-Sectional study included surviving children age 6 to 18 years who had undergone a Fontan procedure at least 6 months prior to entering the study (9). The subjects were identified through a search of all cases seen in the prior 3 years, and were invited to participate. Study data and testing within 3 months of enrollment included echocardiograms, electrocardiograms (ECGs), exercise testing results, health status questionnaires, and medical history from a review of the medical record. Patients were excluded if they had a coexisting noncardiac health condition that would preclude participation in the study protocol or otherwise confound study end points.

Abbreviations and Acronyms

CHQ = Child Health Questionnaire

ECG = electrocardiogram

IART = intra-atrial re-entrant tachycardia

PHN = pediatric heart network

VT = ventricular tachycardia

Study participants. A total of 1,078 records were screened for potential participation in the Fontan Cross-Sectional study, and 644 patients (60%) were found to be eligible. Of these, 546 consented (85%) to participate in the Fontan Cross-Sectional study. From this study sample, a subgroup was selected for this analysis: 17 were excluded because they had no ECG or exercise test completed; 7 were excluded because they had undergone Fontan conversion, which would alter the natural history of the Fontan operation; 1 patient was excluded due to second-degree heart block; and 1 because the nature of the arrhythmia was indeterminate. Thus, 520 patients were included in the analytic dataset (Table 1).

Medical record review. Standardized data forms were used to extract pertinent data from the medical record regarding details of cardiac anatomy, type of surgical repair, as well as both early and late complications since Fontan surgery, including onset of supraventricular or ventricular arrhythmias. In all patients who were identified as having supraventricular arrhythmias, the relevant rhythm documentation was reviewed at the local center and the arrhythmia classified more specifically as ectopic atrial tachycardia, atrioventricular re-entrant tachycardia, or IART (a primary macro-re-entrant atrial arrhythmia).

Table 1 Fontan Cross-Sectional Study Subject Characteristics

Characteristic	Total, n	All	History of IART	No History of IART	p Value
n		520	38	482	
Age at study, yrs	520	11.9 ± 3.4 (11.3)	14.4 ± 3.6 (15.6)	11.7 ± 3.3 (11.1)	<0.001
Age at most recent Fontan, yrs	520	3.4 ± 1.9 (2.8)	3.9 ± 1.9 (3.4)	3.3 ± 1.9 (2.8)	0.01
Years since most recent Fontan	520	8.6 ± 3.4 (8.2)	10.7 ± 3.9 (12.1)	8.5 ± 3.3 (8.1)	<0.001
Male	315	61%	55%	61%	0.50
Race/ethnicity					0.27
Hispanic	35	7%	11%	7%	
Non-Hispanic, white	375	76%	81%	76%	
Non-Hispanic, black	49	10%	8%	10%	
Non-Hispanic, other	33	6%	0%	7%	
Unknown	28				
Type of Fontan operation					0.002
Atriopulmonary connection	72	14%	37%	12%	
Intracardiac lateral tunnel	306	59%	53%	59%	
Extracardiac lateral tunnel	62	12%	8%	12%	
Extracardiac conduit	69	13%	3%	14%	
Other	11	2%	0%	2%	

Values are n, mean ± SD (median), or percentage.
IART = intra-atrial re-entrant tachycardia.

ECG and exercise testing. A standard 12-lead ECG was performed at rest in the supine position and recorded at 25 mm/s sweep speed with a 10 mm/mV amplitude. Bradycardia was defined as a resting heart rate less than the 5th percentile for age (12). Predominant rhythm was classified as atrial-based (sinus and atrial escape) versus junctional escape versus paced (14 with other or unknown type were excluded from analyses specific to predominant rhythm). Exercise testing was performed using a standard ramp protocol on an electronically braked cycle ergometer with continuously monitored 12-lead ECGs.

Echocardiography. Two-dimensional echocardiograms and Doppler evaluations of standard short- and long-axis views of the ventricle(s) were centrally interpreted by 1 of 2 readers, and included assessment of ventricular morphology, ventricular systolic and diastolic function, and atrioventricular and semilunar valve regurgitation. Total ejection fraction was expressed as a *z*-score relative to age in normal children (13).

Child Health Questionnaire (CHQ). The CHQ Parent Report PF-50 has been validated in healthy children aged 5 to 18 years and in cardiac cohorts (10,14). This instrument provides summary scores for physical (CHQ-p) and psychosocial (CHQ-ps) well-being.

Statistical methodology. Groups were defined by history versus no history of IART after the Fontan procedure. The Fisher exact test was used to compare the distributions of categorical variables by group, and 2-sample *t* test and Wilcoxon rank sum test were used for comparison of the distributions of continuous variables by group. Time to IART was defined as the number of years from the Fontan procedure to first IART diagnosis. Follow-up was censored at study enrollment for patients having no history of IART. The Kaplan-Meier method was used to estimate the distribution of time to first episode of IART after Fontan (15). Hazard estimation was performed using kernel-based smoothing, with bootstrapped estimates of the pointwise standard errors. Independent associations of time to development of IART were identified using multivariable stepwise Cox proportional hazards regression. All variables in Tables 1 to 3 were considered as candidates in multivariate modeling if the p value was <0.20 on univariate analysis. A p value <0.05 was considered statistically significant. Analyses were performed using SAS version 9.2 (SAS Institute, Cary, North Carolina) and R (kernel-based hazard estimation).

Results

Prevalence of supraventricular tachycardias. As described in our recent PHN publication (6), the overall prevalence of supraventricular tachyarrhythmia was 9.6%, with 50 of 521 patients having had a history of at least 1 episode following hospital discharge after the Fontan procedure. One subject had an indeterminate type of supraventricular tachyarrhythmia and was excluded. Among the remaining 49, 4 (0.8%) had ectopic atrial tachycardia, 7 (1.8%) had re-entrant atrioventricular tachycardia, and 38 (7.3%, representing 78% of all those with supraventricular tachyarrhythmia) had IART.

Clinical associations of IART. AGE AND TIME SINCE FONTAN OPERATION. The prevalence of IART increased with age; the mean age of patients with IART was 14.4 ± 3.6 years, whereas the mean age of those without IART was 11.7 ± 3.3 years ($p < 0.001$). The hazard of a first occurrence of IART was lowest 4 to 6 years after completion of the Fontan operation, with the risk of IART high in the first 2 years after Fontan and then increasing again in later childhood (Fig. 1). Patients who underwent Fontan at an older age were also more likely to have a history of IART. However, these patients had more follow-up time post-Fontan during which IART might occur, because those who had Fontan at an older age (≥4 years) were in the upper age quartile (≥15 years) at study enrollment (Table 1).

Table 2 Association of Predominant Rhythm on Resting ECG and Exercise Testing With History of IART

Variable	n	History of IART*	n	No History of IART*	p Value†
Predominant rhythm on ECG	36		468		<0.001
Atrial-based rhythm		67%		87%	
Junctional escape		3%		6%	
Paced		31%		7%	
Resting HR‡	11	75.3 ± 15.4 (80.0)	433	75.7 ± 16.6 (75.0)	.87
Resting HR <5th percentile‡	11	27%	433	28%	1.00
Maximum HR, beats/min§	11	146 ± 30 (152)	359	157 ± 21 (160)	.31
%Predicted max HR§	11	71.2 ± 14.6 (73.0)	359	75.7 ± 10.1 (77.0)	.44
%Predicted max HR <75§	11	55%	359	41%	.37
P-axis, °‖	23	17.3 ± 58.2 (40.0)	421	29.3 ± 49.8 (38.0)	.54
P-axis 0° to 90°‖	23	65%	421	77%	.21
Right atrial enlargement on ECG	27	7%	431	8%	1.00
Left atrial enlargement on ECG	27	26%	430	14%	.09

*Values are percentage or mean ± SD (median); †p value is from Fisher exact test for categorical variables and Wilcoxon rank sum test for continuous variables; ‡excluded patients with a paced rhythm and patients on heart rate medications; §excluded patients with rate-responsive pacemakers and patients on heart rate medications; ‖excluded patients with a paced or junctional rhythm.

ECG = electrocardiogram; HR = heart rate; IART = intra-atrial re-entrant tachycardia.

Table 3 Association of Anatomy and Function With History of IART

Variable	n	History of IART	n	No History of IART	p Value*
Anatomic diagnosis	38		482		0.24
Single LV: DILV and TA		47%		36%	
Single RV: DIRV; MA and HLHS		13%		28%	
SV, unbalanced AV canal defect		3%		4%	
Other		26%		24%	
SV, heterotaxia syndrome		11%		7%	
Ventricular morphology	38		482		0.13
Left ventricular		61%		49%	
Right ventricular		18%		34%	
Mixed		21%		17%	
L loop anatomy	38	32%	482	18%	0.05
AV valve regurgitation	37	78%		74%	0.70
AV valve regurgitation severity	37		465		0.38
None		22%		27%	
Mild		54%		54%	
Moderate/severe		24%		19%	
Semilunar valve regurgitation	22	55%	279	49%	0.66
Semilunar valve regurgitation severity	22		279		0.94
None		46%		51%	
Mild		50%		39%	
Moderate		5%		9%	
Echocardiographic ejection fraction, %	27	58.1 ± 10.7	366	58.3 ± 10.4	.95
Echocardiographic ejection fraction *z*-score	27	−1.0 ± 2.1	366	−0.9 ± 2.0	.94

*p value is from Fisher exact test for categorical variables and Wilcoxon rank sum test for continuous variables. For valve regurgitation severity grade, the p value is from the Mantel-Haenszel test for linear trend.

AV = atrioventricular; DILV = double inlet left ventricle; DIRV = double inlet right ventricle; HLHS = hypoplastic left heart syndrome; IART = intra-atrial re-entrant tachycardia; MA = mitral atresia; SV = single ventricle; TA = tricuspid atresia.

HEART RATE. We found no association between the prevalence of IART and resting heart rate, bradycardia (see Methods), or maximum heart rate achieved on exercise test (Table 2).

