

## Original Article

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# Utilisation of a post-Fontan management pathway reduces chest tube drainage and hospital readmission rates

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## Abstract

**Background:** Complications following the Fontan procedure include prolonged pleural drainage and readmission for effusions. To address these complications, a post-Fontan management pathway was implemented with primary goals of reducing chest tube duration/reinsertion rates and decreasing hospital length of stay and readmissions. **Methods:** Fontan patients were identified by retrospective chart review (2017–2019) to obtain baseline data for chest tube duration/reinsertion rates, hospital length of stay, and readmission rates for effusion. A post-Fontan management pathway was implemented (2020–2021) utilising post-operative vasopressin, nasal cannula oxygen until chest tube removal, and discharge regimen of three times daily diuretics, sildenafil, and afterload reducing medications. Patients were followed to evaluate primary outcomes. **Results:** The pre- and post-pathway groups were similar in single ventricle morphology, demographics, and pre-operative haemodynamics. Forty-three and 36 patients were included in the pre- and post-pathway cohorts, respectively. There were statistically significant reductions in chest tube duration (8 vs. 5 days,  $p \leq 0.001$ ), chest tube output on post-operative day 4 (20.4 vs. 9.9 mL/kg/day,  $p = 0.003$ ), and hospital readmission rates for effusion (13[30%] vs. 3[8%],  $p = 0.02$ ) compared to baseline. There was an absolute reduction in hospital length of stay (11 vs. 9.5 days,  $p = 0.052$ ). When combining average cost savings for the Fontan hospitalisations, readmissions for effusion, and cardiac catheterisations within 6 months of Fontan completion, there was a \$325,144 total cost savings for 36 patients following pathway implementation. **Conclusion:** Implementation of a post-Fontan management pathway resulted in significant reductions in chest tube duration and output, and readmission rates for effusion in the perioperative period.

## Introduction

Patients undergoing Fontan palliation have been known to have complications including pleural effusions, thromboembolic events, liver dysfunction, arrhythmias, congestive heart failure, and progressive ventricular dysfunction.<sup>1</sup> Our study focused on decreasing the duration of persistent pleural drainage to improve hospital length of stay, readmission rates, and need for chest tube reinsertions.

Pre-operative and intraoperative factors have previously been shown to be associated with increased risk of persistent pleural effusions following Fontan palliation.<sup>2</sup> Pre-operative factors include lower pre-operative oxygen saturations, higher pulmonary vascular resistance or pulmonary artery pressures, and presence of significant aortopulmonary collateral vessels.<sup>2</sup> Intraoperative factors include longer cardiopulmonary bypass times and the presence or absence of a fenestration within the Fontan conduit.<sup>2–4</sup> Perioperative management of patients after Fontan palliation has also been studied extensively. Fontan protocols for post-operative care have been shown to reduce time to chest tube removal, and hospital length of stay.<sup>3,5,6</sup> Protocol interventions studied have included fat-restricted diets, limited total fluids, standardised diuretic therapy, implementation of pulmonary vasodilation (sildenafil),<sup>2,7,8</sup> or use of vasopressin in the immediate post-operative period.<sup>9,10</sup>

The purpose of our study was to establish and implement a post-operative management pathway for medical management of patients after Fontan palliation with pathway changes incorporated from the conclusion of the surgical procedure, through hospital recovery, and at the time of discharge home. We hypothesise that implementation of the post-operative management pathway would decrease hospital length of stay, duration of chest tube and pleural effusions, and decrease in hospital readmission rates for effusions.

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## Materials and methods

### Study population

This was a single-centre study conducted at a quaternary care children's hospital comparing patients who underwent fenestrated extracardiac or lateral tunnel Fontan operation before and after the implementation of a protocolised post-Fontan management pathway. This project was approved by the Vanderbilt University Medical Center Institutional Review Board (IRB# 200686; 4/16/2020). Individual consent was waived for this study. Patients were excluded from analysis if their Fontan palliation was performed outside of 2 to 6 years of age, was performed as a single-stage Fontan operation, or if the procedure was complicated by early in-hospital Fontan takedown or death.

