

The Fontan procedure in adults

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Abstract

Setting—Tertiary adult congenital cardiac referral centre.

Design—Retrospective cross sectional analysis.

Objectives—To report our 20 year experience with adult Fontan operations, and to compare late outcome in patients with single ventricle with definitive aortopulmonary or cavopulmonary shunt palliation.

Patients and main outcome measures—Patients older than 18 years undergoing Fontan operation between 1 January 1982 and 31 December 1998 were identified. Mortality and late outcome were derived from hospital records. These patients were compared with a cohort of 50 adults with single ventricle who had not undergone a Fontan operation.

Results—61 adults, median age 36 years (range 18–47 years), with a median follow up of 10 years (range 0–21 years) were identified. Actuarial survival was 80% at one year, 76% at five years, 72% at 10 years, and 67% at 15 years. Compared with before the Fontan operation, more patients were in New York Heart Association (NYHA) functional class I or II at the latest follow up (80% *v* 58%, $p < 0.001$). Systolic ventricular function deteriorated during follow up such that 34% had moderate to severe ventricular dysfunction at the latest follow up compared with 5% before Fontan ($p < 0.001$). Arrhythmia increased with time (10% before Fontan *v* 57% after 10 years, $p < 0.001$). Fontan patients had improved NYHA functional class, ventricular function, atrioventricular regurgitation, and fewer arrhythmias than the non-Fontan group at the latest follow up.

Conclusion—The Fontan operation in adults has acceptable early and late mortality. Functional class, systolic ventricular function, atrioventricular regurgitation, and arrhythmia deteriorate late after surgery but to a lesser degree than in non-Fontan patients with a single ventricle.

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Keywords: Fontan operation; univentricular heart; cavopulmonary shunt; aortopulmonary shunt

The Fontan operation allows total bypass of the right heart.¹ It was first described in 1971 in three patients with tricuspid atresia.¹ The ideal age to undergo the Fontan operation is thought to be between 18 months and 6 years.^{2–4} Not infrequently, however, adult patients with or without surgical palliation are encountered who are suitable for the operation.⁵ Under these circumstances volume unloading the single ventricle and simultaneously improving its oxygen delivery may permit better ventricular preservation and may result in less atrioventricular (AV) valve regurgitation, better functional capacity, and potentially improved longevity. Late outcome is, however, not well understood and comparative data with more conservative management are not well documented.⁶ We report our experience with the Fontan operation in adults over a 20 year period. We compared the study patients with a cohort of patients who had had either cavopulmonary or aortopulmonary shunts as their definitive palliation.

Patients and methods

PATIENTS

Fontan group

Consecutive patients older than 18 years at the time of surgery who underwent a modified Fontan procedure at the University of Toronto Congenital Cardiac Centre for Adults (UTC-CCA), Toronto, Canada, between 1 January 1982 and 31 December 1998 were identified from the UTCCCA database. Hospital

records, and reports of cardiac catheterisation and echocardiography were reviewed for patient characteristics. The cardiac morphology was reviewed and categorised according to operative notes, and echocardiographic and cineangiographic descriptions of the anatomy. Patients were assigned one or more of the following diagnostic categories based on their underlying anatomy: tricuspid atresia, double inlet left ventricle, pulmonary atresia with intact ventricular septum, double outlet right ventricle, atrial isomerism, transposition of the great arteries complex, or other hypoplastic ventricle.

Aortic saturations, mean pulmonary artery pressures, systolic ventricular function, the presence of pulmonary arterial stenoses (any focal or diffuse stenosis evident on either cardiac catheterisation or echocardiogram), dominant ventricular anatomy (left or other), and the severity of AV valve regurgitation were recorded from cardiac catheterisation and two dimensional (cross sectional) echocardiography reports before Fontan operation. Surgical history was obtained from operative notes. Recent follow up status was documented from hospital records and contact with the patient and referring physicians. Thirty seven study patients also formed part of a concurrent study looking at outcomes of late atrial arrhythmias after the Fontan operation.⁷ Ethics approval was obtained from the Toronto General Hospital research ethics board.