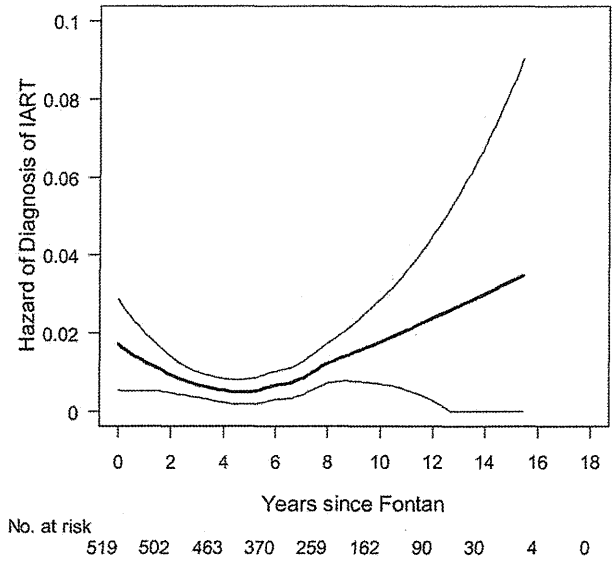

Figure 1 Hazard of IART Following the Fontan Operation, With 95% Confidence Bands

Date of diagnosis was missing for 1 patient, thus the number at risk initially is 519. IART = intra-atrial re-entrant tachycardia.

RESTING RHYTHM. Predominant rhythm on resting ECG was associated with IART ($p < 0.001$), with a higher prevalence of IART in patients with a paced rhythm (26% of those actively paced on ECG) compared with those with an atrial-based (6%) or junctional escape rhythm (3%). This effect was independent of age. Among those with atrial rhythm, there was no significant difference in the P-wave axis between those with and without IART (Table 2).

ANATOMY, CARDIAC FUNCTION, AND PROTEIN-LOSING ENTEROPATHY. There was no association of IART with anatomic diagnosis or with morphology of the systemic ventricle. The prevalence of IART was marginally higher in those with L-looping compared with those without (12% vs. 6%, $p = 0.052$). Neither the degree of atrioventricular valve regurgitation nor the systemic ventricular ejection fraction was associated with a history of IART (Table 3). Protein losing enteropathy occurred in 7.9% of those with IART and 2.9% of those without ($p = 0.12$; $p = 0.19$ after age adjustment) and was not significantly associated with rhythm (data not shown).

FUNCTIONAL STATUS ASSESSMENT. CHQ-p scores were lower ($p = 0.001$) in those with IART than those without (Table 4). The difference remained significant after adjusting for age. There was no significant difference in psychosocial scores (CHQ-ps).

Table 4 Multivariable Cox Regression Model for IART (n = 464)*

Cox Regression Model	Hazard Ratio	95% CI	p Value
CHQ physical summary score	1.23 per 5-U decrease	1.09–1.37	<0.001
Predominant rhythm			0.002
Paced vs. atrial-based	4.01	1.84–8.75	<0.001
Paced vs. junctional escape	4.85	0.61–38.70	0.14
Atrial-based vs. junctional escape	1.21	0.16–9.07	0.85
Type of Fontan operation			0.04
Atriopulmonary connection	—	—	—
Intracardiac lateral tunnel	0.35	0.17–0.75	0.007
Extracardiac lateral tunnel	0.75	0.20–2.87	0.68
Extracardiac conduit	0.22	0.03–1.77	0.16

*The data of 56 of 520 subjects were excluded from the model: 11 patients with "other" type of Fontan, 14 with other/unknown type of predominant rhythm 30 with missing CHQ score, and 1 with unknown date of discharge after Fontan.
CHQ = Child Health Questionnaire; CI = confidence interval; IART = intra-atrial re-entrant tachycardia.

FONTAN OPERATION. The type of Fontan was associated with the presence of IART. Patients with an atriopulmonary connection were more likely to have IART (19%) than those with intracardiac lateral tunnel (7%), extracardiac lateral tunnel (5%), or extracardiac conduit (2%) type (p = 0.002). The prevalence of IART did not differ between patients with an extracardiac conduit (2%) and those with a lateral tunnel procedure (6%, p = 0.22).

Because age has been noted to be a strong predictor of IART, and patients with atriopulmonary connections tended to be older, the association of IART with type of Fontan was reanalyzed by modeling time to first occurrence of IART as a function of number of years since the Fontan operation. We constructed a multivariable Cox regression model (n = 464) to determine whether type of Fontan operation (4 categories) was an independent association of time to occurrence of IART (Table 4). Among these types, only the atriopulmonary connection was independently associated with IART. There was no difference in the covariate-adjusted event-free distributions of time to IART for intracardiac lateral tunnel versus extracardiac conduit (hazard ratio: 1.58, 95% confidence interval: 0.20 to 12.24; p = 0.66). Two additional factors were independently associated with IART: lower CHQ physical summary score and a paced rhythm on ECG. Figure 2 displays the covariate-adjusted (CHQ and predominant rhythm) event-free curves.

PACING. Overall, 68 of 520 patients (13%) had a pacemaker or defibrillator at study enrollment (although were not necessarily actively paced on resting ECG as described in the previous section). Of these 68 patients, 19 (28%) had a history of IART. IART was determined to be the primary indication for pacemaker placement in 8 of 19. Other indications for device placement included bradycardia (26), sinus node dysfunction (7), ventricular tachycardia (VT) (5), junctional rhythm (2), low cardiac output (3), and complete heart block (1). No indication was recorded in 9 patients, and 7 had pacemaker implantation at the time of Fontan without further specific indication. Anatomic diagnosis was examined in relationship to pacemaker presence, and as would be anticipated, there was a strong association between pacemaker and the presence of ventricular L-looping (p = 0.01). There was no association between pacemaker presence and other anatomic features, or any relationship to type of surgery.

VT. Patients with VT were defined as having had at least 1 documented episode of VT or ventricular fibrillation causing symptoms or requiring therapy. VT was seen in 18 patients (3.5%). Because of the infrequency of these events, we did not have the power to detect clinical associations. We found no relationship between VT and valvular regurgitation (atrioventricular or semilunar) or ejection fraction. No other associations with VT were found to be significant, including indicators of chronotropic status, atrial enlarge-

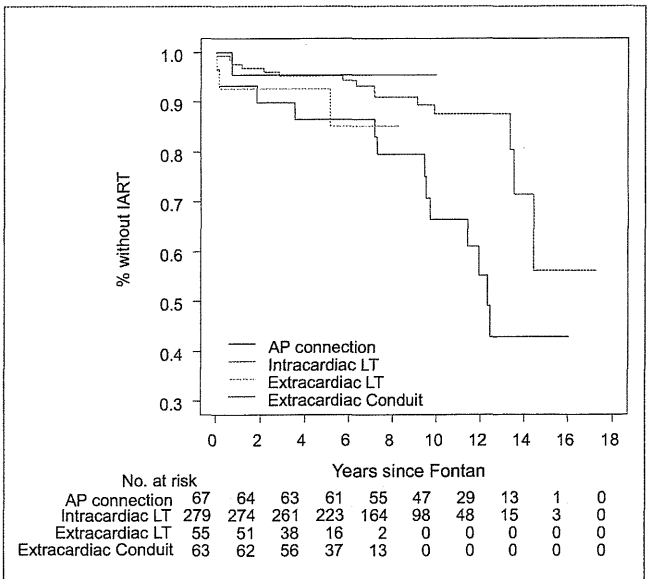

Figure 2 Freedom From IART Following the Fontan Operation by Type of Fontan Procedure

Values are adjusted for predominant rhythm and CHQ physical summary score. Number of patients at risk in each surgical group is shown in the adjoining panel. IART = intra-atrial re-entrant tachycardia.

ment, type of surgical repair, anatomic subgroup, or ventricular morphology.

Discussion

IART. Although long-term results have improved with contemporary modifications of the Fontan operation, IART has remained a frequent (historically 16% to 41%) finding and contributes to ongoing morbidity and mortality. (2,5,7,16–19) In contrast to previous studies, we found only 7.3% of subjects had experienced IART at a mean of 8.6 years follow-up. These previous studies had mean follow-up periods of 3 to 11 years (median 5 years), with the highest rates of IART seen in those studies with longer follow-up periods and a greater proportion of atriopulmonary connection patients. This improvement is likely multifactorial, and changes in surgical strategy have probably played an important role. We found that patients with an atriopulmonary connection-type Fontan operation were at higher risk of developing IART when compared with other surgical strategies, even after adjustment for functional status and predominant rhythm. However, we hypothesize that the majority of the risk in this subgroup is conferred by older age, and patients with types of Fontan procedures other than an atriopulmonary connection were younger in this study. The high hazard that we observed at 10 years post-Fontan and beyond may have an anatomic, electrophysiologic, and hemodynamic basis. Nearly all Fontan patients have atrial incisions and suture lines that may provide the substrate for the development of IART over time. Second, elevated atrial pressures present in nearly all Fontan patients at various stages of palliation lead to atrial dilation and stretch. Atrial fibrosis occurs over time and can result in the development of both anatomic conduction barriers and regions of functional block. These conditions create an anatomic and physiologic substrate that facilitates the development and maintenance of intra-atrial re-entrant circuits. (7,18,20)

Interestingly, there was no significant difference in the event-free distributions of time to IART among the intracardiac lateral tunnel, extracardiac lateral tunnel, and the extracardiac conduit. It has been postulated that the extracardiac conduit, by excluding the majority of atrial tissue, would be less arrhythmogenic than the lateral tunnel (21–23). However, this was not borne out in our cohort. It is important to note that the lateral tunnel and the extracardiac conduit Fontan patients represent a relatively young cohort. Longer follow-up is needed to substantiate these findings.

Sinus node dysfunction has been previously described as associated with IART (7). In contrast, in the present study, even marked bradycardia (heart rate less than the 5th percentile for age) was not associated with IART. Thus, isolated bradycardia per se, as a measure of sinus node dysfunction, is not independently associated with the development of IART. However, this lack of association may be an artifact of excluding the patients with a paced rhythm from heart rate analysis for bradycardia, as this subgroup was more prevalent in the IART group. In contrast, chronotropic competence is not limited by pacemaker implantation. This measure of sinus node dysfunction was also not found to be associated with IART. Nonsinus intrinsic rhythm on ECG, for example atrial or junctional escape rhythm, did not associate with IART. We were not able to discern from our data whether patients had antitachycardia devices placed for IART or for bradycardia.

Lower CHQ physical summary score was the third measure independently associated with IART. Due to the cross-sectional nature of this study, it is not possible to identify causality. It may be that those patients with worse physical function have impaired hemodynamics that contribute to the development of IART. Alternatively, the burden of IART itself may limit physical function. Further follow-up within this study cohort will be useful in identifying any causal relationship or link between these variables.