We established a pre-pathway group through a retrospective chart review of patients undergoing Fontan palliation from 2017–2019 to serve as control. Patients who followed the implementation of the pathway from June 2022 to May 2023 were included in the post-pathway group. The post-Fontan management pathway was developed utilising the Key-Driver Diagram format in collaboration with cardiothoracic surgery, cardiology, cardiac anaesthesia, and cardiac ICU providers (Supplemental Figure 1). Our pathway was adapted from Pike and colleagues' protocol<sup>3</sup> at Children's Hospital of Los Angeles. The protocol was revised with provider input to reflect institutional practice variation prior to implementation. The primary pathway management changes included utilisation of vasopressin in the post-operative period, judicious use of crystalloid fluids for fluid resuscitation, early recognition of prolonged/large volume chest tube output, discharge on at least diuretics three times daily, sildenafil, and afterload reducing medications (angiotensin-converting enzyme inhibition).

### Post-operative management/protocol implementation

After departmental approval of the post-Fontan management pathway, the pathway was disseminated to members of the Pediatric Heart Institute prior to implementation in June 2020 (Table 1). Adherence to the pathway was monitored continuously by the primary investigator (Adam M. Skaff) with each Fontan patient from the time of surgery to discharge. Patients were removed from the pathway if an unexpected surgical complication occurred. Following hospital discharge, patients were followed as outpatients for readmission within 30 days of discharge for effusions, or catheterisation intervention within 6 months of Fontan completion. Referring cardiologists were contacted to inquire about local hospitalisations or complications with medical therapies after patient discharge. Discharge medications were managed at the primary cardiologist's discretion.

The criterion for chest tube removal was  $<9$  mL/kg/day for two consecutive days in both the pre- and post-pathway groups to limit variation. The patients were also monitored with daily metabolic panels as is customary in post-operative care to assess for kidney injury.

### Data collection

Chart review of the electronic health record was used to collect variables of interest including patient demographics (age, weight, gender), cardiac history (single ventricle morphology, diagnosis, history of prolonged effusions), pre-operative haemodynamics (catheterisation and echocardiographic data), and post-operative outcomes (post-operative length of stay, chest tube drainage

duration, chest tube reinsertion rate, and readmission rates) in both groups.

The primary outcome measures were chest tube duration, ICU and hospital length of stay, and readmission rate within 30 days of discharge for pleural effusions. Secondary outcome measures included chest tube output, catheterisation intervention within 6 months of Fontan, and average cost per hospitalisation, readmission, and catheterisation. Cost estimate information reflecting facility technical charges was obtained utilising the Pediatric Health Information System database (Lenexa, KS). Individual patient encounters and associated total costs for the Fontan admission, readmissions for effusion within 30 days, and cardiac catheterisations within 6 months were able to be identified.

### Statistical methods

Continuous data are reported as medians with interquartile ranges, and categorical data are reported as frequencies with percentages. Descriptive statistics were used to perform preliminary analysis of patient characteristics and outcomes in the pre- and post-pathway groups. Categorical variables were compared utilising  $\chi^2$ , and continuous variables were compared using the Mann-Whitney  $U$  test. Post-operative day one chest tube output and chest tube duration were monitored with statistical process control charts (Supplemental materials). Statistical analysis was performed using STATA/IC version 15.1 (StataCorp, LLC, College Station, TX). Statistical process control charts were created using quality improvement Macros 2023 (KnowWare International, Inc., Denver CO). Western Electric rules were used to identify special cause variation.

### Results

Forty-three and 36 consecutive patients were included in our pre- and post-pathway groups, respectively, minus exclusions. There were no significant differences between baseline and surgical patient characteristics including single ventricular morphology, underlying cardiac diagnosis, pre-operative haemodynamic parameters (mean pulmonary artery pressures, transpulmonary gradient, pulmonary vascular resistance, ventricular function, pre-operative saturations, atrioventricular valve function, and need for aortopulmonary collateral embolisation at pre-Fontan cardiac catheterisation), Fontan type, Fontan conduit size (18 mm), cardiopulmonary bypass, and aortic cross-clamp times (Table 2). Patients in the post-pathway were older in age (3.5 [3–4] vs. 3 [3–4] years,  $p = 0.02$ ) and had higher weight (15.6 [13.9–17.2] vs. 13.7 [12.3–15.4] kg,  $p = 0.02$ ) compared to the pre-pathway group.

