Long-term outcomes following Fontan takedown in Australia and New Zealand

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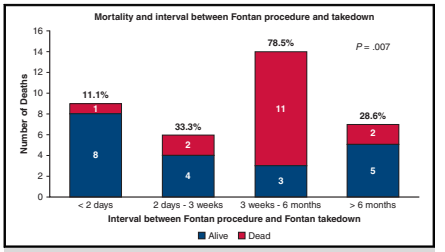
ABSTRACT

Objective: Fontan takedown remains an option for the management of Fontan failure. We sought to evaluate early and late outcomes after Fontan takedown.

Methods: The Australia and New Zealand Fontan Registry was interrogated to identify all patients who had a Fontan takedown.

Results: Over a 43-year study period (1975-2018), 36 of 1540 (2.3%) had a Fontan takedown. The median age at takedown was 5.1 years (interquartile range [IQR], 3.7, 7.0). Nine (25%) patients had a takedown within 48 hours, 6 (16%) between 2 days and 3 weeks, 14 (39%) between 3 weeks and 6 months, whereas 7 (19%) had a late takedown (>6 months). Median interval to takedown was 26 days (IQR, 1.5, 127.5). Sixteen (44%) patients died at a median of 57.5 days (IQR, 21.8, 76.8). The greatest mortality occurred between 3 weeks and 6 months (<2 days: 1/9, 11%; 2 days to 3 weeks: 2/6, 33%; 3 weeks to 6 months: 11/14, 79%; >6 months: 2/7, 28%; $P = .007$). At median follow-up of 9.4 years (IQR, 4.5, 15.3), 11 (31%) patients were alive with an intermediate circulation (10 in New York Heart Association class I/II). Five (14%) patients underwent a successful second Fontan. Freedom from death/transplant after Fontan takedown was 59%, 56%, and 52% at 1, 5, and 10 years, respectively.

Conclusions: The incidence of Fontan takedown is low, but mortality is high. The majority of takedowns occurred within 6 months. Mortality was lowest when takedown occurred <2 days and highest between 3 weeks and 6 months. A second Fontan is possible in a small proportion of survivors. (J Thorac Cardiovasc Surg 2021;161:1126-35)



Early Fontan takedown had the lowest mortality.

CENTRAL MESSAGE

Early Fontan takedown has the lowest mortality. A second Fontan is possible in a small proportion of survivors.

PERSPECTIVE

The incidence of Fontan takedown is low. Factors influencing takedown remain unclear. The majority of takedowns occurred within 6 months (80%). Mortality was lowest when takedown occurred <2 days (11%) and greatest between 3 weeks and 6 months (79%). Midterm outcomes after takedown to an intermediate circulation are acceptable. A second Fontan is possible in a small proportion of survivors.

See Commentaries on pages 1136 and 1137.

Survival with a Fontan circulation has significantly improved in the current era.[1-3] However, morbidity continues to be an unyielding challenge.[4] Early Fontan failure presents in a variety of ways, including low cardiac output state, high Fontan circuit pressures, large fluid requirements, and prolonged high-volume pleural effusions.[5] Late Fontan failure often presents as intractable arrhythmias, protein-losing enteropathy (PLE), plastic bronchitis,

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Abbreviations and Acronyms
BCPS = bidirectional cavopulmonary shunt
CI = confidence interval
DORV = double outlet right ventricle
IQR = interquartile range
NYHA = New York Heart Association
PLE = protein-losing enteropathy

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or poor functional capacity. Irrespective of the etiology or presentation, Fontan failure often leads to death and needs to be managed aggressively.[6,7]

One of the options of managing Fontan failure is takedown of the Fontan circuit to an intermediate palliation stage in the single-ventricle pathway (bidirectional cavopulmonary shunt and/or systemic-pulmonary artery shunt).[8] There is limited information about patients undergoing Fontan takedown.[8] The aim of our study was to identify and study all Fontan takedowns from the entire population of Australia and New Zealand over a period of 43 years.

METHODS

Sample

The institutional review board approval number for this study is HREC/18/QRCH/105, dated March 28, 2018. Fontan takedowns were initially identified from the Australia and New Zealand Fontan Registry, which includes all patients undergoing the Fontan operation in either country.[9] However, Fontan patients are included in the Registry only after hospital discharge. Patients who underwent Fontan takedown in the same hospital admission were identified by interrogating individual hospital databases in each contributing pediatric cardiac center in Australia and New Zealand.

Takedowns were stratified into immediate, early, intermediate, and late groups depending on the interval since the Fontan operation (immediate <48 hours; early: 2 days to 3 weeks; intermediate: 3 weeks to 6 months; late: >6 months).

Statistical Analysis

Descriptive statistics are presented using percentages, median, and interquartile range (IQR). Statistical comparisons were performed with nonparametric tests including the Fisher exact binomial P values for categorical independent variables, and Wilcoxon rank-sum P values for continuous independent variables. A P value $< .05$ was considered statistically significant.

RESULTS

Over a 43-year study period (1975-2018), 1540 patients underwent a Fontan procedure. Thirty-six (2.3%) patients had subsequent takedown of the Fontan.