VT. VT occurred in 3.5% of the population. Given the prevalence of VT, interpretation of possible associations must be undertaken cautiously. Although there was a trend towards lower ejection fraction in those with VT, this did not reach significance, and none of the patients with VT were found to have an ejection fraction of <30%. This should not be interpreted to mean that patients with Fontan palliation and low ejection fractions are at low risk for VT. Patients with single-ventricle anatomy may be vulnerable to VT with ejection fractions higher than associated with increased risk in adult heart failure populations. A survival bias may have a particularly strong effect in this subset of patients: it may be that patients with a Fontan, low ejection fraction, and VT did not survive to be included in this cohort.

Study limitations. The cross-sectional nature of this study limits the chronological data that are available, and thus, causality cannot be shown. All arrhythmia interpretation was conducted at the individual centers, with local electrophysiologists. All participating patients were cared for at tertiary care centers; however, this is the case for the majority of Fontan patients, and thus external validity can be expected to be reasonable. Due to the cross-sectional design of this study and a minimum enrollment age of 6 years, inferences from this study are subject to survivor bias; some associations may be attenuated because patients with the most severe course could not contribute to analysis. Finally, as noted previously, we had limited power to detect differences by type of Fontan operation for events potentially occurring more than 6 years post-Fontan, because the subgroups undergoing lateral tunnel and extracardiac conduit procedures had shorter follow-up than the patients who underwent an atriopulmonary connection. Furthermore, the number of patients receiving an extracardiac conduit was relatively small, one-quarter the size of the cohort undergoing intracardiac lateral tunnel Fontan, rendering 80% power to detect hazard ratios for IART of 2.5; therefore, our

observed hazard ratio of 1.6 may have been not statistically significant due to power.

Conclusions

This contemporary cohort of Fontan survivors (mean 8.6 years post-procedure) represents one of the largest datasets available in this unique population. Overall prevalence of IART (7%) is lower in the current cohort than in previous reports. Independent associated factors of IART development include a paced rhythm, lower functional status, and an atriopulmonary connection Fontan, a previously suspected risk factor for atrial tachycardia. In our study, the atriopulmonary connection does confer a higher risk of IART; however, some of that association is explained by the older age of the patients at the time of the study. We observed no significant difference in time to development of IART between the lateral tunnel and extracardiac types of surgery in this study.

Reprint requests and correspondence: Dr. Elizabeth A. Stephenson, The Hospital for Sick Children, Pediatrics, Division of Cardiology, 555 University Avenue, Toronto, Ontario M5G 1X8, Canada. E-mail: Elizabeth.stephenson@gmail.com.

REFERENCES

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971;26:240–8.
2. Gelatt M, Hamilton RM, McCrindle BW, et al. Risk factors for atrial tachyarrhythmias after the Fontan operation. J Am Coll Cardiol 1994;24:1735–41.
3. Gardiner HM, Dhillon R, Bull C, de Leval MR, Deanfield JE. Prospective study of the incidence and determinants of arrhythmia after total cavopulmonary connection. Circulation 1996;94: II17–21.
4. Triedman JK, Saul JP, Weindling SN, Walsh EP. Radiofrequency ablation of intra-atrial reentrant tachycardia after surgical palliation of congenital heart disease. Circulation 1995;91:707–14.
5. Gentles TL, Mayer JE Jr., Gauvreau K, et al. Fontan operation in five hundred consecutive patients: factors influencing early and late outcome. J Thorac Cardiovasc Surg 1997;114:376–91.
6. Blaufox AD, Sleeper LA, Bradley DJ, et al. Functional status, heart rate, and rhythm abnormalities in 521 Fontan patients 6 to 18 years of age. J Thorac Cardiovasc Surg 2008;136:100–7.
7. Fishberger SB, Wernovsky G, Gentles TL, et al. Factors that influence the development of atrial flutter after the Fontan operation. J Thorac Cardiovasc Surg 1997;113:80–6.
8. Durongpisitkul K, Porter CJ, Cetta F, et al. Predictors of early- and late-onset supraventricular tachyarrhythmias after Fontan operation. Circulation 1998;98:1099–107.
9. Sleeper LA, Anderson P, Hsu DT, et al. Design of a large cross-sectional study to facilitate future clinical trials in children with the Fontan palliation. Am Heart J 2006;152:427–33.
10. McCrindle BW, Williams RV, Mitchell PD, et al. Relationship of patient and medical characteristics to health status in children and adolescents after the Fontan procedure. Circulation 2006;113:1123–9.
11. Anderson PA, Sleeper LA, Mahony L, et al. Contemporary outcomes after the Fontan procedure: a Pediatric Heart Network multicenter study. J Am Coll Cardiol 2008;52:85–98.
12. Davignon A, Rautaharju P, Boisselle E, et al. Normal ECG standards for infants and children. Pediatr Cardiol 1979;1:123–31.
13. Sluysmans T, Colan SD. Theoretical and empirical derivation of cardiovascular allometric relationships in children. J Appl Physiol 2005;99:445–57.
14. Landgraf JM, Abetz L, Ware JE. The Child Health Questionnaire (CHQ) User's Manual. Boston, MA: Health Act, 1999.
15. Muller HG, Wang JL. Hazard rate estimation under random censoring with varying kernels and bandwidths. Biometrics 1994;50:61–76.
16. Fishberger SB, Wernovsky G, Gentles TL, et al. Long-term outcome in patients with pacemakers following the Fontan operation. Am J Cardiol 1996;77:887–9.
17. Freedom RM, Hamilton R, Yoo SJ, et al. The Fontan procedure: analysis of cohorts and late complications. Cardiol Young 2000;10:307–31.
18. Gewillig M, Wyse RK, de Leval MR, Deanfield JE. Early and late arrhythmias after the Fontan operation: predisposing factors and clinical consequences. Br Heart J 1992;67:72–9.
19. Ghai A, Harris L, Harrison DA, Webb GD, Siu SC. Outcomes of late atrial tachyarrhythmias in adults after the Fontan operation. J Am Coll Cardiol 2001;37:585–92.
20. Driscoll DJ, Offord KP, Feldt RH, et al. Five- to fifteen-year follow-up after Fontan operation. Circulation 1992;85:469–96.
21. Petrossian E, Reddy VM, McElhinney DB, et al. Early results of the extracardiac conduit Fontan operation. J Thorac Cardiovasc Surg 1999;117:688–96.
22. Marcelletti CF, Hanley FL, Mavroudis C, et al. Revision of previous Fontan connections to total extracardiac cavopulmonary anastomosis: s multicenter experience. J Thorac Cardiovasc Surg 2000;119:340–6.
23. Azakie A, McCrindle BW, Van Arsdell G, et al. Extracardiac conduit versus lateral tunnel cavopulmonary connections at a single institution: impact on outcomes. J Thorac Cardiovasc Surg 2001;122:1219–28.

Key Words: arrhythmia ▪ congenital heart disease ▪ Fontan ▪ intra-atrial re-entrant tachycardia ▪ prevalence.

APPENDIX

For a complete list of the authors and participating centers, please see the online version of this article.

JACC Vol. 28, No. 3
September 1996:763–7

Pregnancy Outcomes After the Fontan Repair

MARY M. CANOBBIO, MN, RN, DOUGLAS D. MAIR, MD,* MARY VAN DER VELDE, MD,†
BRIAN J. KOOS, MD, DPHIL‡

Los Angeles, California; Rochester, Minnesota; and Boston, Massachusetts

Objectives. This study sought to determine risks and outcome of pregnancy and delivery after the modified Fontan operation.

Background. Increasingly, female Fontan patients reaching child-bearing years are interested in having children. To date, the number of reported pregnancies is small, and pregnancy has therefore been discouraged.

Methods. One hundred ten of 126 female patients from the Fontan registries of the Mayo Clinic and University of California Los Angeles Medical Center responded to a mailed questionnaire. An additional six patients with a reported pregnancy from other centers were identified and reviewed to assess pregnancy outcomes.

Results. Among the participating centers, a total of 33 pregnancies after Fontan operation for various types of univentricular heart disease were reported. There were 15 (45%) live births from 14 mothers, with 13 spontaneous abortions and 5 elective terminations. In the 14 women with live births, the median number of years between operation and pregnancy was 4 (range 2 to 14). Reported prepregnancy problems in these gravidas included atrial flutter in one patient and ventricular dysfunction, aortic regurgitation and atrioventricular valve regurgitation in another. One patient developed supraventricular tachycardia during pregnancy and had conversion to sinus rhythm. No maternal cardiac complications were reported during labor, delivery or the immediate puerperium. There were six female and nine male infants (mean gestational age 36.5 weeks; median weight 2,344 g). One infant had an atrial septal defect. At follow-up, mothers and infants were alive and well.

Conclusions. Pregnancy after the Fontan operation appears to have been well tolerated in 13 of 14 gravidas. There does appear to be an increased risk of miscarriage. The tendency to routinely discourage pregnancy may need to be reconsidered.

(J Am Coll Cardiol 1996;28:763–7)

The Fontan and now the modified Fontan procedures have become recognized as the most definitive operation available for a number of complex congenital heart defects characterized by a functional single ventricle. Increasing numbers of postoperative women are now reaching their childbearing years, and many desire to have children. However, to date, the number of reported pregnancies and live births in this population has been small (1–7). Because of the unlikelihood of any one center having numerous pregnancies, it has been difficult to determine the true risk of pregnancy and delivery to the post-Fontan patient and her infant. A multicenter study was therefore undertaken to determine the pregnancy outcomes after modified Fontan operation and to determine the risk of pregnancy and delivery to the post-Fontan female patient and her infant.

From the University of California Los Angeles School of Nursing, Los Angeles, California; *Department of Pediatric Cardiology, Mayo Clinic, Rochester, Minnesota; †Department of Cardiology, Children's Hospital, Boston, Massachusetts; and ‡Department of Obstetrics and Gynecology, University of California Los Angeles Medical Center, Los Angeles, California. This study was presented at the 66th Annual Scientific Sessions of the American Heart Association, Atlanta, Georgia, November 1993.

Manuscript received February 16, 1995; revised manuscript received April 17, 1996, accepted May 3, 1996.

Address for correspondence: Ms. Mary M. Canobbio, University of California Los Angeles School of Nursing, 700 Tiverton Avenue, Box 951702, Los Angeles, California 90095-1702.