Comparisons between the pre- and post-Fontan pathway groups are summarised in Table 3. The median chest tube duration was significantly lower in the post-pathway group compared to the pre-pathway group (5 [5–7] vs. 8 [7–10],  $p \leq 0.001$ ). Chest tube output was also decreased in the post-pathway group with 9.9 [5.1–21.5] mL/kg/d compared to the pre-pathway group with 20.4 [9.7–45.9] mL/kg/d, ( $p = 0.003$ ) (Figure 1). Characteristic of the chest tube drainage was not significantly different between the two groups. The need for chest tube reinsertion during hospitalisation was lower in the post-pathway group, however, did not reach statistical significance. Patients in the post-pathway were more likely to be extubated in the operating room (39% vs. 21%,  $p = 0.08$ ), had less amount of post-operative ventilator hours (6 [0–9] vs. 10 [5–27] hours,  $p = 0.02$ ), had decreased post-operative fluid balance in the first 24 hours (73.9 [27.6–93.1] vs. 92.5 [41.7–154.3] mL/kg/day,

**Table 1.** Vanderbilt Children's Hospital post-Fontan clinical management pathway

<p><b>Immediately Post-op:</b></p> <ul style="list-style-type: none"> <li>– Diuretics: IV furosemide q8h versus infusion after initial fluid resuscitation</li> <li>– Total Fluids at 25% maintenance</li> <li>– Vasopressin (0.02–0.04 units/kg/hr)- started in OR and continued for 12–24 hours post-op or until extubated</li> <li>– Epinephrine/Dobutamine in place of milrinone</li> <li>– Systolic blood pressure goal at least 25<sup>th</sup> percentile for age <ul style="list-style-type: none"> <li>• Plasmalyte for volume boluses up to 30 mL/kg, using 5% albumin after first 30 mL/kg</li> </ul> </li> <li>– FiO2 60% while intubated/NIPPV or 2L/min NC</li> <li>– Pace to maintain AV synchrony</li> <li>– Fentanyl/hydromorphone for pain (avoid morphine given histamine release)</li> </ul> <p><b>Step 2: after extubation</b></p> <ul style="list-style-type: none"> <li>– Discontinue vasopressin once through with volume resuscitation phase</li> <li>– PVR management- NC oxygen and sildenafil is started at 1–2 mg/kg divided TID</li> <li>– Diuretics: PO HCTZ/chlorothiazide + spironolactone BID, furosemide IV versus PO TID</li> <li>– Afterload reduction: enalapril/lisinopril/captopril**</li> <li>– Diet: Regular diet (low fat diet only for chylous/prolonged drainage), 50% MIVF until PO goal achieved</li> <li>– Respiratory support: extubate to NC and stay on O2 until chest tubes out. Minimum 1–2L/min NC (if agitated try while sleeping)</li> <li>– Chest tube removal: when on feeds and drainage is &lt;9 mL/kg/day for 2 consecutive days- per cardiothoracic surgery</li> <li>– Echo prior to discharge when chest tubes removed/PRN for ICU needs</li> <li>– Start ASA POD 1- send platelet inhibition assay after second dose. If not therapeutic, increase ASA dose until platelet inhibition adequate per assay parameters.</li> </ul> <p><b>Step 3: Preparation for discharge</b></p> <ul style="list-style-type: none"> <li>– Full enteral feeds or stability on therapeutic diet (low-fat diet for chylous drainage)</li> <li>– Continue minimum of TID loop diuretic ± thiazide ± potassium sparing diuretics (furosemide, HCTZ/chlorothiazide, bumetanide, spironolactone)</li> <li>– Continue ACE inhibitor at discharge if no contraindications</li> <li>– Continue sildenafil at time of discharge- discontinue per primary cardiologist at outpatient follow up.</li> </ul> <p><b>Daily studies during post-op:</b></p> <ul style="list-style-type: none"> <li>– AM CXR, BMP, Magnesium, Albumin</li> <li>– Echo post-op TEE and when chest tubes removed- assess AVVR, function, effusion</li> </ul> <p>*Wean diuretics or liberalise maintenance fluids for BUN/Cr changes including BUN&gt;40 or rise in Cr by&gt;100%.</p> <p>**Discontinuation of ACE inhibitor on individual basis for hypotension or AKI. If on multiple diuretics, cut back as tolerated on diuretics prior to ACE inhibitor discontinuation.</p> <p>***Chylous effusions: If concern for chylothorax/increased CT output- initiate low-fat diet and consult with surgeon of record.</p> <ul style="list-style-type: none"> <li>– Chylous output/prolonged drainage- defined as drainage volume of greater than 30 mL/kg/day on POD4, CT duration of &gt;8 days with increasing drainage volume, or a change in character AND volume of drainage with initiation of enteral feeds</li> <li>– Consider head/neck ultrasound ± Echo for thrombosis</li> <li>– Discuss need for lymphangiogram with persistent effusion despite treatment with low fat diet for 5 days</li> </ul> <p><b>Outpatient follow up needs:</b></p> <ul style="list-style-type: none"> <li>– Clinic follow up within 1 week of discharge with CXR and BMP</li> <li>– If started on therapeutic dietary modifications for chylous/prolonged effusions, coordinate nutrition visits with follow up visit.</li> <li>– All medications continued at discharge can be managed at the primary cardiologist's discretion at follow up.</li> </ul>
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IV, intravenous; NIPPV, noninvasive positive pressure ventilation; NC, nasal canula; AV, atrioventricular; PVR, pulmonary vascular resistance; PO, per mouth; BID, twice daily; TID, three times daily; HCTZ, hydrochlorothiazide; MIVF, maintenance IV fluids; ASA, aspirin; POD, post-operative day; ACE, angiotensin-converting enzyme; CT, chest tube; AM, morning; CXR, chest X-ray; BMP, basic metabolic panel; TEE, transesophageal echocardiogram; Cr, creatinine; AKI, acute kidney injury.