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Non-Fontan group

All patients with single ventricle physiology and either cavopulmonary or aortopulmonary shunt as their definitive palliation were identified from the UTCCCA database. Fontan modification was not performed because of any combination of the following factors: patient or family refusal, complex venous anatomy, hypoplastic or distorted pulmonary arteries, impaired ventricular function, AV valve regurgitation, or raised pulmonary vascular resistance. Data from patient records were analysed in a similar fashion to the Fontan group. Details of these patients have been described in a report on outcome after aortopulmonary and cavopulmonary shunt palliation in patients with single ventricle.⁸

STUDY END POINTS

The primary end point was death or transplantation. Secondary end points were functional class, sustained arrhythmia (at least one episode of sustained atrial or ventricular arrhythmia as documented on 12 lead ECG or Holter studies), systolic ventricular function, systemic AV valve regurgitation, diuretic requirement, freedom from reoperation, thrombotic phenomena of clinical significance (warranting either surgical intervention or institution of anticoagulation), protein losing enteropathy, and arterial O₂ saturations at rest. End points were determined at latest follow up—that is, within one year of the study start date (1 January 2000). Thirty seven (86%) Fontan survivors had less than one year old follow up data, five (12%) had one to two year old follow up data, and two (5%) had no recent follow data. Of the surviving non-Fontan patients only one did not have recent follow up information. Systolic ventricular function was visually graded as I (normal function, ejection fraction > 60%), II (mild dysfunction, ejection fraction 40–59%), III (moderate dysfunction, ejection fraction 20–40%), and IV (severe dysfunction, ejection fraction < 20%). AV regurgitation was graded as I (absent or trivial), II (mild), III (moderate), or IV (severe).

DATA ANALYSIS

Descriptive statistics including mean, SD, median, and range were used to describe patient characteristics and to denote secondary outcome variables. Paired *t* test was used to compare continuous variables and a χ^2 test or

Fisher's exact test was used to compare dichotomous values. Each set of data was compared with pre-Fontan values. Functional class, systolic ventricular function, AV regurgitation, and presence of arrhythmia were compared between the Fontan and non-Fontan groups. Differences between Fontan survivors and non-survivors were documented using a univariate Cox regression model. New York Heart Association (NYHA) functional class, systolic ventricular function, and systemic AV valvar regurgitation were subclassified as I and II, and III and IV, respectively, for statistical analysis and data representation. A probability value of *p* < 0.05 was considered significant.

We used the Kaplan-Meier method to describe actuarial survival and used survival beyond 18th birthday to document differences between the groups because age at definitive palliation varied widely. We assessed risk factors for mortality with a multivariate Cox regression model, using initial Fontan operation as time zero and death or transplantation as the event. Highly correlated candidate variables were identified and the most significant factor was used in the model. Demographic parameters used in the analysis were sex and age at initial Fontan operation. Continuous variables were year of operation, length of follow up (time from Fontan, independent of age, to death or transplantation, or latest follow up), arterial saturations, mean pulmonary arterial pressures, and bypass and cross clamp times.

We entered dominant ventricle, Blalock-Taussig, Potts or Waterston shunt, Glenn anastomosis, pulmonary arterial banding, transposition of the great arteries complexes, tricuspid atresia, pulmonary atresia with intact ventricular septum, double inlet left ventricle, double outlet right ventricle, cyanosis, presence of arrhythmia (atrial and or ventricular), pulmonary arterial stenoses, subaortic stenosis, AV regurgitation, functional class (NYHA classification), and the need for early reoperation because of bleeding as dichotomous variables.

Results

PATIENT CHARACTERISTICS

Fontan group

Between 1 January 1978 and 31 December 1998, 61 adults underwent a modified form of the Fontan operation at the Toronto General Hospital (table 1). Thirty two (52%) were male