Baseline Characteristics

Of the 36 patients who underwent Fontan takedowns, 25 (69.4%) were male (Table 1). Six patients (16.7%) had heterotaxy whereas 4 patients (11.1%) had dextrocardia. The dominant ventricular morphology was left ventricle in 17 patients (47.2%), right ventricle in 17 (47.2%), biventricular in 1 (2.8%), and indeterminate in 1 (2.8%). Underlying diagnoses were as follows: hypoplastic left heart syndrome (n = 10, 27.8%), double inlet left ventricle (n = 7, 19.4%), double outlet right ventricle (DORV) (n = 5, 13.9%), tricuspid atresia (n = 4, 11.1%), complete atrioventricular septal defect (n = 4, 11.1%), pulmonary atresia with intact ventricular septum (n = 3, 8.3%), complete atrioventricular septal defect/DORV (n = 2, 5.6%), congenitally corrected transposition of the great arteries (n = 1, 2.8%).

Nineteen (52.7%) patients had bidirectional cavopulmonary shunt (BCPS) whereas 2 (5.5%) had Hemi-Fontan as stage II procedure. Fifteen (41.6%) patients were single-stage Fontan procedures. The median age at stage II procedure was 5.4 months (IQR, 3, 16.8) whereas the median interval from stage II procedure to Fontan was 3.5 years (IQR, 2.7, 5.1). Five (14%) patients had an atriopulmonary Fontan, 14 (39%) had a lateral tunnel Fontan, and 17 (47%) had an extracardiac conduit Fontan ($P = .039$). Pre-Fontan hemodynamic data were available in 31 (86%) patients of which 27 (87%) had mean pulmonary artery pressures $\leq$15 mm Hg. No patient had moderate or severe ventricular dysfunction or moderate or greater atrio-ventricular valve regurgitation preoperatively. An examination of era distribution of Fontan takedowns as compared to total Fontan procedures during the same period revealed no era effect ($P = .069$) (Table 1).

Takedown Characteristics

The median age at takedown was 5.1 years (IQR, 3.7, 7.0). Nine (25%) patients had a Fontan takedown within 48 hours, 6 (16%) had an early takedown (2 days to 3 weeks), 14 (39%) had intermediate takedown between 3 weeks and 6 months, whereas 7 (19%) had a late takedown (>6 months). The median interval to Fontan takedown was 26 days (IQR, 1.5, 127.5). Indications for takedown were low cardiac output syndrome (19, 53%), intractable pleural effusions (12, 33%), PLE (3, 8%), thrombosis (1, 2.7%), and unknown (1, 2.7%).

Timing of Takedown

Immediate (<48 hours; n = 9; 25%). All 9 patients developed intractable low cardiac output immediately after the Fontan. One patient could not be weaned off cardiopulmonary bypass and the Fontan was taken down intraoperatively.
Early (2 days to 3 weeks; n = 6, 16.7%). All 6 patients presented with intractable low cardiac output. One patient also developed thrombosis of the Fontan circuit.
Intermediate (3 weeks to 6 months; n = 14; 38.9%). Eleven of the 14 patients (79%) presented with intractable pleural effusions whereas 3 patients (21%)

CONG

TABLE 1. Baseline characteristics of patients undergoing Fontan takedown

Characteristic	n (%)
Male	25 (69.4%)
Heterotaxy	6 (16.7%)
Dextrocardia	4 (11.1%)
Dominant ventricular morphology	
Left ventricle	17 (47.2%)
Right ventricle	17 (47.2%)
Biventricular	1 (2.8%)
Indeterminate	1 (2.8%)
Baseline diagnosis	
HLHS	10 (27.8%)
DILV	7 (19.4%)
DORV	5 (13.9%)
Tricuspid atresia	4 (11.1%)
CAVSD	4, (11.1%)
PA-IVS	3 (8.3%)
CAVSD + DORV	2 (5.6%)
ccTGA	1 (2.8%)
Type of Fontan	
AP	5 (14%)
LT	14 (39%)
ECC	17 (47%)
Era of Fontan takedowns and concomitant Fontans during the same period (%)*	
1975-1989	3/189 (1.5%)
1990-1999	15/361 (4.1%)
2000-2009	12/518 (2.3%)
2010-2017	6/472 (1.2%)

HLHS, Hypoplastic left heart syndrome; *DILV*, double inlet left ventricle; *DORV*, double outlet right ventricle; *CAVSD*, complete atrioventricular septal defect; *PA-IVS*, pulmonary atresia/intact ventricular septum; *ccTGA*, congenitally corrected transpoistion of the great arteries; *AP*, atriopulmonary; *LT*, lateral tunnel; *ECC*, extracardiac conduit. *$P = .069$.

presented with low cardiac output. All 14 patients underwent multiple procedures in an attempt to ameliorate the pleural effusions including multiple chest tube insertions and pleurodesis.

Late (>6 months, n = 7, 19.4%). These patients had a varied presentation (PLE, n = 3; low cardiac output, n = 1, pleural effusion, n = 1, thrombosis of the Fontan circuit, n = 1, unknown, n = 1). Overall, 80% of all Fontan takedowns occurred within 6 months of the index procedure.

Hemodynamics Post-Fontan Takedown

Post-Fontan takedown catheterization data were available in 11 (30%) patients. A catheter study was not performed in the remaining patients. Median pulmonary artery pressure was 13 mm Hg (IQR, 13, 18 mm Hg) whereas the median ventricular end diastolic pressure was 10 mm Hg (IQR, 8, 12 mm Hg). Median transpulmonary gradient was 4 mm Hg (IQR, 4, 5 mm Hg).

Fate After Fontan Takedown (Table 2)

Stratified by timing of takedown. *Immediate (n = 9).* There was 1 death in a 5-year-old patient with a lateral tunnel Fontan despite mechanical support. A 4-year-old patient with DORV and hypoplastic left ventricle underwent an unsuccessful second attempt at the Fontan and ultimately underwent a heart transplant. Two patients underwent a successful second Fontan completion. Five patients are still alive with an intermediate circulation.