$p = 0.03$ ), and a significant reduction in incidence post-operative arrhythmia (36% vs. 60%,  $p = 0.03$ ) compared to the pre-pathway group.

Following the implementation of the protocolised post-Fontan management pathway there was special cause variation with a decrease in both chest tube duration and output noted on process control charts. A centreline shift with a decrease in total volume of post-operative day 1 chest tube output from an average of 56 to 37 mL/kg. average chest tube duration experienced two centreline shifts from 11.9 to 5.6 days (Supplemental Figure 2).

All patients in the post-pathway cohort were assessed for acute kidney injury as defined by Kidney Disease Improving Global Guidelines (KDIGO) Clinical Practice Guidelines for Acute Kidney Injury.<sup>11</sup> There was one admission for acute kidney injury in the setting of decreased oral intake following discharge. There were no other patients with acute kidney injury noted on outpatient follow up laboratory testing.

The total hospital length of stay was lower in the post-pathway group compared to the pre-pathway group, although did not reach statistical significance (9.5 [7.5–13.5] vs. 11 [9–17] days  $p = 0.052$ ). There was also a decrease in ICU length of stay in the post-pathway

group compared to the prepathway although not statistically significant (3 [2–6.5] vs. 5 [3–8],  $p = 0.13$ ). The readmission within 30 days was lower for the post-pathway group compared to the pre-pathway group (3 [8%] vs. 13 [30%],  $p = 0.02$ ). None of the patients readmitted in the post-pathway group required chest tube insertion compared to 38% in the pre-pathway group with one readmission in the post-pathway group related to diuretic non-compliance at home after discharge.

