



Dominant Ventricular Morphology and Early Postoperative Course After the Fontan Procedure

Uri Pollak, MD^{1,2,3,4} , Inbar Abarbanel, MD⁵, Yishay Salem, MD^{5,6}, Alain E. Serraf, MD, PhD^{5,7}, and David Mishaly, MD^{5,7}

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Abstract

Background: Single ventricle heart disease comprises a wide variety of critical heart defects that lead to the provision of systemic cardiac output by one dominant ventricle. It requires staged surgical palliation that culminates in Fontan circulation. Dominant ventricular morphology in single ventricle patients reportedly has an impact on postoperative morbidity and mortality with varying results. The objectives of this study were to examine the association between ventricular morphology and the early postoperative course after the Fontan procedure. **Methods:** A retrospective cohort study in a tertiary referral pediatric medical center that included 98 consecutive patients who underwent Fontan procedure between October 2009 and May 2016. Postoperative outcomes were compared between patients with left ventricular morphology and those with right ventricular morphology (crude effect and regression analysis). **Results:** Patients with right ventricular morphology had longer postoperative hospitalizations compared to patients with left ventricular morphology (26.5 days vs 18.2 days, respectively, $P=.028$), higher postoperative maximal vasoactive-inotropic scores (25.6 vs 12.4, $P=.02$), higher serum lactate levels (7.7 mmol/L vs 6.4 mmol/L, $P=.03$), higher proportions of ventilation throughout 24 h or more (16 patients [38%] vs 8 patients [14%], $P=.009$), higher proportions of ventricular dysfunction (12 patients [29%] vs 5 patients [9%], $P=.0001$), and lower blood oxygen saturation levels at discharge (87% vs 92%, $P=.03$). **Conclusions:** The Fontan procedure in patients with right ventricular morphology is associated with longer postoperative hospitalization and worse early postoperative characteristics (ventricular dysfunction and atrioventricular valve regurgitation) as well as higher rates of early, transient signs of sub-optimal postoperative hemodynamics compared to those with left ventricular morphology.

Keywords

congenital heart disease, pediatric cardiac surgery, single ventricle, ventricular morphology, Fontan procedure, perioperative outcomes

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Introduction

The Fontan procedure for the palliation of tricuspid atresia and an underdeveloped nonfunctioning right ventricle was first introduced in 1968.¹ Since the initial descriptions of the procedure, several surgical modifications have been made, along with the extension of its application for other types of functional univentricular congenital heart defects.^{2,3} Performing the anastomosis between the superior vena cava and the right pulmonary artery (the bidirectional Glenn procedure) as a transitional step at an earlier age decreased both mortality and morbidity to achieve a final Fontan circulation. The most recent modification of the technique was the insertion of an extracardiac conduit between the inferior vena cava and the right pulmonary artery.³

Dominant ventricular morphology in patients with a single ventricle has been designated to influence postoperative morbidity and mortality, but the results have been inconsistent.⁴⁻⁹ Some studies described right ventricular morphology (RVM) as being associated with increased mortality⁴ and prolonged

hospitalization⁵ following the Fontan procedure. The reports by Tweddell et al⁶ and Zhou et al⁷ on long-term survival, heart transplantation, and Fontan takedown found no

¹ Pediatric Cardiac Critical Care Unit, Hadassah University Medical Center, Jerusalem, Israel

² Pediatric Cardiology, Hadassah University Medical Center, Jerusalem, Israel

³ Pediatric Extracorporeal Support Program, Hadassah University Medical Center, Jerusalem, Israel

⁴ The Hebrew University Hadassah Medical School, Jerusalem, Israel

⁵ The Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

⁶ Pediatric Cardiology, Edmond J. Safra International Congenital Heart Center, The Edmond and Lily Safra Children's Hospital, The Chaim Sheba Medical Center, Tel Hashomer, Israel

⁷ Pediatric and Congenital Cardiac Surgery, Edmond J. Safra International Congenital Heart Center, The Edmond and Lily Safra Children's Hospital, The Chaim Sheba Medical Center, Tel Hashomer, Israel