Early (n = 6). Two patients died immediately following Fontan takedown (both procedures were performed in the 1980s). Two patients are alive with an intermediate circulation. Two patients underwent a successful Fontan at the second attempt 4.4 and 15.5 years after the initial Fontan takedown.

Intermediate (n = 14). Before the decision to takedown the Fontan, 13 (27%) patients had 14 rescue procedures including Fontan revision (n = 5), enlargement of fenestration (n = 4), coiling of aortopulmonary collaterals (n = 2), pacemaker insertion (n = 1), left pulmonary artery stenting (n = 1), and removal of bronchial cast (n = 1). Other nonrescue procedures included thoracotomy and pleurodesis (n = 4), drainage of pleural effusion (n = 2), and drainage of pericardial effusion (n = 1). Eleven patients died, 9 of whom presented with pleural effusions. Of the 3 survivors, 2 are alive with an intermediate circulation and 1 patient with pulmonary atresia/intact ventricular septum was converted to one and a half ventricle repair at Fontan takedown.

TABLE 2. Fate after Fontan takedown stratified by timing of Fontan takedown

Timing of takedown after Fontan	n (%)	Death	Transplant	Intermediate circulation	Second Fontan	Biventricular/one and a half ventricle repair
<2 d	9 (25%)	1	0	5	3	0
2 d to 3 wk	6 (16.7%)	2	0	2	2	0
3 wk to 6 mo	14 (38.9%)	11	0	2	0	1
>6 mo	7 (19.4%)	2	1	2	0	2
Total	36	16	1	11	5*	3

*One patient subsequently underwent a heart transplant.

Late (n = 7). Two patients died immediately after Fontan takedown. One patient had a successful transplant. Two patients are alive with an intermediate circulation, whereas 2 patients were successfully converted to a biventricular circulation. Of the 3 patients who presented with protein losing enteropathy, 1 patient died after Fontan takedown while in the remaining 2 patients the protein losing enteropathy improved after Fontan takedown.

Entire cohort. *Death (n = 16).* Sixteen (44%) patients died at a median of 57.5 days (IQR, 21.8, 76.8) after the Fontan procedure (Figure 1). Patients who had a takedown between 3 weeks and 6 months had the greatest mortality and patients who had the Fontan taken down within 48 hours had the lowest mortality (<2 days: 1/9, 11%; 2 days to 3 weeks: 2/6, 33%; 3 weeks to 6 months: 11/14, 79%; >6 months: 2/7, 28%; $P = .007$) (Figure 2). Transplant-free survival at 15 years was 47% (95% confidence interval [CI], 28-64). We found no association between any of the pre-takedown characteristics and death (Tables 3 and 4).

Transplant (n = 1). One patient with double inlet left ventricle underwent a heart transplant. The patient had a late Fontan takedown 14.3 years after an atriopulmonary Fontan. He presented with thrombosis of the Fontan circuit, which was taken down to a bidirectional cavopulmonary shunt, which was not tolerated. He successfully underwent a transplant 5 months after takedown. He was lost to follow-up at the time of the study.

Intermediate circulation (n = 11). All 11 (31%) patients remained alive with an intermediate circulation after a median follow-up 9.4 years (IQR, 4.5, 15.3). The sources of pulmonary blood flow in this group included BCPS (n = 8), BCPS and systemic to pulmonary artery shunt (n = 1), BCPS and arteriovenous fistula (n = 1), and Hemi-Fontan, systemic to pulmonary artery shunt and arteriopulmonary fistula (n = 1). Ten of these patients were in New York Heart Association (NYHA) class I/II, with 1 patient in NYHA III. No patient in this group has had a heart transplant. Eight patients were deemed unsuitable for a second Fontan on repeat cardiac catheterization (pulmonary artery pressure >16 mm Hg [n = 6], elevated ventricular