The need for catheterisation within 6 months of the Fontan operation was significantly decreased in the post-pathway compared to the pre-pathway group (0 [0%] vs. 11 [26%],  $p = 0.004$ ). The primary indications for catheterisation were haemodynamic assessment due to clinical concerns including recurrent or persistent effusions (4 patients), hypoxia (6 patients), or concern for left pulmonary artery stenosis by echocardiography (2 patients). Interventions at the time of catheterisation included left pulmonary artery angioplasty/stenting (8 patients), Fontan tunnel angioplasty/stenting (2 patients), and collateral embolisation (2 patients).

Thirty-two patients (89%) were discharged home on an angiotensin-converting enzyme inhibitor and three times daily

**Table 2.** Fontan patient characteristics and procedural information pre- and post-pathway implementation

Variable	Pre-pathway (n = 43)	Post-pathway (n = 36)	p-value
Age (years)	3.0 (3–4)	3.5 (3–4)	0.02
Gender			0.046
Male	19 (44%)	24 (67%)	
Female	24 (56%)	12 (33%)	
Weight (kg)	13.7 (12.3–15.4)	15.6 (13.9–17.2)	0.002
Ventricle type			0.29
Left	24 (56%)	15 (42%)	
Right	19 (44%)	20 (56%)	
Indeterminate	0 (0%)	1 (2%)	
Diagnosis			0.8
TA	12	7	
DILV	6	5	
DORV	5	6	
Unbalanced AVSD	2	3	
HLHS	15	10	
PA/IVS	1	3	
Other	2	2	
History of chylous/prolonged output	6 (14%)	6 (17%)	0.74
Pre-operative haemodynamics			
Mean PAP (mmHg)	9 (8–11)	10 (9–11)	0.08
TPG (mmHg)	4 (4–5)	4 (3.5–5)	0.73
PVR (iWU)	1.4 (1.2–1.7)	1.5 (1.2–1.7)	0.94
Pre-op collateral embolisation	15 (35%)	12 (33%)	0.89
≥Moderate AVVR	4 (9%)	2 (6%)	0.53
≥Mild ventricular dysfunction	5 (12%)	9 (25%)	0.12
Pre-operative saturations, %	83 (81–85)	83 (80–86)	0.68
Fontan type			0.71
Extracardiac	37	32	
Lateral tunnel	6	4	
Fenestrated	43	35	0.27
CPB time (minutes)	121 (108–165)	123 (98–154)	0.47
AoCC time (minutes)	0 (0–54)	0 (0–8)	0.14

Values are presented as n (%) or median (interquartile range).  $\chi^2$  p-values for categorical variables; Mann-Whitney U p-values for continuous variables. TA, tricuspid atresia; DILV, double inlet left ventricle; DORV, double outlet right ventricle; AVSD, atrioventricular septal defect; HLHS, hypoplastic left heart syndrome; PA/IVS, pulmonary atresia/intact ventricular septum. PAP, Pulmonary artery pressure; TPG, transpulmonary gradient; PVR, pulmonary vascular resistance; iWU, indexed Wood unit; AVVR, atrioventricular valve regurgitation; CPB, cardiopulmonary bypass; AoCC, aortic cross-clamp.

diuretics. Patients that were not discharged on an angiotensin-converting enzyme inhibitor or three times daily diuretics had medication adjustments due to medication contraindications per protocol. There were several patients with unrelated post-operative complications including complete heart block requiring pacemaker placement and management of anti-coagulation with a mechanical atrioventricular valve. These individual patient complications, which are not directly related to our study interventions, prolonged hospitalisation, and led to variation in

post-operative management from the post-Fontan management pathway.

Follow-up showed that most patients were weaned off diuretics over the first 1–2 months post-discharge. Sildenafil was typically weaned off slowly over 4–6 months post-discharge, using trends in oxygen saturations and fenestration patency/gradient as a marker of improving pulmonary pressures and Fontan haemodynamics. All patients in our study cohort were able to be prescribed sildenafil after prior authorisations were completed and approved.