Table 1 Characteristics of the study and control groups

	Fontan (n = 61)	CP shunt (n = 35)	<i>p</i> Value	AP shunt (n = 15)	<i>p</i> Value
Length of follow up (years) Range	9.1 (6.4) (0–21)	17.9 (6.4) (8–31)	0.349	18 (6.9) (5–33)	0.722
Length follow up after 18th birthday (years) Range	17.1 (8.8) (3–46)	12.2 (5) (1.6–22.7)	0.173	8.9 (4.9) (2.5–20.5)	–
Age at definitive palliation (years) Range	26 (7) (18–47)	12.6 (7.7) (1–35)	0.431	12.7 (14.5) (0.1–53)	0.320
Male:female (%)	53:47	51:49	0.903	73:27	0.105
TGA complex	32 (53%)	17 (49%)	0.790	6 (40%)	0.402
Tricuspid atresia	30 (49%)	14 (40%)	0.200	8 (53%)	0.846
Pulmonary atresia IVS	4 (7%)	13 (37%)	<0.001	5 (33%)	<0.001
Atrial isomerism	3 (5%)	5 (15%)	0.010	2 (13%)	0.132
Left ventricle dominant	48 (79%)	32 (91%)	0.066	13 (87%)	0.451
Gr I&II vfx: (baseline)	55 of 58 (95%)	20 of 35 (57%)	<0.001	13 of 15 (87%)	0.182
Gr I&II AV regurgitation (baseline)	57 of 59 (97%)	31 of 35 (89%)	0.016	14 of 15 (93%)	0.405
Age at latest follow up (years) Range	35.1 (9) (20–66)	31.4 (5.9) (19.6–46.5)	0.753	30.5 (10.3) (20.5–57.8)	0.848

p < 0.025 was considered significant; this value is corrected for multiple comparisons.

AP, aortopulmonary; CP, cavopulmonary; Gr I&II AV regurgitation, mild to moderate systemic atrioventricular valve regurgitation before definitive palliation; Gr I&II vfx, proportion of patients with mild to moderate ventricular dysfunction before definitive palliation; IVS, intact ventricular septum; TGA, transposition of the great arteries.

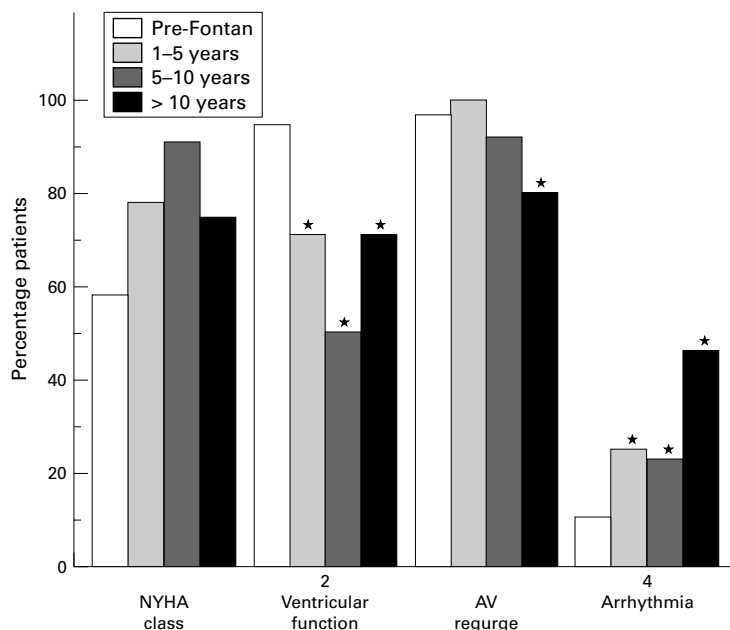


Figure 1 Proportion of patients in grades I and II for each parameter over time. *p* Values reflect comparisons with baseline data. **p* < 0.020 corrected for multiple comparisons. Twelve patients were at 1–5 years, 13 at 5–10 years, and 25 at more than 10 years of follow up.

patients. Median age at Fontan was 24 years (range 18–47 years). Forty two patients (69%) had atriopulmonary anastomoses, nine (15%) had AV connection via a conduit to a rudimentary right ventricle, and nine (15%) had lateral tunnel modification.