Published by Elsevier Science Inc.

0735-1097/96/$15.00
PII S0735-1097(96)00234-3

Methods

Patients. The study cohort was drawn in part from the Fontan registries of the Mayo Clinic and the University of California Los Angeles (UCLA) Medical Center. Post-Fontan female patients who had reached menarche were asked to respond to a mailed questionnaire designed to determine pregnancy and contraceptive experiences after modified Fontan repair. Of the 126 eligible female patients, 110 responded, with 16 nonresponders who were lost to follow-up. Additionally, six centers (see Appendix) with Fontan pregnancies were invited to participate. For subjects who reported a live birth, the medical records and, where possible, the obstetric records were reviewed.

Results

This collaborative effort identified 21 of 118 subjects as having a total of 33 pregnancies after the Fontan procedure. Fifteen (45%, 95% confidence interval [CI] 28% to 60%) pregnancies resulted in live births, 5 (15%, 95% CI 6% to 29%) in elected therapeutic abortions for noncardiac reasons and 13 (39%, 95% CI 23% to 54%) in spontaneous abortions during the first trimester. One woman was taking the fertility drug clomiphene citrate and had three spontaneous miscarriages. If this patient is removed from the series, the first-trimester miscarriage rate was 33% (95% CI 18% to 49%).

Table 1. Diagnosis and Surgical History of 14 Post-Fontan Mothers

Pt No.	Defect	Type of Connection	Age at Operation (yr)	Postoperative Complications Before Pregnancy	Postoperative Medications During Pregnancy
1	T. atresia	Atriopulmonary anastomosis	22	Atrial flutter	Digoxin, quinidine
2	T. atresia	Atriopulmonary anastomosis	14	None	Digoxin
3	T. atresia	Atriopulmonary anastomosis	15	None	None
4	T. atresia	RA-RV Hancock conduit	24	None	None
5	DILV	#1 RA-RV Hancock conduit	23		
		#2 Atriopulmonary anastomosis	32	None	None
6	T. atresia	Atriopulmonary anastomosis	26	None	None
7	T. atresia	RA-RV Hancock conduit	14	None	None
8	DILV	Atrial pulmonary Hancock conduit	11	None	None
9	T. atresia	#1 RA-RV Hancock conduit	5		
		#2 RA-RV Carpentier conduit	10	None	None
10	DORV/hypoplastic RV	Cavopulmonary anastomosis	22	None	None
11	TGA/straddling TV/hypoplastic RV	#1 Atriopulmonary anastomosis	14		
		#2 Redo patch closure RAVV	15	None	None
12	DORV/hypoplastic RV	Cavopulmonary anastomosis	21	AVVI, LV dysfunction	Quinidine
13	DORV/hypoplastic RV, polysplenia	Cavopulmonary anastomosis	18	Periodic SVT	Digoxin
14	UVH of LV type with right AVV atresia	Atriopulmonary anastomosis	17	Atrial flutter with 1:1 conduction; ventricular rate 265 beats/min	Tenormin/digoxin

AI = aortic valve insufficiency; AVVI = atrioventricular valve insufficiency; DIRV = double-inlet left ventricle; DORV = double-outlet right ventricle; LV = lef ventricle; Pt = patient; RA-RV = right atrial to right ventricle; RAVV = right atrioventricular valve patch closure; RV = right ventricle; SVT = supraventricula tachycardia; T = tricuspid; TGA = transposition of the great arteries; TV = tricuspid valve; UVH = univentricular heart.

Fourteen gravidas carried their pregnancies to a live birth, with 1 having two successful pregnancy outcomes. These 14 women and their 15 children form the study cohort for the present report.

Cardiac anomaly and surgical history. The median age at Fontan operation of patients subsequently becoming pregnant was 17 years (range 10 to 32). Diagnoses and surgical history of the patients are summarized in Table 1. Two patients (Patients 5 and 9) required reoperation after their original Fontan procedure. Postoperative complications also are listed in Table 1. Atrial arrhythmias after the Fontan operation were reported in four patients. One patient (Patient 1) developed atrial flutter that required treatment initially with digoxin (0.125 mg twice daily) followed by cardioversion and then quinidine (300 mg three times daily) (2). At the time of her pregnancy she was in sinus rhythm. Patient 12 was given quinidine preoperatively and postoperatively for supraventricular and ventricular tachycardia. Patient 13 had periodic supraventricular tachycardia that has been controlled with digoxin; Patient 14 was treated for atrial flutter with intermittent 1:1 conduction and a ventricular rate of 265 beats/min. The remaining patients were reported to be free of significant heart rate abnormalities. Apart from arrhythmias, the only other reported post-Fontan cardiac problem involved Patient 12. At the time of her pregnancy she was known to have mild aortic regurgitation, atrioventricular (AV) valve regurgitation and some degree of ventricular dysfunction.

Pregnancy history. At the time of pregnancy the 14 study subjects ranged in age from 18 to 36 years (median age 24). These 14 patients had 25 of the total 33 pregnancies. Pregnancy, for the 15 live births, occurred at a median of 4 year (range 2 to 13) after the Fontan procedure.

The median gestational age of the newborns was 38 week (range 28 to 40). Footling breech presentation, failure t progress and preterm labor at 28 weeks were the obstetri indications for 3 of the 11 cesarean sections. For the remainin eight, the reason for cesarean delivery was unclear from revie of the obstetric records. Epidural anesthesia was used for 1 deliveries, spinal for 2 and general anesthesia for 1. On woman delivered by natural childbirth. The anesthesia metho for the remaining delivery is unknown.

Cardiac complications. For 12 of the 14 mothers, ther were no significant cardiac complications reported throughou the antepartum, labor, delivery or postpartum period. Patier 7 developed supraventricular tachycardia in her 27th gest tional week, requiring hospital admission. She was started o digoxin (0.25 mg/day) and verapamil (80 mg/day) therapy. A 38.5 weeks, she developed recurrent supraventricular tachyca dia with a heart rate of 180 beats/min and complained weakness and breathlessness. She had conversion to sinu rhythm with 5 mg of intravenous verapamil. Patient 12 repor edly complained of mild shortness of breath throughout h pregnancy and immediately after delivery. She was treated wi oxygen for 1 week and discharged. Subsequently, she w treated for congestive heart failure with afterload-reducir agents. There were no other reported cardiac complicatio during the postpartum period. No cardiac medications we initiated during any of the other pregnancies. However Patient 1, the quinidine dose was increased from three to fo times per day during the pregnancy. Two patients complain

of periodic palpitations during the pregnancy, but no treatment was necessary.

Noncardiac complications. Peripheral edema occurred in two patients. Patient 6 had spontaneous premature rupture of the membranes at 34 weeks and later developed an incarcerated left inguinal hernia involving the left ovary, necessitating a herniorrhaphy. The patient recovered without further consequences.

Maternal outcomes. At 18 months after delivery, no adverse clinical sequelae were evident for 13 of the 14 women. Patient 12 has continued to be treated for ventricular failure, but at 20 months, her cardiovascular function has returned to prepregnancy status.

Infant outcomes. There were six female and nine male infants. Birth weight ranged from 1,050 to 3,575 g (median 2,344). Only one child was born prematurely at 28 weeks' gestation and weighed 1,050 g. At 4 years old, the boy has developed normally. One infant had a congenital anomaly, an atrial septal defect that has since been surgically closed. All infants are currently alive and well.

Prepregnancy counseling. Because physicians tend to advise the female Fontan patient against pregnancy, included in the UCLA/Mayo Clinic questionnaire were questions that addressed pregnancy counseling. Of the 76 patients who responded to this question, 50 (66%, 95% CI 54% to 75%) indicated that they had been advised by a physician not to become pregnant despite a clinically stable surgical outcome and a strong desire to have children. The remaining 26 patients (34%, 95% CI 23% to 44%) indicated they had not been advised about pregnancy. With respect to contraception, there were 83 responses. Thirty-two indicated that they used no form of contraception; 21 said that they used some form of barrier method; and 14 used oral contraception. Fourteen had undergone tubal ligation (seven before and seven after their Fontan procedure), and two had a hysterectomy after their Fontan procedure.

Discussion

The goals of the original Fontan procedure (8) and its subsequent modifications have been to eliminate intermixing of pulmonary and systemic venous return, thereby achieving a normal oxygen tension in the systemic arterial circulation, and normalizing the volume load on the functional single ventricle. These goals can be accomplished only at the price of elevated systemic venous and right atrial pressures. In the absence of a ventricle pumping to the lungs, a minimal pressure of 12 to 14 mm Hg is necessary in the right atrium and systemic veins to achieve an adequate driving pressure across the pulmonary capillary bed (9). Fontan patients with a mean right atrial pressure ≥20 mm Hg immediately after repair have been reported to have an increased surgical mortality and late morbidity (9,10). Late postoperative complications associated with the Fontan procedure include arrhythmias, protein-losing enteropathy and thromboembolic events, with atrial arrhythmias the most frequently cited (11). Probably due to persistent elevated right atrial pressure and increased wall tension, the most prominent arrhythmias have been supraventricular tachycardia and atrial flutter or fibrillation. In most series, including ours, arrhythmias have been well controlled with standard medical therapy.

In women with congenital heart disease, pregnancy outcomes have been shown (12) to be related to functional cardiac status, anatomic diagnosis, pulmonary vascular resistance, type of operation and degree of residual impairment. In women with cyanotic heart disease, maternal mortality ranges between 4% and 16% in uncorrected lesions (13). Tetralogy of Fallot, the only cyanotic lesion for which postoperative pregnancy data are widely available, carries a postrepair pregnancy risk similar to that of the general population (14,15). Little pregnancy data are available for other corrected cyanotic lesions.

For post-Fontan patients, questions have focused on the ability of the right atrium and functioning single ventricle to generate and tolerate the normal cardiovascular adjustment to pregnancy, including increases in cardiac output (30% to 40% above nonpregnant levels), circulating volume (up to 50%) and heart rate that normally occur during pregnancy. Because the post-Fontan patient has limited ability to increase cardiac output despite an elevated venous pressure, concern has been raised about her ability to cope with the increased physiologic demands of pregnancy and delivery. The present study and others report a total of 22 term pregnancies. Among these, there have been no reported maternal deaths or serious morbidity. There have been two reported neonatal deaths due to prematurity (4,5). In both cases the mothers developed arrhythmias during pregnancy. In one case the mother had persistent junctional tachycardia (4), and in the other the mother developed atrial flutter with 2:1 AV block (5). The current series has the only reported case of congenital heart disease in the offspring.