**Table 3.** Post-Fontan pathway compliance (A) and clinical outcomes (B) pre- and post-pathway implementation

Variable	Pre-pathway (n = 43)	Post-Pathway (n = 36)	p-value
<b>A</b>			
Post-operative milrinone	32 (74%)	5 (14%)	<0.001
Post-operative vasopressin	18 (42%)	36 (100%)	<0.001
Post-operative epinephrine	38 (88%)	29 (81%)	0.34
Discharge Medications			
Sildenafil	7 (16%)	36 (100%)	<0.001
ACE inhibitor	21 (49%)	32 (89%)	<0.001
TID diuretics	24 (56%)	32 (89%)	0.001
Loop diuretic dosing (mg/kg/day)	3 (2–3)	3 (3–3)	0.03
<b>B</b>			
Post-operative arrhythmia	26 (60%)	13 (36%)	0.03
Need for pacing?	18 (42%)	13 (36%)	0.6
Post-operative CVP (mmHg)	16 (15–19)	14 (13–18)	0.02
Discharge echo ≥ Moderate AVVR	1 (2%)	0 (0%)	0.36
Discharge echo ≥ Mild ventricular dysfunction	6 (14%)	13 (36%)	0.02
Patent fenestration at discharge by echo	40 (93%)	33/35 (94%)	0.82
Extubation in operating room	9 (21%)	14 (39%)	0.08
Post-operative ventilator hours	10 (5–27)	6 (0–9)	0.02
Post-operative fluid balance, 24 hours (mL/kg)	92.5 (41.7–154.3)	73.9 (27.6–93.1)	0.03
Chest tube duration (days)	8 (7–10)	5 (5–7)	<0.001
POD 4 Chest tube output (mL/kg/day)	20.4 (9.7–45.9)	9.9 (5.1–21.5)	0.003
Chylous drainage	7 (16%)	6 (17%)	0.71
Chest tube reinsertion	7 (16%)	3 (8%)	0.29
ICU length of stay (days)	5 (3–8)	3 (2–6.5)	0.13
Total length of stay (days)	11 (9–17)	9.5 (7.5–13.5)	0.052
Readmission within 30 days	13 (30%)	3 (8%)	0.02
Readmission length of stay (days)	5.5 (4–10)	5.0 (5–6)	0.94
Readmission chest tube insertion	5/13 (38%)	0/3 (0%)	0.2
Cath intervention within 6 months	11 (26%)	0/33* (0%)	<0.001

Values are presented as *n* (%) or median (interquartile range).  $\chi^2$  *p*-values for categorical variables; Mann-Whitney *U* *p*-values for continuous variables. CVP, central venous pressure; ACE, angiotensin-converting enzyme; TID, three times daily; AVVR, atrioventricular valve regurgitation; POD, post-operative day; ICU, intensive care unit. \*At the time of submission 33 post-pathway patients were 6 months post-Fontan.