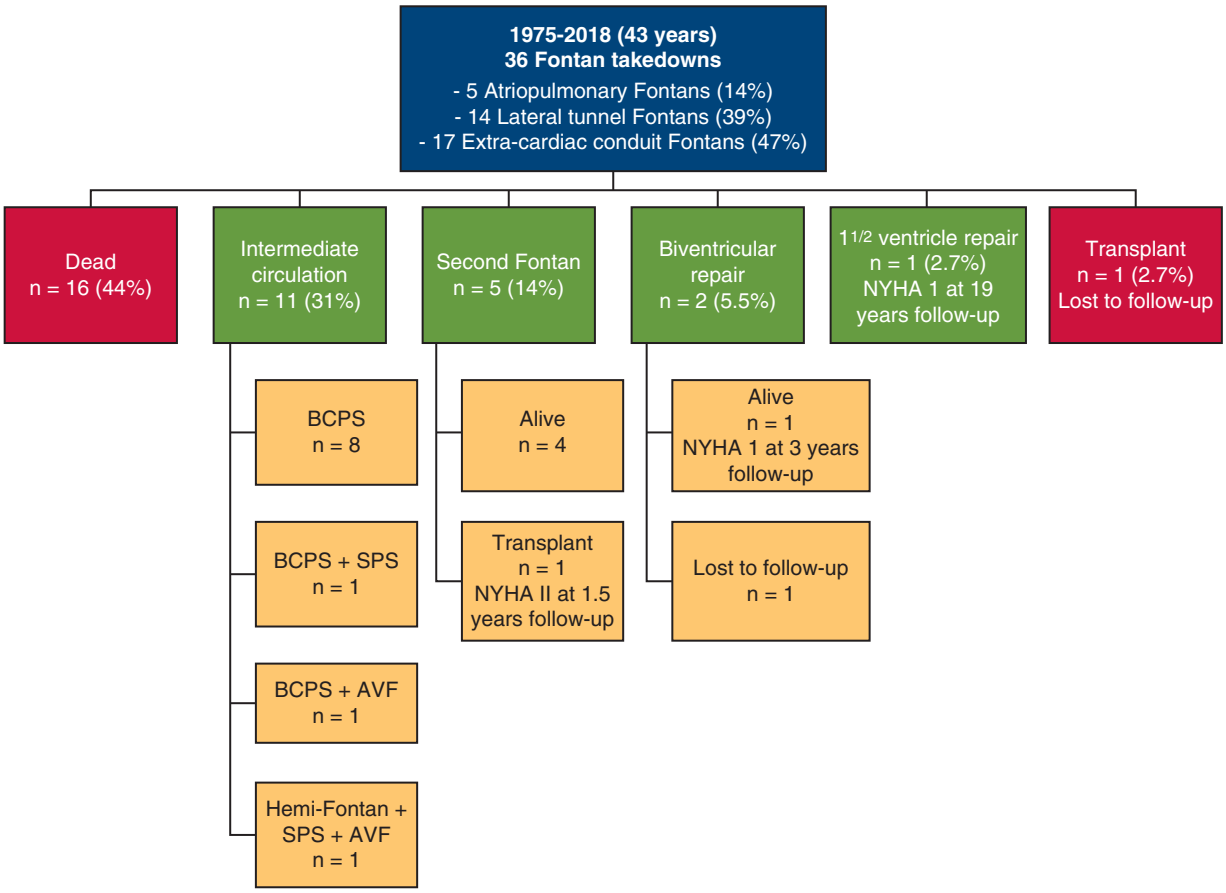


FIGURE 1. Fate of all 36 Fontan takedown patients operated between 1975 and 2018. Overall mortality was 44%; 31% were alive with an intermediate circulation whereas 14% patients had a second Fontan. *NYHA*, New York Heart Association; *BCPS*, bidirectional cavopulmonary shunt; *SPS*, systemic to pulmonary artery shunt; *AVF*, arteriovenous fistula.

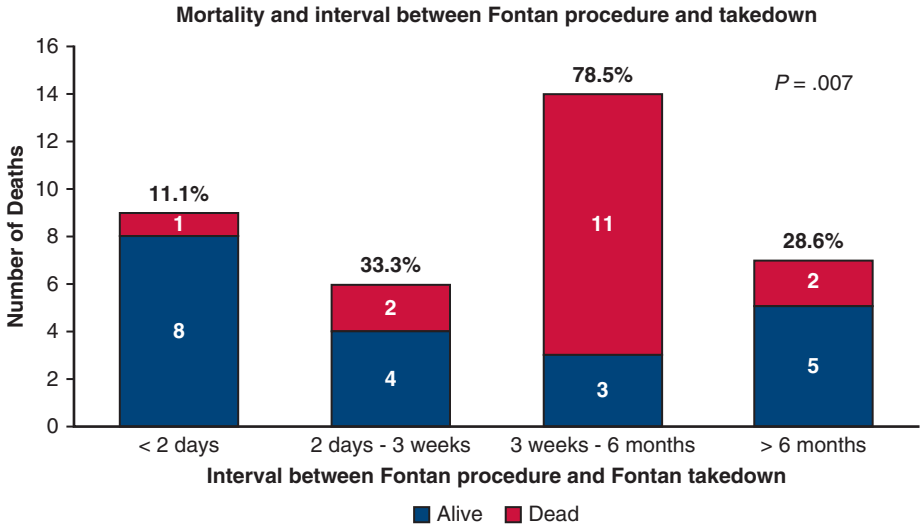


FIGURE 2. Mortality and interval to Fontan takedown (n = 36, 1975-2018): patients who had Fontan takedown within 48 hours had the lowest mortality (11.1%), whereas patients who had takedown between 3 weeks to 6 months after Fontan had the greatest mortality (78.5%) ($P = .007$).

TABLE 3. Association between categorical independent predictors and death following Fontan takedown

Predictor	Death				Fisher *P**
	No		Yes		
	%	n	%	n	
Sex					
F	30	6	31.25	5	1
M	70	14	68.75	11	
Heterotaxy					
N	80	16	87.5	14	.672
Y	20	4	12.5	2	
Dextrocardia					
N	80	16	100	16	.113
Y	20	4	0	0	
Dominant ventricle					
LV	61.11	11	37.5	6	.303
RV	38.89	7	62.5	10	
Fenestration					
N	35	7	37.5	6	1
Y	65	13	62.5	10	
Pulmonary artery reconstruction					
N	80	16	68.75	11	.47
Y	20	4	31.25	5	
Pre-Fontan ≥ moderate atrioventricular valve regurgitation					
N	100	20	93.75	15	.444
Y	0	0	6.25	1	
Single-stage Fontan					
N	68	13	47	8	.311
Y	32	6	53	9	

F, Female; *M*, male; *N*, no; *Y*, yes; *LV*, left ventricle; *RV*, right ventricle. *Fisher *P* = Fisher exact *P* value.

end-diastolic pressure [12 mm Hg; n = 1], and left pulmonary vein stenosis [n = 1]). The anatomic and hemodynamic parameters in these 8 patients were favorable before the first Fontan procedure and the reasons for the transition from favorable to unfavorable hemodynamics remain unclear. The reason for not attempting a second Fontan was unclear in the remaining 3 patients.