Although the number of cases is still small, it appears that the post-Fontan patient who is clinically well before pregnancy is able to tolerate the additional hemodynamic burden of a term pregnancy and delivery and to return to her prepregnancy level of function once her convalescence is complete. However, this same experience suggests that the incidence of first-trimester spontaneous abortion in post-Fontan patients (33%) appears to be twice that in the general population (15%) (16).

Still unknown is whether pregnancy will have any long-term residual effect on a myocardium previously exposed to years of hypoxia and volume overload. With Fontan surgery being performed at younger ages, these effects on the functional single ventricle may be less important in the future.

For the infant, the risk of congenital heart disease remains unknown. In the present series, the incidence rate was 6%, which is within the 3% to 8% range often quoted for infants born to women with congenital heart disease (17,18). Advances in fetal echocardiography, including transvaginal echocardiography, should eventually permit identification of most major fetal anomalies during the first trimester (19).

This clinical experience suggests that women who are in clinically stable condition after the Fontan operation wil

tolerate pregnancy and delivery. A woman who is doing well and desires to have a child should not be routinely discouraged from doing so. Rather, she should be made aware of the potential risk to her and her child, including the possible increased incidence of first-trimester miscarriage. To provide the mother with accurate information about the risk that a pregnancy may pose, a comprehensive evaluation, including electrocardiography and Doppler echocardiography to evaluate ventricular function and AV valve competency and to check patency of the right-sided Fontan pathway, should be performed before pregnancy. If potential concerns are identified by Doppler echocardiography, a short cardiac catheterization to provide more precise information, including cardiac output and right-sided pressures, may be indicated.

Contraception. Prepregnancy counseling should be an integral part of routine cardiac follow-up care. Patients should be advised of the importance of planning a pregnancy and counseled about appropriate forms of contraception. In the UCLA/Mayo study, 38% (95% CI 27% to 49%) of the respondents reported using no form of birth control, whereas an additional 14% (95% CI 7% to 23%) used oral contraception. Because of the increased potential for intracardiac thrombus formation, estrogen-based contraception is contraindicated (20). Although the risk of infective endocarditis in post-Fontan patients is low, use of intrauterine devices may still be ill-advised. Combined barrier methods (i.e., diaphragm with spermicide), controlled-release progestin (Norplant) or Depo-Provera are probably the safest contraceptives for these patients (21).

Management. A pregnant post-Fontan patient requires collaborative management, including a cardiologist, a high risk obstetrician and anesthesiologist familiar with adult congenital heart disease and the cardiovascular changes of pregnancy and parturition. Ideally, the gravida should be followed up at a specialized center that has experience both with post-Fontan patients and high risk pregnancy. Peripheral edema, breathlessness and fatigue are common findings in normal pregnancy and do not necessarily indicate cardiac decompensation (22). Because of their limited cardiac reserve, it would be prudent for these patients to avoid strenuous exercise (such as heavy lifting), to take daily rest periods and to limit their salt intake.

Observed maternal cardiovascular changes occurring during labor and delivery pose additional risk. For example, during the first stage of labor, cardiac output normally increases by ~25% between uterine contractions, with further elevations during contractions. However, because of compression of the inferior vena cava by the gravid uterus, the supine position at term gestation decreases stroke volume and cardiac output by ~25% and may result in maternal bradycardia and hypotension (23). To minimize these effects, it is important to have the gravida labor in a lateral position. Epidural anesthesia is recommended because it promotes hemodynamic stability by reducing pain-related increases in sympathetic activity. However, epidural anesthesia should be used cautiously, or not at all, if cardiac output is suspected to be unusually sensitive to decreases in preload. In such high risk patients, narcotic epidural and continuous narcotic spinal anesthesia are a good alternative during labor, with general anesthesia reserved for cesarean delivery. A vaginal delivery should be expected with an assisted second stage of labor so as to avoid the reduction of venous return caused maternal Valsalva maneuvers. Cesarean delivery is reserved for obstetric indications (23,24). Immediately after delivery, patients should be closely monitored because cardiac output in normal women increases by as much as 75% over predelivery values (25). This increase is of particular concern in women who may not tolerate volume shifts or who are unable to increase their cardiac output in response to increased demand (26). Therefore, volume shifts should be monitored carefully and the mother left on her left side during the immediate postpartum period. The effect may persist for ~1 week but returns toward normal within 2 weeks (23–25).

Study limitations. To our knowledge, this is the largest clinical series reported on pregnancy outcomes in post-Fontan patients; however, certain limitations need to be considered. Patient selection was a sample of convenience and is representative, therefore, only of post-Fontan patients followed up principally at a few centers; there are undoubtedly other patients who have become pregnant and have not been identified by this relatively informal sampling. In turn, if some of these patients had developed complications, the overall outlook is less favorable. Another potential bias is that for logistic reasons, obstetric care data were obtained through correspondence and often relied on the memory of the obstetrician or cardiologist rather than direct inspection of the medical record.

Another limitation is that little is known of patient hemodynamic status after the Fontan procedure and before pregnancy; data were not consistently available, in part because several pregnancies were not planned, and in part because hemodynamic studies are not part of routine follow-up care. Optimally, however, a clinical and hemodynamic baseline of functional status would be established before conception. Finally, the numbers of patients in the present study are too few to predict safety; however, with one exception all patients were in functionally stable condition before pregnancy.

Conclusions. The number of female post-Fontan patients reaching adulthood and desiring to have children will continue to increase. These women are limited in their ability to increase cardiac output in response to exertion or the increased physiologic demands of pregnancy and have generally been counseled by their physicians to avoid pregnancy. A larger patient series and longer follow-up periods are clearly needed before the risk of pregnancy and delivery to these patients and their infants can be precisely defined; based on our experiences with 14 patients, however, it seems unreasonable to prohibit pregnancy in the patient who has a good postoperative result. Such patients should be made aware of the clinical experiences to date and the potential risks to them and their fetus, including the possibility of a reported increased risk for first-trimester abortion, which will help them to make an informed decision regarding pregnancy.

Appendix

Participating Centers and Investigators

Jack A. Colman, MD, *Mount Sinai Hospital, Toronto, Ontario, Canada;* Thomas G. Di Sessa, MD, *Department of Cardiology, University of Tennessee, Le Bonheur Children's Medical Center, Memphis, Tennessee;* Derek A. Fyfe, MD, PhD, *Emory University School of Medicine, Children's Heart Center, Atlanta, Georgia;* Michelle Moss, MD, *University of Arkansas, Little Rock, Arkansas;* William B. Strong, MD, *Medical College of Georgia, Augusta, Georgia;* Mary van der Velde, MD, *Children's Hospital of Boston, Boston, Massachusetts.*

References

1. Carmona F, Martinez S, Periz A, Cararach. Pregnancy after surgical correction of tricuspid atresia. Acta Obstet Gynecol Scand 1993;72:498–9.
2. Fyfe DA, Gillette PC, Jones JS, Danielson GK. Successful pregnancy following modified Fontan procedure in a patient with tricuspid atresia and recurrent atrial flutter. Am Heart J 1989;117:1387–8.
3. Girod DA, Fontan F, Deville C, et al. Long-term results after the Fontan operation for tricuspid atresia. Circulation 1987;75:605–10.
4. Hess DB, Hess LW, Heath BJ, et al. Pregnancy after Fontan repair of tricuspid atresia. South Med J 1991;84:532–4.
5. Cohen AM, Mulvein J. Obstetric anesthesia management in a patient with Fontan circulation. Br J Anesth 1994;73:252–5.
6. Carp H, Jayaran A, Vadhera R, Nichols M. Epidural anesthesia for Cesarean delivery and vaginal birth after maternal Fontan repair: report of two cases. Anesth Analg 1994;78:1190–2.
7. Gerardin B, Houyel L, Discazeaux B, et al. Successful pregnancy after Fontan complicated by recurrent junctional tachycardia. Arch Mal Coeur 1993;86:935–8.
8. Fontan F, Baudet E. Surgical repair for tricuspid atresia. Thorax 1971;26: 240–8.
9. Mair DD, Puga FJ, Danielson GK. Late functional status of survivors of the Fontan procedure performed during the 1970's. Circulation 1992;86 Suppl II:II-106–9.
10. Driscoll DJ, Offord KP, Feldt RH, et al. Five-to-fifteen-year follow-up after Fontan operation. Circulation 1992;85:469–6.
11. Mair DD, Hagler DT, Julsrud PR. Early and late results of the modified Fontan procedure for double-inlet left ventricle. The Mayo Clinic experience. J Am Coll Cardiol 1991;18:1727–32.
12. Whittemore R, Hobbins JC, Engle MA. Pregnancy and its outcome in women with and without surgical treatment of congenital heart disease. Am J Cardiol 1982;50:641–51.
13. Shime J, Mocarski EJM, Hastings D, Webb GD. Congenital heart disease in pregnancy: Short and long-term implications. Am J Obstet Gynecol 1987; 156:313–21.
14. Ralston JH, Dunn M. Pregnancies after surgical correction of tetralogy of Fallot. JAMA 1976;235:2627–8.
15. Singh H, Ballon PJ, Oakley CM. Pregnancy after surgical correction of tetralogy of Fallot. BMJ 1982;285:106–8.
16. Cavanagh D, Comas MR. Spontaneous abortion. In: Danforth D, editor. Obstetrics and Gynecology, 6th ed. Philadelphia: Lippincott, 1994:378–94.
17. Nora JJ, Nora AH. Maternal transmission of congenital heart disease: new recurrence risk figure and the question of cytoplasmic inheritance and vulnerability to teratogens. Am J Cardiol 1988;59:459–63.
18. Whittemore R. Congenital heart disease: its impact on pregnancy. Hosp Pract 1983;18:65–8.
19. Fyfe DA, Kline CH. Fetal echocardiographic diagnosis of congenital heart disease. Pediat Clin N Am 1990;37:45–65.
20. Jahangiri M, Ross DB, Redington AN, et al. Thromboembolism after the Fontan procedure and its modifications. Ann Thorac Surg 1994;58:1409–14.
21. Craig S, Hepburn S. The effectiveness of barrier methods of contraception with and without spermicide. Contraception 1982;26:347–56.
22. Hytten Ef, Thomson AM, Taggart N. Total body water in normal pregnancy. J Obstet Gynaecol Br Commonw 1966;73:553–61.
23. Ueland K, Hansen PM. Maternal cardiovascular dynamics III. Labor and delivery under local and caudal anesthesia. Am J Obstet Gynecol 1969;103: 8–18.
24. Metcalfe J, Ueland K: Maternal cardiovascular adjustments to pregnancy. Prog Cardiovas Dis 1974;16:363–74.
25. Ueland K, Hansen JM. Maternal cardiovascular dynamics III. Labor and delivery under local and caudal analgesia. Am J Obstet Gynecol 1969;103: 8–18.
26. Gewellig MH, Lundstrom UR, Bull C, et al. Exercise responses in patients with congenital heart disease after Fontan repair: patterns and determinants of performance. J Am Coll Cardiol 1990;15:1424–32.