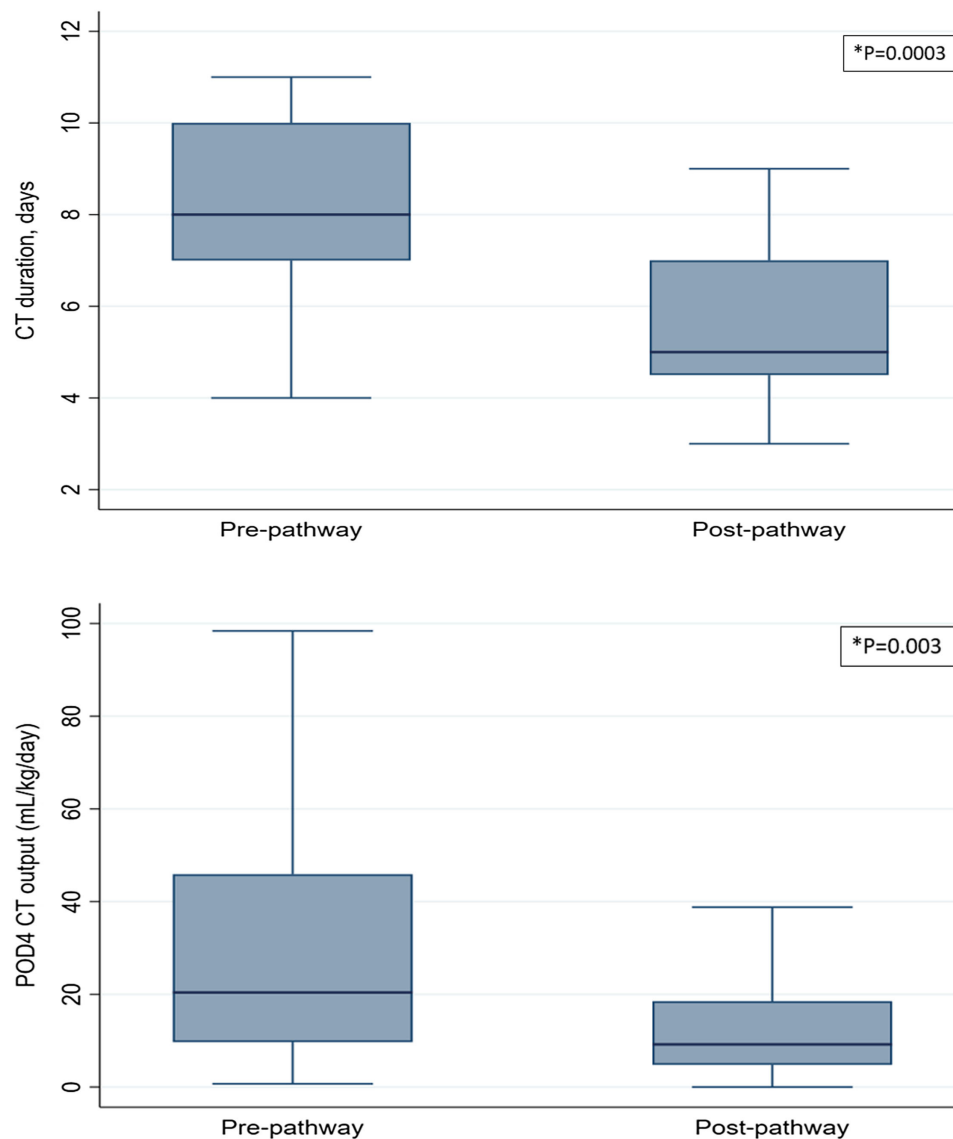
Following implementation of the post-Fontan management pathway, the total average costs for the Fontan surgical hospitalisation and readmissions for effusion were reduced by \$1,948 and \$3,187 per patient, respectively. Eleven patients underwent cardiac catheterisation within 6 months of Fontan completion for a total average cost of \$19,831 per catheterisation, compared to no catheterisations in the post-pathway group (Supplemental Table S4). When combining total average cost savings per patient for the Fontan hospitalisations, readmissions within 30 days for effusion, and cardiac catheterisations within 6 months of Fontan completion, there was a \$325,144 institutional cost savings following pathway implementation when adjusting for expected outcomes and post-pathway cohort size.

## Discussion

Our study highlights the benefits of implementing a protocolised approach to post-Fontan management. Our results demonstrated significant reduction in chest tube duration by 3 days, chest tube output, hospital length of stay by 1.5 days, and readmission rates within 30 days of discharge for effusion by 23%. There were no patients who required readmission for chest tube reinsertion.

Our findings support post-Fontan management protocolisation such as those described by Cava, Sunstrom, and Pike<sup>3,5,6</sup> which all showed successful protocol implementation and reduction in chest tube duration, chest tube output and a decrease in hospital stay with reduction in hospital costs. Our study adds to this limited literature additional findings including the addition of new pharmacologic therapies to the protocol that have been





**Figure 1.** Comparison between pre- and post-pathway chest tube duration and chest tube output on post-operative day 4 (POD4). The bold horizontal line indicates the median. The borders of the boxes indicate the 25th and 75th percentiles for each patient group. The whiskers indicate the minimum and maximum values, excluding the outliers. Outliers are not depicted to better display differences in pre- and post-pathway cohorts.

independently reported to improve chest tube drainage in the Fontan circulation, as well as the introduction of a quality improvement approach to the standardisation of the process in a quaternary care centre. At our centre, there were significant differences in daily management between individual providers which ultimately affected length of stay and the need for readmission with chest tube reinsertion prior to study protocol implementation.