Corresponding Author:

Uri Pollak, Section of Pediatric Critical Care, Hadassah University Medical Center, Ein Kerem, POB 12000, Jerusalem 9112001, Israel.
Email: uripol@hadassah.org.il; uri.pollak@gmail.com

Abbreviations

CHD	congenital heart defect
CPB	cardiopulmonary bypass
VIS	vasoactive inotropic score
LVM	left ventricular morphology
RVM	right ventricular morphology
PVR	pulmonary vascular resistance
AVVR	atrioventricular valve regurgitation
MV	mechanical ventilation
LOS	Length of hospital stay
PTFE	polytetrafluoroethylene.

differences between patients with left ventricular morphology (LVM) and those with RVM, whereas Moon et al⁸ demonstrated that RVM was negatively associated with long-term survival following the Fontan procedure, possibly due to a tendency toward progressive atrioventricular valve regurgitation (AVVR) and deterioration of single ventricle function. Ovroutski et al⁹ and then Nordmeyer et al¹⁰ identified RVM of the dominant ventricle to be associated with prolonged inotropic support and prolonged mechanical ventilation (MV) after the Fontan procedure.

The objective of this study was to retrospectively explore the association between dominant ventricular morphology and the early postoperative course and hospital outcomes of a cohort of patients who underwent the Fontan procedure in a single center between 2009 and 2016.

Patients and Methods

This study was approved by the Institutional Review Board of the Chaim Sheba Medical Center, Tel Hashomer, Israel. The requirement for informed consent was waived. This retrospective cohort study included all patients younger than 18 years of age who underwent the Fontan procedure at the Edmond and Lily Safra Children's Hospital of the medical center between October 2009 and May 2016. The sole exclusion criterion was indeterminate morphology of the dominant ventricle.

The Fontan Procedure

The uniform type of Fontan procedure in our cohort was the completion of the extracardiac conduit Fontan. Most of the patients underwent continuous cardiopulmonary bypass (CPB) with moderate hypothermia. First, the inferior vena cava was transected approximately 1 cm above the diaphragm and the cardiac end was oversewn, after which a polytetrafluoroethylene (PTFE) tube graft was connected to the inferior vena cava in an end-to-end anastomosis by means of a 5-0 monofilament suture. The conduit was then cut to the appropriate length with a slight bevel at the superior end. An anastomosis was constructed between that end and the pulmonary arteries. If the patient had previously undergone a bidirectional Glenn, the conduit was spatially connected to precisely conform to the underside of the right pulmonary artery.

If a fenestration was indicated, as in cases of pulmonary vascular resistance greater than 3 wood U/m², small pulmonary

arteries (Nakata Index < 250 mm²/m²), pulmonary artery pressure greater than 15 mm Hg, or according to the surgeon's impression and preferences, a side-to-side anastomosis between the conduit and the atrium was performed, preferably with a side hole of 6 mm or, if the anatomy does not permit direct anastomosis, by placing a 6 to 8 mm intervening segment of PTFE tube graft. If a fenestration had not been performed during the initial Fontan procedure but at a later occasion (usually within 48 h) due to a state of low cardiac output, it was referred to as a "late" fenestration.

Data Collection

Data were collected from the patients' electronic medical records with the application of Microsoft Excel 2016 (Microsoft Corporation) spreadsheets. Preoperative data included patient characteristics, as well as echocardiography and cardiac catheterization reports. Intraoperative data included the duration of CPB, the aortic cross-clamp time, and the creation of fenestration between the Fontan conduit and the atrium. Postoperative data included maximal vasoactive inotropic support (VIS: dopamine [μg/kg/min] + dobutamine [μg/kg/min] + 100 × epinephrine [μg/kg/min] + 10 × milrinone [μg/kg/min] + 10,000 × vasopressin [U/kg/min] + 100 × norepinephrine [μg/kg/min]), time to tracheal extubation, chest tube output, and length of hospital stay (LOS). All postoperative echocardiograms were independently and blindly interpreted by two pediatric cardiologists (UP and YS). Perioperative major adverse events included cardiac arrest, neurologic deficits, and the need for renal replacement therapy. Our follow-up period for each patient was defined as the time from the procedure until hospital discharge (which was our primary outcome). All variables were collected according to this time period. If patients were hospitalized prior to their surgery that period of time was not counted as part of our follow-up.