Thorax (1971), **26**, 240.

Surgical repair of tricuspid atresia

F. FONTAN and E. BAUDET

Centre de Cardiologie, Université de Bordeaux II, Hôpital du Tondu, Bordeaux, France

Surgical repair of tricuspid atresia has been carried out in three patients ; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, 'ventriclized', to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.

Only palliative operations (systemic vein to pulmonary artery anastomosis ; systemic artery to pulmonary artery anastomosis) have been performed in tricuspid atresia. Although these procedures are valuable, they result in only a partial clinical improvement, because they do not suppress the mixture of venous and oxygenated blood.

We have initiated a corrective procedure for tricuspid atresia, which completely suppresses blood mixing. The entire vena caval return undergoes arterialization in the lungs and only oxygenated blood comes back to the left heart. This procedure is not an anatomical correction, which would require the creation of a right ventricle, but a procedure of physiological pulmonary blood flow restoration, with suppression of right and

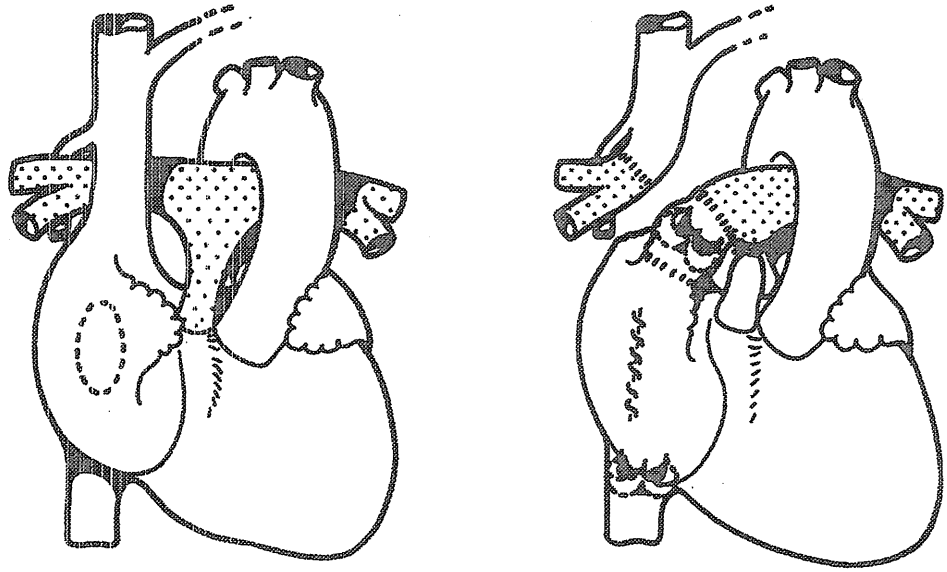

FIG. 1. *Case 2. Tricuspid atresia type II B. Drawing illustrates steps in surgical repair:* (1) *end-to-side anastomosis of distal end of right pulmonary artery to superior vena cava;* (2) *end-to-end anastomosis of right atrial appendage to proximal end of right pulmonary artery by means of an aortic valve homograft;* (3) *closure of atrial septal defect;* (4) *insertion of a pulmonary valve homograft into inferior vena cava; and* (5) *ligation of main pulmonary artery.*

left blood mixing. This new surgical procedure has been used in three patients and has been successful in two of them; the first case has been followed satisfactorily for 30 months. The indications for this procedure apply only to children who are sufficiently well developed, without pulmonary arterial hypertension.

Palliative operations remain valuable in other patients and will permit many of them to have a secondary corrective procedure.

SURGICAL TECHNIQUE

The purpose of the operation is to drain the whole vena caval blood to the pulmonary arteries (Fig. 1): the superior vena cava is anastomosed to the distal end of the right pulmonary artery, according to Glenn's procedure; the proximal end of the right pulmonary artery is anastomosed to the right atrium; so, after the atrial septal defect has been closed, the blood of the inferior vena cava is drained towards the left pulmonary artery. The main pulmonary artery is ligated at the point where it leaves the right hypoplastic ventricle, to prevent ventricular blood entering the left lung. In short, the right atrium is used to propel inferior vena caval blood through the left lung. To facilitate this function, the right atrium is provided with two aortic or pulmonary valve homografts: one is inserted into the inferior vena cava at its junction with the right atrium, to prevent blood reflux into the inferior vena cava during atrial systole; the other is used as an anastomosis between the right atrial appendage and the proximal end of the right pulmonary artery, so that, during atrial diastole, there is no reflux from the left pulmonary artery into the right atrium.

The operation is performed through a median sternotomy. After the pericardium has been opened, the heart is examined to confirm the preoperative diagnosis of tricuspid atresia type. The pulmonary arteries also have to be examined carefully to ensure that their size is large enough to permit a cava-pulmonary anastomosis. In addition, it is necessary to measure the pressures in the pulmonary artery, thus making sure that there is no pulmonary arterial hypertension, which would be a contraindication to cava-pulmonary anastomosis. This information, suspected from catheterization and angiocardiography, can only be corroborated during operation.

The surgical repair begins with the classic cava-pulmonary anastomosis between the distal end of the right pulmonary artery and the right posterolateral aspect of the superior vena cava. End-to-side anastomosis is made, using a Blalock continuous suture (Fig. 2). But the superior vena cava is not yet transected at its entry into the right atrium, because it must be used for superior vena caval cannulation during cardiopulmonary bypass. This transection must be carried out as the last step of the operation.

The proximal end of the right pulmonary artery is then anastomosed to the right atrium by means of an aortic valve homograft (Fig. 3); the aortic wall is tailored to an adequate length; the origin of the right pulmonary artery can also be enlarged by bevelling up to the main pulmonary artery in order to achieve a good fit with the homograft (Fig. 2). End-to-end anastomosis is made using a continuous suture. The homograft (a short segment of the anterior mitral leaflet and septum below the aortic cusps has been kept) is end-to-end anastomosed to the right atrial appendage. There is no problem of fit with the atrial appendage which is, in tricuspid atresia, widely dilated, but fleshy tissues in the atrial appendage should be resected so that they do not hinder blood flow. Such a homograft was used in our second and third cases. In the first case, a younger child, we did not have a small enough homograft. We anastomosed the proximal end of the right pulmonary artery directly to the left lateral side of the upper part of the right atrium (Figs 7 and 8).

The operation then proceeds under cardiopulmonary bypass (Fig. 4), at flow rates of 2 to 2·2 litres/min/m^2 at normothermia. The duration of cardiopulmonary bypass is about 40 minutes. The ascending

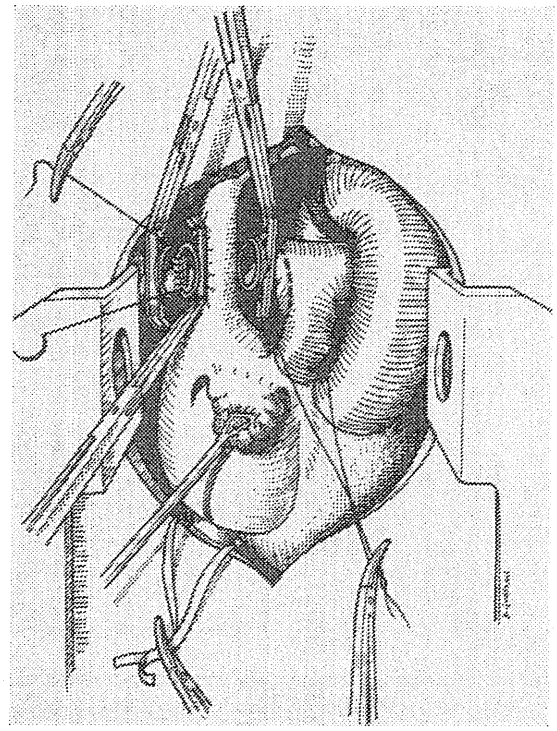

FIG. 2. *First step of the repair: end-to-side anastomosis of distal end of right pulmonary artery to superior vena cava (Glenn, 1958). Drawing illustrates bevelling of proximal end of right pulmonary artery to ensure a correct fit with the aortic valve homograft (see Fig. 3).*

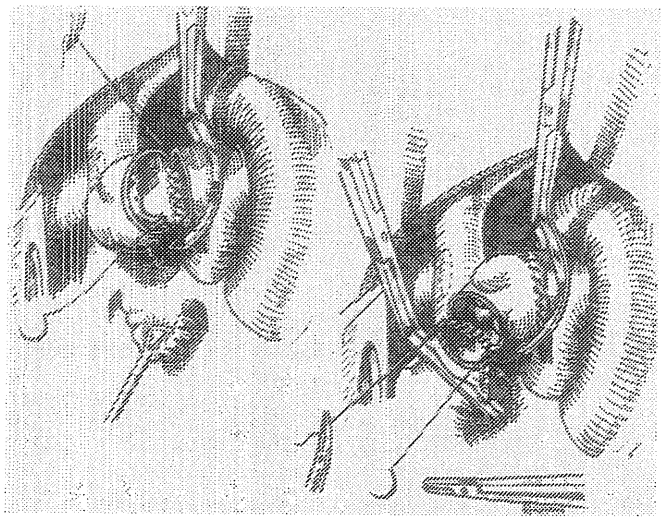

FIG. 3. *Second step of the repair: end-to-end anastomosis of right atrial appendage to proximal end of right pulmonary artery by means of an aortic valve homograft. Drawing illustrates superior vena cava to right pulmonary artery anastomosis, but superior vena cava is not yet transected at its entry into right atrium because it must be used for superior vena caval cannulation during cardiopulmonary bypass.*