Second Fontan (n = 5). Five (14%) patients underwent the Fontan for a second time after a median of 4.4 years (IQR, 1.9, 13). No remediable cause was identified in any patient before the second Fontan. One patient with DORV and hypoplastic left ventricle underwent an ECC fenestrated Fontan at 4 years of age. It was taken down within 24 hours for low cardiac output. He subsequently underwent a second Fontan after 2.1 years. He underwent a heart transplant approximately 7 years later for Fontan failure. The remaining 4 patients were alive in NYHA I/II at a median of 10.8 years IQR (3.1, 19.1). Before the second Fontan, cardiac catheter studies in these 4 patients demonstrated a median pulmonary artery pressure of 11 mm Hg (range, 9-13 mm Hg) and median ventricular end diastolic pressure of 9 mm Hg (range, 8-10 mm Hg). The hemodynamic parameters were similar to the findings on the cardiac catheter studies undertaken before the first Fontan in all 4 patients.

Biventricular/1½ ventricle repair (n = 3). Two (5.5%) patients had a biventricular repair, of whom 1 patient was alive in NYHA 1 after 3.1 years and 1 patient was lost to follow-up. One patient had a 1½ ventricle repair and was alive in NYHA 1 after 19 years. Freedom from death/transplant after Fontan takedown was 59% at 1 year (CI, 41%-74%), 56% at 5 years (CI, 38%-71%) and 52% at 10 years (CI, 34%-68%) (Figure 3).

TABLE 4. Associations between continuous independent predictors and death following Fontan takedown

Predictor	Dead				Rank-sum *P**
	No		Yes		
	Median	IQR	Median	IQR	
PA pressure	13	11-14	13	11-15	.968
Ventricular EDP	9	8-11	9	6-12	.621
TPG	5	4.75-6	5	4.75-9	.418
Hemoglobin oxygen saturation	83.5	79-87	87	84.5-87.25	.394
Age at Fontan	4.41	3.33-5.95	5.01	3.86-6.24	.417
Weight at Fontan	17.8	15-19.7	15	12.9-20.2	.339
BSA at Fontan	0.713	0.66-0.75	0.68	0.58-0.75	.362

IQR, Interquartile range; *PA*, pulmonary artery; *EDP*, end-diastolic pressure; *TPG*, trans-pulmonary gradient; *BSA*, body surface area. *Rank-sum *P* = Wilcoxon rank-sum test *P* value.

DISCUSSION

From a clinical standpoint, having to take down the Fontan is disappointing. For patients undergoing single-ventricle "palliation," and their families, it is a catastrophe. Fontan takedown has been performed since the early years of our experience with this procedure and has been successful in salvaging the acutely unstable patient. However, the role of Fontan takedown is difficult to assess, as it is rarely performed and risk factors remain unknown. Furthermore, most patients have favorable pre-Fontan hemodynamics. There are only a limited number of reports on this rare procedure and long-term outcomes remain poorly defined.

The largest and the most comprehensive study to date has been reported by Almond and colleagues,[10] who described 53 Fontan takedowns over 27 years from 1979 to 2006. Takedown was performed intraoperatively in 12 (22%) patients, within the first postoperative month in 31 (58%) and between 1 month and 1 year in the remaining 10 (18%) patients.[10] Iyengar et al reported 14 (3%) Fontan takedowns from 413 procedures performed in Melbourne over 27 years from 1980 to 2007.[8] The incidence of Fontan takedown is similar to our study (2.3%) The incidence of takedown for atriopulmonary, lateral tunnel and extracardiac conduit Fontan procedures was also similar in both studies.[8,10] Murphy and colleagues[5] reported a lower incidence of Fontan takedown among 592 Fontan procedures performed between 1995 and 2009.[5] Early Fontan failure occurred in 11 patients (1.9%) of which 5 (0.8%) underwent early takedown to a superior cavopulmonary connection.

The timing of Fontan takedown varies between reports. In the study by Almond and colleagues,[10] takedown was performed during the Fontan procedure itself in 12 patients (22%) with a further 31 patients (58%) occurring within the first postoperative month and the remaining 10 (18%) occurring between 1 month and 1 year.[10] In the study by Iyengar and colleagues,[8] 8 (57%) of the takedowns were performed within 2 weeks, 4 (28%) between 2 weeks and 6 months with the remaining 2 occurring between 1 and

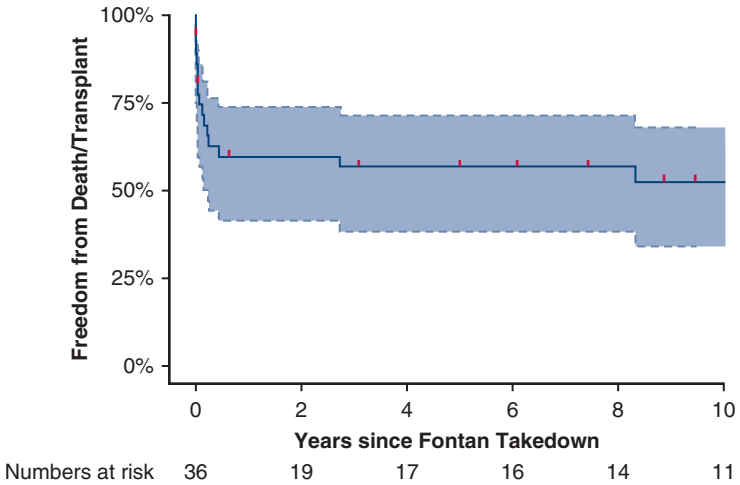


FIGURE 3. Freedom from death/transplant after Fontan takedown was 59% at 1 year (CI, 41%-74%), 56% at 5 years (CI, 38%-71%), and 52% at 10 years (CI, 34%-68%) (n = 36, 1975-2018).