Statistical Analysis

The distributions of variables were presented by means and standard deviations for continuous variables and by frequencies for categorical ones. LOS was transformed into a log scale to improve agreement with normal distribution. The effect of the VM on the continuous variables was tested twice, first by a *t* test with unequal variances and then by backward linear regression. The initial list of variables in the regression included all potentially important covariates. As for the binary outcomes, the relationship between the outcomes and the VM was tested by the Fisher exact test for the corresponding 2 × 2 frequency table. The backward logistic regression was applied with the same list of potential covariates. The crude effect of VM on discrete variables was calculated with the Fisher test instead of the χ^2 test due to the low occurrence of adverse outcomes in the study population. The variable of VM was fixed in the model to observe its effect after adjustment for other covariates in each regression. Only the final condensed model was presented for each outcome. All tests were two-sided. *P* values were

calculated either by Fisher test for discrete variables or the *t* test for continuous variables and considered significant at $<.05$. Corrections for multiple comparisons were also performed. All calculations were carried out with STATA SE software (StataCorp LLC), and all results are described as mean (standard deviation), unless stated otherwise.

Results

All 98 consecutive patients met the inclusion criteria and were enrolled in the study. Fifty-six patients (57.1%) had LVM and 42 patients (42.9%) had RVM. The preoperative and intraoperative characteristics of both groups are summarized in Table 1. The preoperative anatomical findings and any previous surgical interventions are summarized in Table 2.

Crude Effect of the VM

The crude effect of the VM was tested on several postoperative clinical, laboratory, and echocardiographic parameters. Overall,

patients with RVM had longer LOS (26.5 days vs 18.2 days for patients with LVM, $P = .028$), a higher proportion of prolonged (24 h or more) MV (16 patients [38%] vs 8 patients [14%], respectively, $P = .009$), a higher proportion of ventricular dysfunction (12 patients [29%] vs 5 patients [9%], $P = .0001$), a higher postoperative maximal VIS (25.6 vs 12.4, $P = .02$), a higher mean serum lactate level (7.7 mmol/L vs 6.4 mmol/L, $P = .03$), and a lower mean blood oxygen saturation level at discharge (87% vs 92%, $P = .03$). The clinical results are summarized in Table 3 and the laboratory results are summarized in Table 4. Other adverse outcomes included the use of extracorporeal membrane oxygenation in one patient who died in close proximity to the surgery and a tracheostomy in another patient, both from the RVM group. The low prevalence of those adverse outcomes precluded testing their statistical significance.

Regression Analysis

Simple linear regression was used for continuous variables, and logistic regression was used for categorical variables (Table 5). The effect size for VM refers to the RVM group, thus a positive effect size indicates a larger average value of that variable among patients with RVM. LOS (in log scale due to its more closely resembling Gaussian distribution) was significantly

Table 1. Pre- and Intraoperative Characteristics According to Ventricular Morphology.

Preoperative data	LV morphology (n = 56)	RV morphology (n = 42)	P value
Sex, male	30 (53.6%)	28 (66.7%)	.22
Age, months	68.2 (40.4)	59.1 (25)	.17
Weight, kg	18.4 (9.3)	16.1 (5)	.11
Time from stage 2 procedure, months	41.4 (24.6)	42.5 (20.4)	.81
Noncardiac anomalies	11 (19.6%)	8 (19%)	.20
Blood oxygen saturation, %	81 (5)	82 (6)	.53
Hematocrit, %	47 (4)	47.6 (6.1)	.54
Mean pulmonary artery pressure, mm Hg	10.9 (1.2)	12.5 (1.8)	.68
End-diastolic pressure, mm Hg	7.8 (1.1)	9.1 (2)	.26
Pulmonary vascular resistance, Wood unit·m ²	1.5 (0.3)	1.76 (0.4)	.41
Atrioventricular valve regurgitation			
Mild	10 (17.9%)	9 (21.4%)	.31
Moderate	3 (5.3%)	4 (9.5%)	
Severe	0	0	
Intraoperative data			
Fenestration: n			
Yes	31 (55.3%)	20 (47.6%)	.54
No	24 (42.8%)	15 (35.7%)	.53
Late	1 (1.8%)	7 (16.7%)	.02
Conduit diameter, mm	19.4 (1.1)	19.2 (1.4)	.43
Cardiopulmonary bypass time, min	66.9 (27.7)	75.2 (41.1)	.27
Aortic cross-clamp	4 (7.1%)	6 (14.3%)	.32