Twenty nine patients (48%) had preceding cavopulmonary shunts with either end-to-end (classic Glenn) or end-to-side (bidirectional Glenn shunt) anastomoses. Twenty eight patients (46%) had a Blalock-Taussig shunt, six (10%) a Potts shunt, and three (5%) patients had a Waterston shunt. Three (5%) patients had undergone prior pulmonary arterial banding during infancy. Twelve patients (20%) had no prior palliation. Baseline NYHA functional class, systolic ventricular function, and AV valvar regurgitation along with changes over time are summarised in fig 1. Four patients had documented atrial fibrillation and two patients had atrial flutter before the Fontan operation. Mean arterial saturation was 85 (7)% (range 56–95%).

Non-Fontan group

There were 50 patients who did not have a Fontan operation. All patients in this group had one or more palliative shunt procedures: 15 had an arterial shunt (Blalock-Taussig, Potts, Waterston, axillary arteriovenous fistula, or aortopulmonary interposition graft) and 35 patients had cavopulmonary shunts. Of the 35 patients with cavopulmonary shunts, nine had a classic connection and the remainder had a bidirectional connection as their definitive palliation. Similarities and differences in relation to the Fontan group are summarised in table 1.

SURVIVAL

Fontan group

Patients were followed up for a median of 10 years (range 0–21 years) after Fontan operation

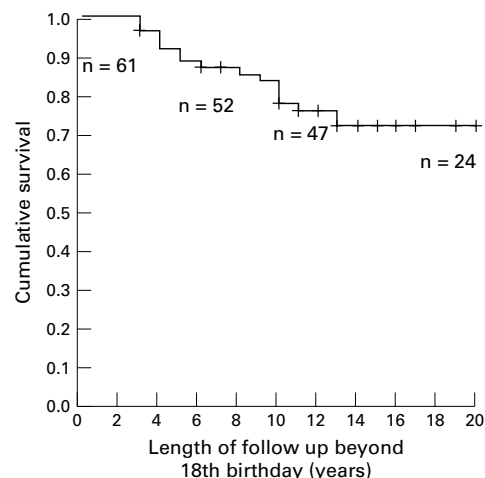


Figure 2 Fontan shunt actuarial survival; 18th birthday is taken as point zero.

(fig 2). Median age at latest follow up was 36 years (range 20–66 years). Actuarial survival determined using Fontan operation as point zero was 80% at one year, 76% at five years, 72% at 10 years, and 67% at 15 years. There were 18 (30%) deaths in the Fontan group. Necropsy data were available in 15. Eight deaths (13%) occurred in the early postoperative period, of which three patients had intractable arrhythmia, one patient had high pulmonary vascular resistance, and the remaining four patients had low output syndromes. One patient had take down of the Fontan connection and reinstitution of an aortopulmonary shunt but died by the second postoperative day. Of the remaining seven late deaths the cause was heart failure in one, sudden death in three (at one, eight, and 11 years, presumably related to arrhythmia), pulmonary embolism in one (eight years after the procedure) and complications of cerebrovascular accident in one. The cause of death was not known in the other patient.

Higher preoperative mean pulmonary arterial pressures (19 (9) *v* 14 (5) mm Hg, *p* = 0.025) and more reoperation for postoperative bleeding (35% *v* 2.3%, *p* = 0.005) occurred in the group that died than in those who survived (univariate Cox regression analysis). Ventricular function, degree of AV valve regurgitation, arterial saturations, bypass times, and functional class were not significantly different between those who did not survive and those who did.

Non-Fontan group

Cumulative survival is summarised in figs 3 and 4. Determinants of survival for this group have been previously described.⁸

SECONDARY OUTCOMES

Fontan group

The latest review was 1–5 years after Fontan operation in 12 patients, 5–10 years in 13 patients, and more than 10 years in 25 patients.

At latest follow up, more patients were in functional class I and II than before the Fontan operation (36 of 45 (80%) *v* 35 of 61 (57%), *p* < 0.001). Twenty four patients had more

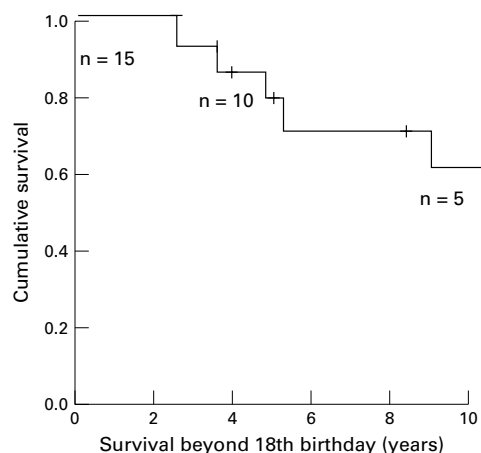


Figure 3 Aortopulmonary shunt actuarial survival; 18th birthday is taken as point zero.