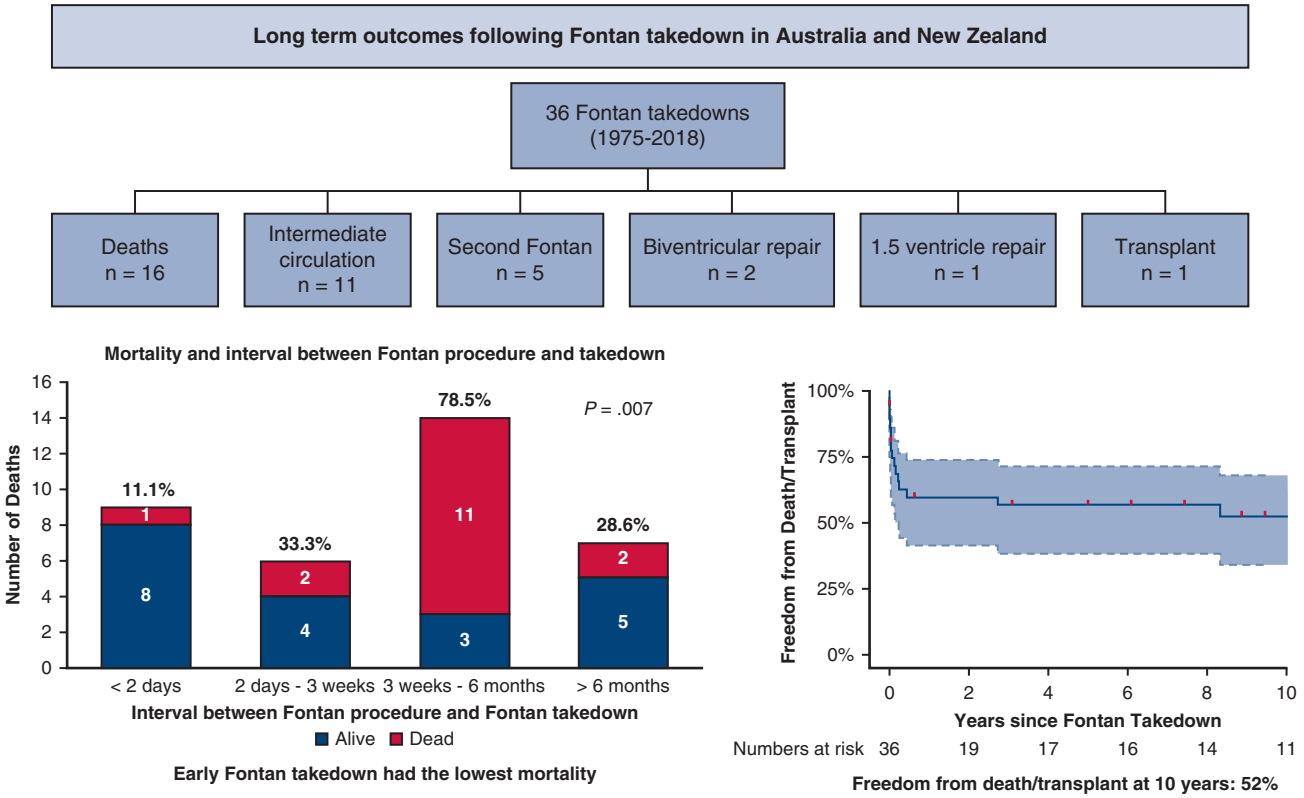


FIGURE 4. Fate of all 36 Fontan takedown patients operated between 1975 and 2018. Overall mortality was 44%, 31% stayed alive with an intermediate circulation, whereas 14% patients had a second Fontan. Mortality after early Fontan takedown was low whereas it was highest for takedowns between 3 weeks to 6 months. Freedom from death/transplant at 10 years was 52%.

2 years after the index procedure. The report from Murphy and colleagues[5] did not describe the incidence of Fontan takedowns beyond 30 days. More recently, Trezzi and colleagues[11] reported 18 Fontan takedowns for early Fontan failure between 1990 and 2015.[11] Takedown was performed during the Fontan procedure itself in 2 patients (12%) and within the first 2 postoperative months in the remaining 16 patients (88%). These reports are similar to our study, in which the majority of takedowns occurred early after the Fontan procedure with only 20% performed after 6 months.

The reported mortality for early Fontan takedown varies between 36% and 66%.[5,8,10,12,13] This is in contrast to our study, where the mortality was comparatively low (11%) when the Fontan was taken down within 48 hours. The highest mortality occurred between 3 weeks and 6 months (79%). This is similar to the report by Trezzi and colleagues[11] wherein 17 (94%) of the 18 Fontan takedowns to a BCPC survived the early post-takedown period. There is limited information on the outcomes of patients who have a takedown beyond 3 to 4 weeks of the Fontan. In the study by Almond and colleagues,[10] patients who died after later Fontan takedown were more likely to have been operated for persistent pleural effusions.[10] In our study the group of patients who underwent a Fontan takedown between 3 weeks and 6 months of age commonly presented with persistent pleural effusions and had a disproportionally high risk of mortality (79%). Most of these patients were subject to multiple procedures in an attempt to treat the effusions before taking down the Fontan circulation. Similar to other reports, our data suggest that there is an early window of opportunity to perform the Fontan takedown with a low mortality. Beyond this period, there is a substantial increase in mortality. We recommend that early takedown be considered sooner than later in patients with high-volume pleural drainage in the early postoperative period.