All data presented as number of patients (%) or mean (standard deviation) unless stated otherwise.

Table 2. Preoperative Anatomy and Previous Surgical Interventions.

Preoperative anatomy	LV morphology (n = 56)		RV morphology (n = 42)	
	TA	15	AA/MA	12
	PA	14	AA/MS	6
	TA/PA	12	AS/MA	5
	DILV	11	AS/MS	4
	DILV/ MGA	2	DORV/PA/ MGA	8
	UCAVC	2	UCAVC	5
	Unknown	0	Unknown	2
Previous surgical interventions				
Stage 1 palliation				
Blalock-Taussig-Thomas Shunt	41		0	
Norwood/Sano Shunt	0		24	
Norwood/ Blalock-Taussig-Thomas Shunt	3		3	
Damus-Kaye-Stansel	5		4	
Pulmonary artery banding	5		3	
None	2		8	
Stage 2 palliation				
Glenn	54		38	
Kawashima	0		2	
Damus-Kaye-Stansel + Glenn	2		1	
"One-step Fontan"	1		1	

Abbreviations: AA, aortic valve atresia; AS, aortic valve stenosis; DILV, double inlet left ventricle; DORV, double outlet right ventricle; MA, mitral valve atresia; MS, mitral valve stenosis; PA, pulmonary valve atresia; SVC, superior vena cava; TA, tricuspid valve atresia; MGA, malposition of the great arteries; UCAVC, unbalanced complete atrioventricular canal.

Table 3. Postoperative Parameters According to Ventricular Morphology.

Postoperative data	LV morphology (n = 56)	RV morphology (n = 42)	P value
Maximal vasoactive-inotropic score	12.4 (3.72)	25.6 (9.38)	.02
Time to extubation, hrs mean (range)	27.9 (0-480)	59.4 (0-480)	.1
Extubation in less than 8 h	31 (55%)	16 (38%)	.1
Extubation after 24 h or more	8 (14%)	16 (38%)	.009
Blood oxygen saturation, %	92	87	.03
Chylothorax	7 (12%)	8 (19%)	.4
Pericardial effusion	9 (16%)	2 (5%)	.11
Ventricular dysfunction			
Mild	4 (7%)	5 (12%)	<.001
Moderate	0	4 (10%)	
Severe	1 (2%)	3 (7%)	
Atrioventricular valve regurgitation			
Mild	18 (32%)	18 (43%)	.01
Moderate	2 (4%)	6 (14%)	
Severe	0	0	
Length of hospital stay, days	18.2 (12.87)	26.5 (20.77)	.028

All data are presented as number of patients (%) or mean (standard deviation) unless stated otherwise.

affected by VM, with the RVM patients having a longer LOS (coefficient = 0.3, $t = 2.6$, $P = .011$).

Comment

Several publications have suggested an effect of the dominant VM on long-term mortality and morbidity following the Fontan procedure.^{11,12} VM also reportedly impacts exercise capacity after the Fontan procedure, with the RVM patients having inferior exercise performance compared to LVM patients.^{13,14} In contrast, there is limited evidence of any differences in the early postoperative course following the Fontan procedure in this population, and the current published data are conflicting.^{10,15} In this retrospective study, we demonstrated that RVM is associated with longer LOS, higher rates of low cardiac output (as shown by means of unfavorable hemodynamics) and prolonged MV, and lower blood oxygen saturation levels (by both crude effect and regression analysis) compared to LVM (Figure 1).