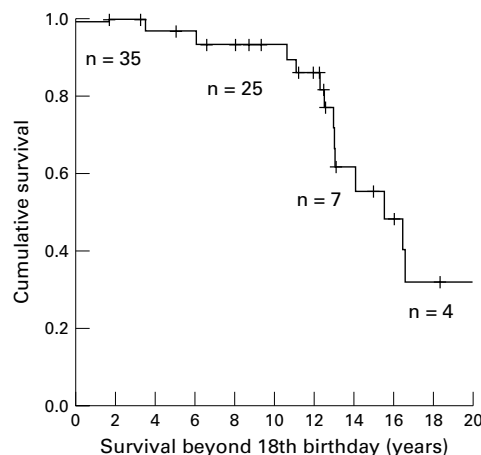


Figure 4 Cavopulmonary shunt actuarial survival; 18th birthday is taken as point zero.

than 10 years of follow up, at which time 18 (75%) were in class I and II, for a relative decline in functional class from 1–5 years following surgery when 10 of 11 (91%) were in class I and II ($p = 0.005$). There were no risk factors for poor NYHA functional class (grades III and IV) at the latest follow up.

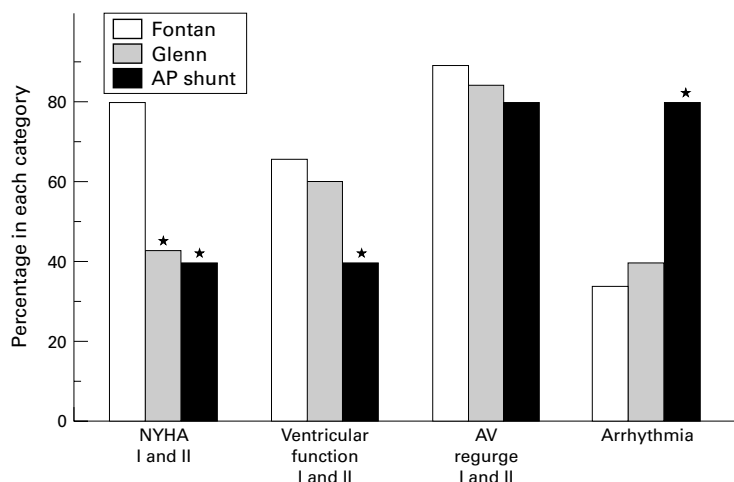


Figure 5 Functional characteristics and latest follow up. *P* values reflect comparisons between the Fontan group and the control groups. * $p < 0.025$, corrected for multiple comparisons.

Systolic ventricular function deteriorated during follow up, being evident at 1–5 years following surgery. After 10 years 15 of 21 (71%) patients had grade I and II function, compared with 55 of 58 (95%) ($p < 0.001$) before Fontan operation. Fontan revision was the only significant determinant of poor systolic ventricular function (grade III and IV) determined by multivariate analysis ($p = 0.017$).

Systemic AV regurgitation worsened with time such that after 10 years 20 of 25 patients (80%) had grade I and II regurgitation compared with 57 of 59 (97%) ($p = 0.010$) before Fontan surgery. AV regurgitation correlated with worsening ventricular function ($r = 0.962$, two tailed Pearson correlation coefficient).

Arrhythmia increased in frequency with longer duration of follow up such that at 1–5 years three of 12 (25%) were affected ($p = 0.005$), at 5–10 years four of 12 (33%) ($p = 0.013$) were affected, and after 10 years 11 of 24 (46%) ($p < 0.001$) were affected.

Arterial saturation at latest follow up was 92 (5)% and was greatly improved from the preoperative saturation of 85 (6)%, as is expected ($p < 0.001$). Saturations in the non-Fontan group were significantly lower at 83 (8)% ($p < 0.001$) for both cavopulmonary and aortopulmonary shunted patients.