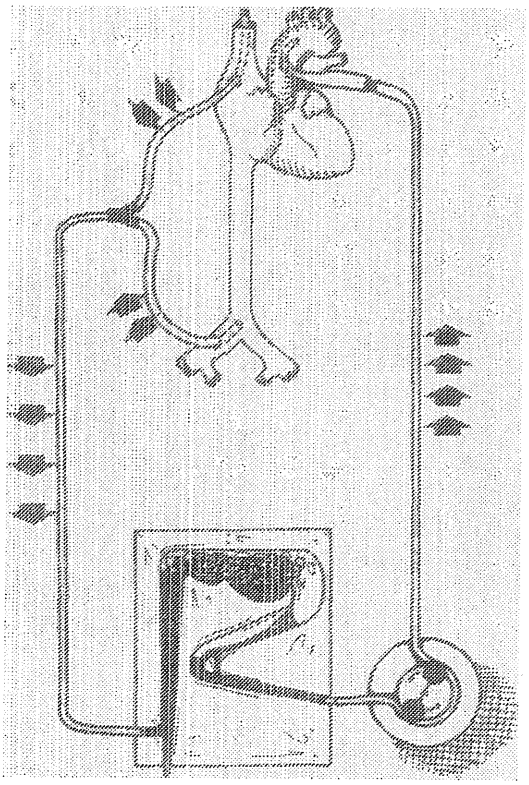

FIG. 4. *Sketch of cardiopulmonary bypass, with cannulation of ascending aorta, superior vena cava, and right iliac vein.*

aorta is cannulated; the superior vena cava is cannulated through a purse-string suture slipped on between the right atrial appendage and the superior vena cava. The inferior vena cava is cannulated by means of the right external iliac vein so that the catheter does not prevent the insertion of the valve homograft into the inferior vena cava level with its junction with the right atrium. When the bypass is started, the superior vena cava is snared by an umbilical tape above the right cava-pulmonary anastomosis and the inferior vena cava is clamped just below its entry into the right atrium. The left ventricle is vented. The aorta is cross-clamped. After the right atrium has been opened (Fig. 5), the atrial septal defect is closed; a pulmonary valve homograft is inserted into the inferior vena caval orifice. This homograft is prepared in the following manner: the whole subvalvular tissue is resected and only 2 to 3 mm of the arterial wall above the cusps is kept to suture the homograft to the atrial wall, using a continuous suture. There is no fear of harm to the bundle of His if the suture is passed sufficiently far behind the coronary sinus.

After the atriotomy has been closed, the air evacuated, and the clamps removed, the main pulmonary artery is ligated or transected. Cardiopulmonary bypass is discontinued as soon as cardiac action is vigorous. When the systemic pressure is above 100 mmHg, the same or slightly higher pressures are looked for in the superior vena cava, the right atrium, and the pulmonary artery as were measured before bypass.

After the cannulae have been removed, the superior vena cava is transected between two clamps at its entry into the right atrium and both ends are sutured (Fig. 6).

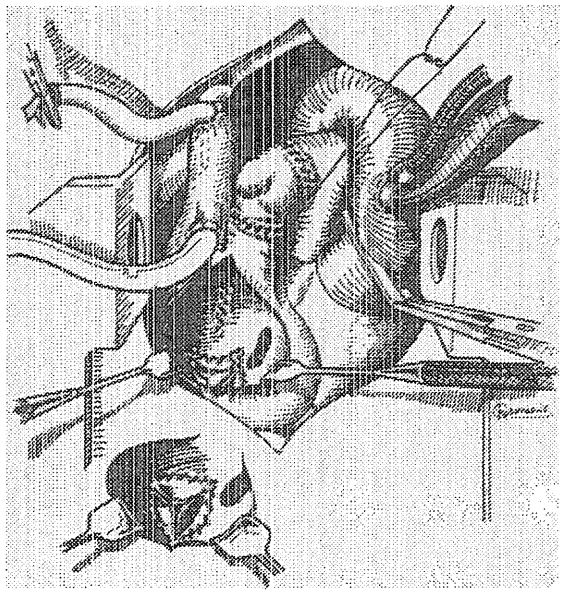

FIG. 5. *Third step of the repair, under cardiopulmonary bypass: closure of atrial septal defect and insertion of a pulmonary valve homograft into inferior vena cava level with right atrium. Superior vena cava is cannulated through a purse-string suture, slipped on between right atrial appendage and superior vena cava, and it is snared by an umbilical tape above cava-pulmonary anastomosis.*

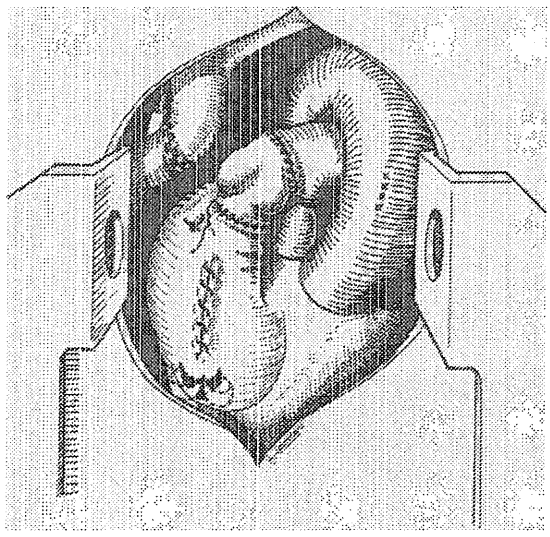

FIG. 6. *The operation at completion, after the last step of the repair: superior vena cava is transected at its entry into right atrium and main pulmonary artery is ligated.*

The pericardium is closed in the upper part without compromising the different anastomoses. The pericardial cavity and the anterior mediastinum are drained; the sternum is reapproximated with wire sutures and the subcutaneous tissue and skin are closed.

CASE REPORTS

CASE 1 Our first patient, C.F., underwent operation at the age of 12 years.

The face and extremities had been cyanosed from the age of 6 months and the cyanosis had gradually progressed; at the same time, exertional dyspnoea had appeared when she was admitted to hospital in 1961, at the age of 6 years, for haemodynamic investigations.

Cyanosis was marked. There was clubbing of the fingers and toes. A loud systolic murmur was heard at the apex, radiating along the left of the sternum, with an accentuated second heart sound. Blood pressure was 100/60 mmHg. Manifestations of cardiac failure were not noted. A blood count showed 7,000,000 RBC/mm^3.

Cardiac catheterization and angiocardiography revealed a type I B tricuspid atresia, with pulmonary arteries of good size. The possibility of a cava-pulmonary anastomosis was noted. The child left hospital and was lost sight of.

She was later readmitted to hospital in April 1968. She was very erythrocyanotic and manifested exertional symptoms—dyspnoea and frequent episodes of tachycardia. No signs of cardiac failure were noted. A blood count showed 7,800,000 RBC/mm^3 and the haematocrit was 80%.

Operation (Fig. 7) was performed on 25 April 1968 through a median sternotomy. The findings were tricuspid atresia without transposition of the great vessels (type I B) but with pulmonary arteries of good size and low intra-arterial pressure (15·0 mmHg). A superior vena cava to pulmonary artery anastomosis and an anastomosis between the atrium and the proximal end of the right pulmonary artery were carried out (Fig 8); the azygos vein was not ligated. Then, under cardiopulmonary bypass, the atrial septal defect was closed, a pulmonary valve homograft was inserted into the inferior vena cava level with the right atrium, and the main pulmonary artery was ligated. After the cardiopulmonary bypass had been discontinued, the superior vena cava was divided level with the right atrium below the cava-pulmonary anastomosis (Fig. 9).

The initial postoperative course was very satisfactory: cyanosis disappeared. The patient was in sinus rhythm at 90 per minute and the blood pressure was 120/60 mmHg. Ventilation and haematosis were satisfactory. The venous pressure was not raised.

Twenty-four hours postoperatively anuria developed quite suddenly, but metabolic disorders were corrected by one haemodialysis only, while a moderate melaena appeared. The following day urinary

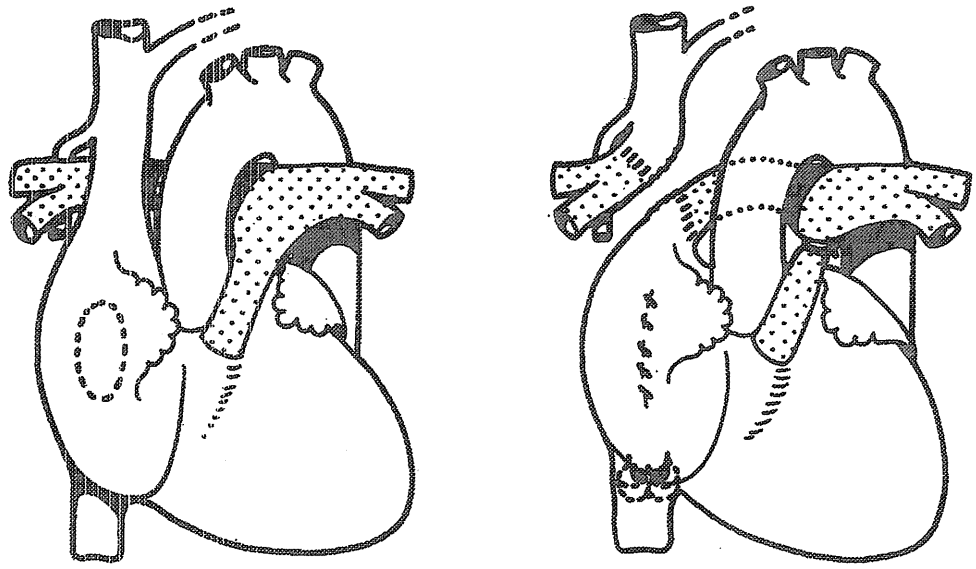

FIG. 7. *Case 1. Tricuspid atresia type I B. Drawing illustrates the repair: anastomosis between right atrium and proximal end of right pulmonary artery was made without interposition of an aortic valve homograft.*