The LOS for both the RVM and LVM groups was longer than previously reported because more than 60% of our cohort were patients from very low-income countries to which they returned after recovering from the Fontan procedure. Notably, our RVM patients had a longer LOS than our LVM patients. This finding is consistent with the data of McGuirk et al⁵ and Kamata et al.¹⁵ Also in accordance with

Table 4. Postoperative Laboratory Results According to Ventricular Morphology.

Laboratory test	LV morphology	RV morphology	P value
Lactate, max, mmol/L	6.4 (3.1-20)	7.7 (2.7-20)	.03
Bicarbonate, min, mmol/L	17.7 (8.1-24.9)	17.5 (8.4-26.6)	.2
Creatinine, max, umol/dL	105.2 (34.5-601.3)	130.9 (42.4-718.8)	.71

All data presented as mean (range).

Table 5. Regression Analysis of Postoperative Variables With Right Ventricular Morphology.

Postoperative variable	Coeff	Stderr	t	P	95% CI
Maximal vasoactive-inotropic score	14.7	5.1	2.88	.005	4.5-24.8
Time to extubation	46.8	15.45	3.03	.003	16.1-77.6
Blood oxygen saturation	-2.8	1.5	-1.89	.048	-5.8-0.1
Chylothorax	1.4	0.9	0.54	.587	0.4-4.8
Pericardial effusion	0.1	0.1	-1.77	.077	0.0-1.3
Ventricular dysfunction	2.6	1.8	1.39	.002	0.7-10.2
Atrioventricular valve regurgitation	3.3	1.6	2.5	.012	1.3-8.6
Length of hospital stay (log scale)	0.3	0.1	2.6	.011	0.1-0.6
Infections	3.1	1.8	2.04	.042	1-9.4

Abbreviations: CI, confidence interval; Coeff, coefficient; P, P-value of t test; Stderr, standard error; t, result of t test.

the literature,^{16,17} we indicated that RVM patients required longer MV and more intensified hemodynamic support, as evidenced by higher VIS, with both interventions leading to prolonging the LOS. Persistent pleural effusion and chylothorax, which sometimes play a role in postoperative LOS after the Fontan procedure, were similar in our two study groups.

Postoperative ventricular dysfunction was associated more often with RVM than with LVM in our cohort. Matsuda et al⁴ described low cardiac output as a major concern in the early postoperative course after the Fontan procedure among their RVM patients. Anderson et al's¹⁸ mid-term results showed poor ventricular function in their Fontan patients with RVM who had higher fiber stress, a higher rate of ventricular dilatation, and lower circumferential fiber shortening, all directly relating to poor ventricular function. Higher ventricular volumes secondary to ventricular dilatation and lower mass-to-volume ratios were also identified by Ghelani et al, suggesting that differences in the myofiber architecture may contribute to the suboptimal adaptation of the RV as a dominant chamber.¹⁹ In a comparison of cardiac functional measures between single ventricle patients with LVM and RVM before first- and second-stage palliation, the progressive reduction in the RVM longitudinal and circumferential function suggested that RVM may have a mechanical disadvantage from birth