It is highly unlikely that meaningful risk prediction for Fontan takedown will ever be possible, given the small number of patients in every study. Previously identified risk factors include atriopulmonary Fontan and right dominant ventricular morphology.[10] In our study, there was no statistical association between any pre-takedown characteristic and mortality. Furthermore, pre-Fontan hemodynamic data were available in 86% of patients. The mean pulmonary artery pressure was <15 mm Hg in the majority of patients and no patient had greater than moderate ventricular dysfunction or greater than moderate atrioventricular valve

regurgitation. In our opinion, the factors contributing to the need to take down the Fontan circulation remain unknown and poorly understood. As technical imperfections cannot be ruled out as a contributing factor, we suggest that these should be aggressively ruled out or surgically addressed in the early postoperative period.

Patients who are taken down to an intermediate circulation appears to have an acceptable survival. In the study from Boston, 8 survivors (27%) were alive at a median of 9.7 years post-takedown.[10] In the study from Melbourne, there were 2 long-term survivors (22%) at a median follow-up of 9 years.[8] Trezzi and colleagues[11] reported 8 survivors (80%) at a median of 6.3 years post takedown, all with reasonable hemoglobin oxygen saturations (median 84%). In our study, there were 11 patients (31%) with an intermediate circulation at a median of 9.4 years post takedown, 10 of whom were in NYHA I/II. Previous reports have suggested that a cavopulmonary shunt or aortopulmonary shunt or both are an acceptable form of long-term palliation in a patient with single ventricle physiology, with 10- and 20-year survival between 80% and 90% and 50%, respectively.[14,15]

Interestingly, Fontan takedown does not appear to preclude a second attempt at Fontan completion.[10] In the series by Almond and colleagues,[10] a high proportion of patients were identified with correctable abnormalities that may have contributed to the initial Fontan failure. The longest follow-up in this group has been reported by Trezzi and colleagues.[11] All 3 patients were alive in NYHA 1 at a median follow-up of 10.7 years.[11] In our study 5 (14%) of patients underwent a second Fontan and 4 of them were alive in NYHA I/II at a median follow up of 10.8 years. Similar to other authors, we recommend that patients undergo investigation to identify and treat potentially remediable factors including branch pulmonary artery stenosis, arrhythmias, thrombus, pulmonary arteriovenous malformations, arch obstruction or hemi-diaphragmatic paresis prior to an attempt at a second Fontan.

Anecdotal reports of enlarging the fenestration in patients with early Fontan failure have been previously described, but the long-term outcome of these patients remains uncertain.[16,17] Another approach includes rescue cardiac transplantation for acute Fontan failure. Chaudhari and colleagues[18] have used this approach in 6 patients with 1 intra-operative death and 5 mid-term survivors (6-81 months).

Limitations

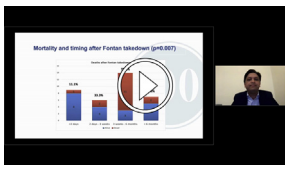
Our study bears all the limitations of a retrospective study spanning over 4 decades. Even though we did not find an era effect, the substantial changes in the approach to the Fontan procedure, decision making, surgical techniques and postoperative management over the past decades are highly likely to have influenced outcomes.

CONCLUSIONS

The incidence of Fontan takedown is low, but overall mortality is high. Factors influencing Fontan takedown remain unclear. The majority of Fontan takedowns occurred within 6 months with the greatest incidence between 3 weeks and 6 months. The mortality of this rescue procedure was 11% when performed within 2 days and increased to 33% and 79% when performed between 2 days and 3 weeks and between 3 weeks and 6 months respectively (Figure 4). Long-term outcomes after takedown to an intermediate circulation are acceptable. A small proportion of survivors can undergo a successful second Fontan. We should have a low threshold for Fontan takedown in patients who demonstrate signs of early failure within days of the procedure.

Webcast

You can watch a Webcast of this AATS meeting presentation by going to: https://aats.blob.core.windows.net/media/20AM/Presentations/Long-Term%20Outcomes%20Following%20Fontan.mp4.



Conflict of Interest Statement

Professor d'Udekem is a consultant with MSD and Actelion. All other authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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Key Words: Fontan palliation, single-ventricle palliation, Fontan failure, Fontan takedown

CONG

Discussion

Presenter: Dr Supreet P. Marathe

Dr Damien J. LaPar *(New York, NY)*. Dr Marathe and his colleagues from Australia and New Zealand present a 43-year experience with a very complicated patient cohort: those with Fontan failure who ultimately undergo Fontan takedown. The authors' centers certainly have an extensive surgical as well as published experience with the Fontan operation, including more than 1500 patients, and they have a leading Fontan registry that has once again served an important purpose in capturing and providing longitudinal outcomes for patients undergoing single-ventricle palliation. For this analysis, the authors have examined the group of patients that have vexed almost every congenital heart surgeon in their practice to provide some insight into outcomes following surgical strategies for Fontan failure.

The authors have analyzed 36 patients who underwent Fontan takedown, which represents an overall incidence of 2.3%. The key findings of this analysis include the incidents of Fontan takedown over time that is nearly unchanged by surgical era, acceptable pre-Fontan hemodynamics, a median time to takedown of 26 days, and a high mortality rate of 44%. Certainly, these characteristics represent the various clinical challenges that patients represent for all surgeons. Based on these findings and their analysis, I do have a couple questions for Dr Marathe, which I'll ask one at a time.