At 1–5 years, four of 12 patients (33%) required diuretics, at 5–10 years, six of 12 (50%) ($p = 0.171$) required diuretics, and after 10 years 13 of 25 (52%) ($p = 0.038$) required diuretics.

Fourteen of the 43 survivors (32%) had experienced a thrombotic event of clinical significance, warranting surgical treatment in three (7%) and institution of anticoagulation in 14 (32%). Two patients (5%) had protein losing enteropathy at latest follow up.

Twelve of the 43 surviving Fontan patients (28%) required reoperation at a median of 11 years (range 2–18). Reasons for reoperation were conduit stenosis in five, conduit regurgitation in one, atrial arrhythmia with secondary heart failure requiring lateral tunnel modification in one, and subaortic stenosis with secondary heart failure in one. Three patients were reoperated on because of thrombotic complications and one patient had early surgical re-exploration for bleeding.

Comparison with non-Fontan group

At latest follow up, functional class, ventricular function, and incidence of arrhythmia were significantly better in the Fontan group (fig 5).

Discussion

Managing the adult with single ventricle physiology and prior palliation can be perplexing. Performing the Fontan operation under these circumstances has incompletely understood risks and benefits. Our study provides insight into outcome late after adult Fontan surgery. To our knowledge it is the longest follow up in this population and additionally draws lessons, albeit limited, from a non-Fontan single ventricle patient cohort. Our data has shown that the

Fontan patients have improved functional class, comparatively good preservation of ventricular function, and excellent maintenance of AV valve integrity with relative reduction in arrhythmia in the intermediate term. However, over time all of these variables deteriorate, although less rapidly than in adults with a single ventricle who have not undergone Fontan operation.

SURVIVAL

It is reassuring that our survival statistics for the adult who has undergone the Fontan operation correspond well with the mixed age cohorts of Fontan *et al*⁹ and Driscoll *et al*¹⁰ with 69–70% 10 year survival and 60–63% 15 year survival, suggesting that higher mortality is not implicit in the adult Fontan operation. Humes *et al*⁶ reached similar conclusions. Interestingly they had an early mortality of only 6% (5/77 patients) and had only eight late deaths.⁶ Their follow up period was shorter, however (4 v 10 years), and this may explain in part the discrepancy in late mortality. Our 13% perioperative mortality occurred secondary to arrhythmia or poor cardiac output states. This corresponds more with the 1973 to 1984 Fontan cohort of Driscoll *et al*.¹⁰ As in other studies, deaths occurred predominantly during our first decade of experience with the procedure.¹⁰ In the current era perioperative mortality at our institution is 4%.¹¹

Late mortality in the non-Fontan group was high (40%). Modes of death and risk factors in this group have been previously ascribed to worsening ventricular function, progressive AV regurgitation, and the development of arrhythmia.⁸ Kopf *et al*¹² described a 22% late (30 years) mortality in patients with single ventricle physiology palliated by cavopulmonary shunt after definitive surgery.

SECONDARY OUTCOMES

Functional class

NYHA functional class was significantly improved at latest follow up compared with before the Fontan operation, substantiating symptomatic improvement, and appeared most favourable in the intermediate term (5–10 years). After 10 years, functional class had declined from the values at 1–5 years, but was not different from before the Fontan operation (fig 1). In the adult Fontan cohort of Gates *et al*,⁵ functional class had improved significantly at 7.4 (3.8) years from preoperative values. They anticipated progressive time related disability as 10 of their 13 patients at more than five years of follow up were in class II and three were in class I. This is in contrast with three of six who were in class II and three who were in class I with less than five years of follow up. Fontan *et al*⁹ described similar trends after the “perfect Fontan” and attributed it to the Fontan state per se.