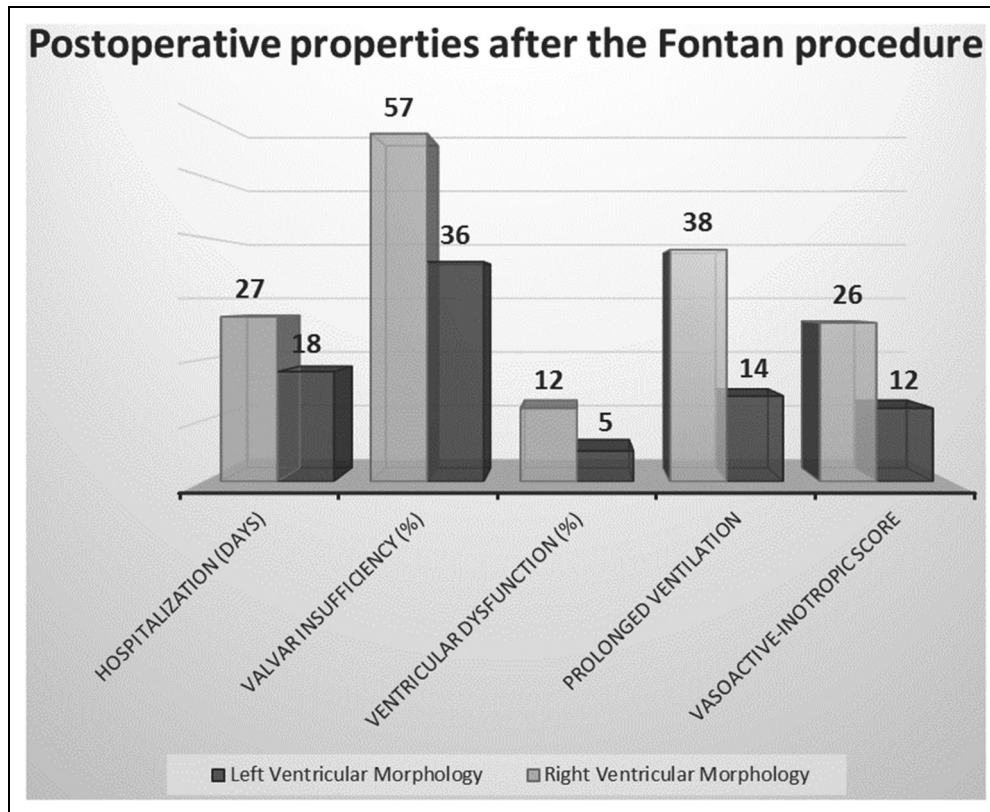


Figure 1. Postoperative properties after the Fontan procedure. Single ventricle patients with right ventricular morphology who undergo the Fontan procedure are prone to unfavorable hemodynamics, higher rates of prolonged mechanical ventilation (24 h or more), lower blood oxygen saturation levels, and longer postoperative hospitalization.

and progressive impairment with age.²⁰ Another study observed no difference in hemodynamics at pre-Glenn catheterization between RVM and LVM, although the RVM patients had higher ventricular end-diastolic pressure levels at pre-Fontan catheterization.²¹ Although not statistically significant (probably related to the small size of the study group), our RVM group of patients evidenced higher mean pulmonary artery pressures and higher end-diastolic pressures compared to the LVM group. These findings suggest that patients with RVM are already at a disadvantage to begin with. Whether this is related to the greater insult to the myocardium from the stage 1 palliative procedure and the need for myocardial incision (in the RV-to-PA conduit group) is a matter of conjecture. Intrinsic differences in morphology, function, and response to performing as the dominant ventricle between RVM and LVM may lead to an elevated ventricular end-diastolic pressure. This could limit pulmonary blood flow and coronary perfusion after the Fontan procedure and potentially lead to poorer performance on the part of the RVM patients.

The magnitude of inotropic support required in children with a single ventricle was shown to be affected by the reduced ventricular function.^{10,22} Nordmeyer et al described the need for longer inotropic support in RVM patients after the Fontan procedure compared with LVM patients.¹⁰ In a study by Ovroutski et al, RVM was an independent risk factor for the need for

prolonged postoperative inotropic support.⁹ In our study, a higher postoperative VIS was associated more often with RVM than LVM. We also measured relatively higher maximal serum lactate levels in the RVM patients, possibly indicating decreased perfusion and tissue oxygen delivery. Unfavorable hemodynamics in this group, in the presence of ventricular dysfunction and a higher VIS, may be the underlying contributing causes.