First, based on your data, you report that the most common indications for Fontan takedown are low cardiac output and intractable pleural effusions, both of which often present in the immediate and early postoperative periods. However, the majority of takedowns in your series occurred between 3 weeks and 6 months following Fontan. Considering that many have argued that outcomes for failing Fontan are improved with as early takedown as possible, how do you think the timing of Fontan takedown in this series ultimately impacted the long-term fate for these patients?

Dr Supreet P. Marathe *(Brisbane, Australia)*. Thank you, Dr LaPar, for your question. I would like to point out that the group of patients who had a takedown between 3 weeks and 6 months had more or less a common theme. All of them presented with intractable effusions, and they underwent multiple interventions—multiple chest tubes and pleurodesis, and they were sat on, and we did not take the Fontan down and then they ultimately died. So that was the most important take-home message for us, that we probably should not wait on these patients. The fact that they are developing these effusions points toward subtle markers that something is not right. Even if it looks fine on echo, it looks optimum on cath, the numbers are all okay, but the circulation is probably not right. It probably indicates that we should not shy away from taking down the Fontan, and as we can see, there are many other options after we take down the Fontan which we can provide them with.

Dr LaPar. Thank you. Also, related to the high frequency of low cardiac output in these patients, what is your center's experience? I didn't see it reported in your series for fenestrations at the time of Fontan.

Dr Marathe. We did not identify the presence of a fenestration to be predictive of takedowns. There was no difference regards to fenestration between the ones who died or had a transplant and the ones who did not.

Dr LaPar. Okay, great. My second question centers upon those patients undergoing takedown: Your late Fontan

failures for protein-losing enteropathy or plastic bronchitis. In this experience, while it's a relatively small number of patients, did the Fontan take down in these patients ultimately improve those symptoms?

Dr Marathe. Yes, they did.

Dr LaPar. Your data also demonstrate that 22% of the Fontan takedown patients ultimately underwent either re-Fontan or one and a half or 2-ventricle repair after takedown, whereas 31% of the patients remain with an intermediate circulation. So, my question is: After Fontan takedown, what is your center's approach or protocol to re-evaluating these patients for either surgical repair or potential re-Fontan? Do you repeat the cath at a certain time interval, or are there certain hemodynamics or characteristics that you look for that might push you toward operating on these patients?

Dr Marathe. So, because there are several member institutes, I would not say that there's a common protocol with regards to evaluation. But, in general, if we talk about the philosophy, we should definitely be doing the cath if there's something obvious. I think we should completely re-evaluate whether these patients really need to be along the single-ventricle pathway, because as we know, around the world, aggressive biventricular repairs are being pursued. We only had 2 biventricular repairs and 1 one-and-a-half ventricle repair in our series, but someone who had a single-ventricle palliation in the previous era might probably be okay with a biventricular circulation in the current era. So, we would also like to do magnetic resonance imaging to evaluate whether these patients are actually suitable for a biventricular repair; I think that should be the first step. The second step: If not, maybe we should consider them for a re-Fontan, and there are many ways; obviously, we can do aggressive atrioventricular valve repairs, we can optimize the pulmonary arteries, and there are many ways where we can make them good Fontan candidates if you don't find any obvious reason. The third option is, if we still are forced to stay with an intermediate circulation, that is where we wait.

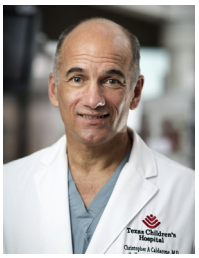
Dr LaPar. My final question is: Do your data, after analyzing it, provide you or your group any insight into which patients ultimately might do best? That is, survive or ultimately achieve a re-Fontan or a complete or partial repair after the Fontan takedown?

Dr Marathe. So, the aim was really this, we wanted to find out is there a common theme in them? Is there a basic diagnosis? Is there a predominant ventricular morphology which points toward these problems? But there's none, and the only difference is the timing of the takedown. Now, all this points to the fact that are there other things that we haven't looked at that we need to think about. They possibly could be subtle technical imperfections where the numbers are okay, but the flows are not optimal. So, what is the role of flow studies and whether we should consider doing them in these patients.

Dr Vaughn A. Starnes *(Los Angeles, Calif)*. Did you notice any era differences about when people took Fontans down? It's a study over 43 years; did you notice in the last decade that people took them down quicker? In the earlier decades, did people go on to die?

Dr Marathe. We did not find such a difference, no. The *P* value is trending toward significance, but not actually significant, though.

Dr. Starnes. Thank you.



Dr Christopher A. Caldarone *(Houston, Tex)*. The rationale for an early take down in a patient with low cardiac output syndrome might be different than one with prolonged pleural effusions. I didn't see that your analysis was stratified that way, but the message that you need to take down quickly may be more urgent in a low cardiac output state than it is in prolonged pleural effusions because, as you know, many patients have prolonged pleural effusions that resolve. So, how do you reconcile those 2 aspects of your analysis?

Dr Marathe. Absolutely. I cannot agree more. The ones who have early takedown, that is really a forced decision. We really don't have any other option. They have such low cardiac output, high inotrope requirement, it's really a last-ditch effort where we have to take them down. But this other group that we are talking about, we think they are fine, and we can potentially sit on them, but what our study shows is that we should probably not, and the most prudent way to go about them is to actually take down the Fontan.