Functional status

Functional status was substantially better preserved than in the non-Fontan group with 80% being in class I or II compared with 40% and 43% in patients with aortopulmonary and

Glenn shunts, despite similar age at latest follow up. Patients with aortopulmonary shunts had more ventricular dysfunction than patients with Fontan operation, and patients with cavopulmonary and aortopulmonary shunts were more desaturated at rest, elucidating at least in part the discrepancy in functional capacity.¹³

Systolic ventricular function

Systolic ventricular function declined with time after the Fontan operation such that at latest follow up a third of patients had moderate or severe reduction in function. This decline was also reflected by an increasing need for diuretics during follow up. The only significant variable that predicted poor ventricular function was reoperation. Patients with cavopulmonary shunts without a Fontan operation had a degree of ventricular dysfunction similar to that of the Fontan group at latest follow up, but in contrast, patients with aortopulmonary shunt had significantly worse ventricular dysfunction. Our findings suggest that partial or complete volume offloading offers protection to the single ventricle but does not prevent late deterioration. Cheung *et al*¹⁴ showed progressive diastolic dysfunction in a cohort of paediatric Fontan patients and it is conceivable that such changes may induce neurohumoral up-regulation that eventually results in late systolic dysfunction.¹⁵

Systemic AV regurgitation

Systemic AV regurgitation correlated with ventricular dysfunction. At latest follow up 10% of patients had moderate or severe regurgitation, presumably reflecting ventricular functional impairment and annular dilation. AV regurgitation tended to be increased in the aortopulmonary shunted group compared with either the Fontan or cavopulmonary shunt patients.

Arrhythmia

There was a progressive increase in the number of patients with arrhythmia: after 10 years, 46% had documented sustained arrhythmia on at least one occasion. Compared with patients with an aortopulmonary shunt the incidence was considerably lower (80% v 40%). Atrial arrhythmia and its underlying mechanisms in patients with Fontan operation are well described.^{16–18} The likely mechanism in patients with aortopulmonary shunt (many of whom had no atrial surgery) is atrial stretch and dilation of the atrial myocardium secondary to chronic volume overload of the single ventricle.⁸

Other complications

One third of our patients with Fontan operation had thrombotic events of clinical significance, three of them requiring surgical thrombectomy. Routine long term anticoagulation was not practised in this cohort, nor were thromboses routinely screened for. There may have been other people with subclinical thromboses, making the cumulative incidence substantial. Data supporting routine use of anticoagulation for this group are lacking¹⁹; however,

this study strongly supports the need for large prospective multicentre trials to document the risk to benefit ratio of routine anticoagulation in this group.

Freedom from reintervention

Late reoperation rate was high in the Fontan cohort (28%), significantly more than the 16% described by Gates *et al.*⁵ Our rates correspond to the 29% reported reoperation rate of Driscoll *et al.*¹⁰ and in part reflect our relatively frequent use of right atrial to right ventricular conduits, which became obstructed or regurgitant. Two patients had successful lateral tunnel conversion after developing atrial arrhythmia and conduit stenosis. Because valved conduits are no longer used and the cavopulmonary anastomosis site is generally positioned away from the anterior chest wall,^{20 21} newer Fontan modifications are less likely to require reoperation for this reason. One quarter of reoperations were secondary to thrombotic complications. This implies that a lower threshold for anticoagulation may be appropriate in adult patients with Fontan operation and that active surveillance for thrombotic phenomena may reduce the need for surgical intervention.

STUDY LIMITATIONS

Data presented in this study were not collected prospectively or serially, and patients were not randomly assigned to Fontan and non-Fontan palliation. The attrition rate over the paediatric years was not included in this study. There was a selection bias in both groups as they were required to reach age 18 years for inclusion in the study. Some patients in the non-Fontan group were thought unsuitable for the Fontan procedure and some have had their definitive palliations during childhood as well as adulthood. However, we believe that the similarities of morphological spectra, age, functional class, and arterial oxygen saturation before definitive surgery allow for limited inferences to be made.

CONCLUSION

The Fontan operation may be offered to adults with acceptable early and late mortality. Outcome continues to improve with experience, better selection criteria, and newer modifications of the procedure. Despite late deterioration, the Fontan state seems to offer improved late mortality, better preservation of functional status and ventricular function, and more freedom from arrhythmia than aortopulmonary or cavopulmonary shunt palliation

alone. All current management options for the adult with functional single ventricle are suboptimal, but the Fontan operation has virtues that make it a reasonable strategy in carefully selected adults.

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