Fontan patients with RVM were shown by Anderson et al to have poor valvar function compared to Fontan patients with LVM.¹⁸ Our results indicated a higher proportion and increased severity of AVVR in the RVM group. The morphology of the tricuspid valve in hypoplastic left heart syndrome can differ from the valve seen in normal individuals.²³ These morphological differences can contribute to regurgitation of the valve, including bicuspid or quadricuspid valves, accessory orifices, leaflet malformation, leaflet prolapse, leaflet restriction or “tethering,” and wide variability of the supporting apparatus.²³ In addition to the morphological characteristics, other factors can contribute to the development and severity of the AVVR in this population, among them annular dilatation, right ventricular dysfunction, and valve prolapse.²⁴

Although the duration of MV after the Fontan procedure did not differ significantly between our RVM patients and LVM patients, there was a trend toward longer MV in the RVM

group. The proportion of patients that required prolonged (24 h or more) MV was significantly higher in the RVM group than the proportion of patients in the LVM group. Nordmeyer et al¹⁰ reported longer MV time in their RVM group. Those authors suggested that the reasons for the longer MV might be postoperative prolonged inotropic support and hemodynamic instability with dominant ventricular dysfunction which were present in their RVM group. Fluid overload, need for dialysis, and supraventricular tachyarrhythmia had been suggested as additional reasons for prolonged MV in this population,¹⁰ but our data were incomplete and not suitable for such an analysis. The need for longer intubation time is known to negatively influence Fontan hemodynamics by decreasing venous drainage into the pulmonary arteries, leading to intensified postoperative therapy and to higher postoperative morbidity as a result. Thus, minimizing the duration of MV remains a key goal in the postoperative management of Fontan patients.

Fenestration is associated with low mortality, less pleural effusion, and shorter hospital stay.^{25,26} In our current retrospective study, the intraoperative creation of fenestration was the same in both study groups, but late fenestration had been placed more often in the patients with RVM. The greater need for cardiovascular support in the RVM group can explain, to some extent, the need for a late fenestration. Nordmeyer et al¹⁰ also reported that fenestration had been placed more often in patients with RVM than in patients with LVM. The finding that the preoperative hemodynamic properties were similar between our two study groups can explain the lack of difference in any fenestration. The higher rate of late fenestration in the RVM group, however, raises the question of whether a fenestration should be placed in RVM patients as a matter of routine.

Blood oxygen saturation was significantly lower among our RVM patients, as similarly shown by others.¹⁰ Several explanations can be suggested for this finding. One is the higher proportion of fenestration in RVM patients causing a right-to-left shunt, and another is that RVM was more often associated with heterotaxy syndrome than LVM,²⁷ which was linked with ciliary dyskinesia, impaired pulmonary function and gas exchange.²⁸ There was a trend toward a greater linkage of heterotaxy syndrome in our RVM group, but without statistical significance (data not shown).

This study had several limitations. Its retrospective design resulted in some incomplete data for a small portion of the patients included in our cohort. Since we collected data over a long period of time, there could have been some alterations in the intra- and postoperative management of these patients. Another limitation was the heterogeneity of the patients with regard to previous cardiac surgical procedures, including a Blalock-Taussig-Thomas shunt, the Norwood procedure, and pulmonary artery banding. Another aspect of heterogeneity is the origin of the patients and the time point of referral for the Fontan procedure. This source of referral bias is planned to be a part of a future study, looking into the longer-term (one- and five-year) postoperative outcomes. Decisions, such as the timing of the Fontan operation and whether to fenestrate, as

well as the type of fenestration, were up to the discretion of the surgeon. We did not take into account the postoperative course of the first-stage palliation nor of the Glenn anastomosis: we tried to compensate for this limitation by standardizing the pre-Fontan properties as much as possible.

To conclude, preoperatively suitable Fontan patients demonstrated good postoperative results after an extracardiac Fontan procedure regardless of the VM. However, patients with RVM more frequently developed early, transient signs of sub-optimal hemodynamics, as indicated by the need for intensified cardiovascular support, the higher incidence of late fenestration creation, and the prolonged MV during the postoperative course. Patients with RVM were more prone to ventricular dysfunction as well as AVVR than patients with LVM, with both promoting early signs of Fontan failure. All the above contributed to prolonged LOS in RVM patients. Our findings support the preemptive optimization of Fontan circulation hemodynamic properties, including the implementation of all strategies for improving RV filling, reduction in the PVR, and maintenance of systemic perfusion pressure.


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ORCID iD

Uri Pollak  <https://orcid.org/0000-0002-4575-3888>

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