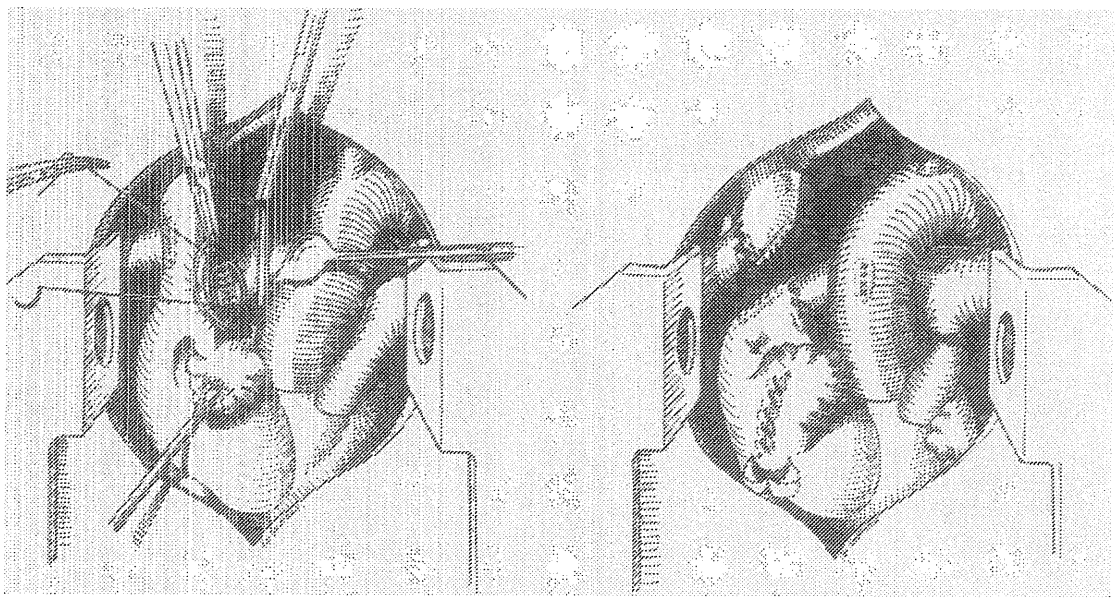

FIG. 8. *End-to-side anastomosis of proximal end of right pulmonary artery to left lateral side of upper part of right atrium.*

FIG. 9. *Case 1. Appearance at completion of operation.*

function returned, after the patient's legs had been raised to improve the stagnant inferior vena caval circulation. From that moment there was no further urinary problem, but intravenous urography revealed a bilateral congenital hydronephrosis.

One month postoperatively a right serofibrinous pleural effusion required suction drainage for a few days.

The child was examined regularly after discharge from hospital and she is quite well after a 30 months' follow-up; she has grown normally and does not show exertional symptoms; she is no longer cyanotic and there is no oedema of the inferior limbs. The liver is just palpable below the costal edge. A chest radiograph shows normal pulmonary vascularity of both lungs. An electrocardiogram indicates a regression of the right atrial hypertrophy. A blood count showed 4,400,000 RBC/mm³ and the haematocrit was 52%. Digitalis and diuretics in standard doses were prescribed. Anticoagulants were not used postoperatively. Physical examination is normal and there is no longer a systolic murmur.

The following postoperative haemodynamic and angiocardiographic investigations were made:

(1) The pressure curve in the superior vena cava fluctuated between 2·0 and 16·0 mmHg with breathing movements. On the other hand, in the inferior vena cava and the right atrium, the pressure was stable, about 15·0 mmHg (Table I).

TABLE I

	RBC per mm³	Haematocrit %	Pressure (mmHg) SVC	IVC	RA
Preoperative 6 yr	7,000,000	70		8	
Preoperative 12 yr	7,800,000	80			
Postoperative	4,400,000	52	2 to 16	15	14

(2) Right atrial angiocardiography showed that, except for a slight flow along the catheter (Figs 10 and 11), the contrast medium did not flow back from the right atrium to the inferior vena cava because the valve homograft performed its antireflux function perfectly. The valvular sinus is well seen on the angiocardiogram (lateral view). The contrast medium flowed through the left pulmonary artery and passage though the left lung was unimpeded.

CASE 2 Our second patient, J.B., was a 36-year-old woman, who had been cyanotic since she was a child but was normally developed.

At the age of 18 years she had had a cerebral abscess which was drained without sequelae.

At the age of 30 years she was referred to hospital because of exertional dyspnoea. Marked generalized cyanosis and clubbing of the fingers were noted. A very loud systolic murmur was heard to the left of the sternum, but there were no signs of cardiac failure.

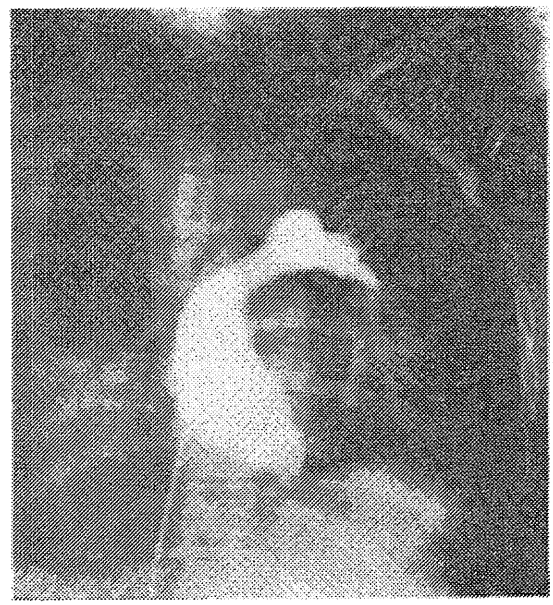

FIG. 10. *Case 1. Postoperative right atrial angiocardiogram, anteroposterior view, showing opacification of right atrium and contrast medium flowing through left pulmonary artery.*

FIG. 11. *Case 1. Postoperative right atrial angiocardiogram, anteroposterior view: passage of contrast medium through left lung is fast, as proved by good opacification of left atrium. Except for a slight flow along catheter, contrast medium does not flow back from right atrium to inferior vena cava, because the pulmonary valve homograft performs its antireflux function perfectly.*

At the age of 33 years exertional dyspnoea, nonproductive cough, and headache appeared, but they were improved by symptomatic therapy.

Finally, at the age of 36 years, her exercise tolerance was reduced and dyspnoea became progressively worse. She was admitted to hospital in January 1970.

Cardiac catheterization and angiocardiography confirmed a type IIB tricuspid atresia, with pulmonary arteries which seemed of small size when compared with an enormous transposed aorta, but the right and left branches of the pulmonary artery were, in fact, of nearly normal size.

Operation (Fig. 1) was performed on 20 January, 1970. The pulmonary arterial pressure was 35·0 mmHg. After superior vena cava to right pulmonary artery anastomosis had been carried out (Fig. 2), the superior vena caval pressure was 10·0 mmHg. Anastomosis of the right atrial appendage to the proximal end of the right pulmonary artery was carried out by means of an aortic valve homograft (Fig. 3). Under cardiopulmonary bypass (Fig. 5) the atrial septal defect was closed, the pulmonary valve homograft was inserted into the inferior vena cava, and finally the main pulmonary artery was ligated. After the cardiopulmonary bypass had been discontinued, the superior vena cava was transected below the cava-pulmonary anastomosis (Fig. 6) at its entry into the right atrium.

The initial postoperative course was very satisfactory: cyanosis disappeared; the patient was in sinus rhythm at 80 per minute and ventilation and diuresis were excellent. To maintain an adequate blood pressure it was necessary to produce hypervolaemia by increasing the transfusion rate and tachycardia by isoproterenol.

A superior vena caval syndrome appeared eight days after the operation but disappeared after a few days under treatment with diuretics, while a bilateral serosanguineous pleural effusion required aspiration. On getting up, the patient had no oedema of the inferior limbs and only a moderate hepatomegaly. There was a considerable biological improvement: the blood count was 4,200,000 RBC/mm³ and haematocrit was 50% (Table II).

TABLE II

		RBC per mm³	Haematocrit %	Pressure (mmHg) SVC	IVC	PA
Preoperative	30 yr	6,750,000	70	5		35
	36 yr	7,200,000	80			
Postoperative		4,200,000	50	10	15	30

It is too soon yet to carry out cardiac catheterization, but these haemodynamic and angiocardiographic investigations will be performed shortly. The clinical course is quite satisfactory 10 months postoperatively: there is no cyanosis and no systolic murmur; cardiac auscultation is normal except for an accentuated second heart sound. There is no venous stasis in the upper half of the body, no pleural effusion, and no oedema of the inferior limbs. There is only a moderate persistent hepatomegaly.

CASE 3 The third patient, N.B., was a 23-year-old woman. Dyspnoea and cyanosis had appeared in childhood. An episode of cardiac failure occurred during pregnancy, in 1969, followed by a premature birth four months before her admission to hospital in December 1969.

Physical examination showed clubbing of the fingers, cyanosis of the extremities, and hepatomegaly. A loud systolic murmur was heard in the whole precordial area. A blood count showed 3,700,000 RBC/mm³ and the haematocrit was 44%.

A diagnosis of tricuspid atresia with dextrocardia, atrial septal defect, and ventricular septal defect was made. There was a low pulmonary arterial pressure of 23·0 mmHg (Table III).

TABLE III

	Pressure (mmHg)			
	RA	PA		
Preoperative				
Maximal		23		
Minimal		13		
Mean	7	16		
	SVC–RPA	IVC	RA	LPA
Postoperative				
Maximal	15		22·5	17·5
Minimal	10		17·5	12·5
Mean		17·5		

Operation was performed in March 1970. First, a superior vena cava to pulmonary artery anastomosis was carried out; then, an anastomosis of the right atrium to the proximal end of the right pulmonary artery, using an aortic valve homograft; finally, under cardiopulmonary bypass, a pulmonary valve homograft was inserted into the inferior vena cava, the atrial septal defect was closed, and the main pulmonary artery was transected and sutured.

The initial postoperative course was satisfactory, with good cardiac action. But, despite blood overcompensation and isoproterenol, her pulse and blood pressure fell slowly 6 hours postoperatively and she died.

At necropsy there was no thrombosis and the anastomoses were patent; but the mitral valve was abnormal, with vegetations and a perforation of 1 cm² in the anterior leaflet. Finally, the right atrium was small and its wall was very thin.

We are of the opinion that failure was due to this mitral insufficiency.

DISCUSSION

A new surgical technique for repair of tricuspid atresia seems worth while. Indeed, in reviewing the literature on the subject, no corrective tech-