Management of pleural effusions in this population continues to be complicated. Gupta *et al.* described hydrostatic, hormonal, and inflammatory mechanisms as contributors to persistent pleural effusions.<sup>2</sup> The aetiology of pleural effusions is multifactorial and influenced by poor pulmonary vascular compliance and the unknown effects of the single ventricle's circulation in series. The alterations implemented in our protocol compared to previous pathways highlight the importance of continuing to target pulmonary haemodynamics and pulmonary vascular compliance.<sup>12</sup> Our protocol also had novel features including performance as a prospective analysis and included updated strategies such as the initiation of vasopressin in the immediate post-

operative period and the standardised use of sildenafil during hospitalisation.

We incorporated the use of vasopressin in our protocol based on the evidence for decreasing chest tube output after the Fontan operation as reported by Kumar *et al.*<sup>9</sup> Their findings suggest that vasopressin ameliorates the effects of inflammation caused by cardiopulmonary bypass and reducing capillary leak due to its ability to increase vascular tone by acting on V1 receptors in skin, skeletal muscle, and small bowel and additional positive effects of coronary and pulmonary vasodilation by promoting release of endothelial nitric oxide while increasing myocardial contractility. Vasopressin safety and efficacy in the post-Fontan management has been described previously with no difference seen in blood pressure, heart rate, catecholamine use, or fluid administration when compared to a placebo-control group and was associated with decreased chest tube drainage and lower transpulmonary gradient.<sup>10</sup> Vasopressin use may allow for blood pressure support without the need for excess fluid resuscitation, helping to maintain a less positive fluid balance in the immediate post-operative period. The combination of tight blood pressure control, less crystalloid

usage during fluid resuscitation, likely helped to maintain a more euvoletic state without the need for extra diuresis after the initial post-operative period without causing renal injury and is likely reflected in the decrease in the reduction of mean central venous pressures in the immediate post-operative period.

Sildenafil is a selective phosphodiesterase type-5 inhibitor that increases cyclic guanosine monophosphate and relaxes vascular smooth muscle and is approved for the treatment of pulmonary hypertension in adults.<sup>13</sup> The use of sildenafil in older paediatric Fontan patients has previously shown a decrease in pulmonary artery pressure, pulmonary vascular resistance, and transpulmonary gradient when assessed by catheterisation without affecting ventricular performance.<sup>7</sup> The effect of the use sildenafil pre-operatively in CHD has been reported and a meta-analysis by Fikri et al., that analysed 233 patients from four studies that were treated pre-operatively with sildenafil 1–2 weeks before repair of a ventricular septal defect that showed decreased mean pulmonary artery pressures, shorter cardiopulmonary bypass time and decreased duration of mechanical ventilation and length of ICU stay.<sup>14</sup> The inclusion of sildenafil in our protocol is based on the potential therapeutic target for decreasing pulmonary vascular resistance in the post-operative period induced likely by the effects of cardiopulmonary bypass. Our study supports the use of sildenafil and vasopressin in this population with a decrease in chest tube output and duration, likely due to improved pulmonary haemodynamics.

Patients in the post-pathway group also had decreased use of milrinone compared to the pre-pathway group, and less incidence of post-operative arrhythmia. Milrinone was only utilised in the post-operative period for ventricular or systemic atrio-ventricular valve dysfunction on post-operative transesophageal echocardiogram. The use of milrinone in the post-operative period has been shown to be independently associated with post-operative tachyarrhythmia.<sup>15</sup> While the cause of post-operative arrhythmia is likely multi-factorial, maintenance of AV synchrony is associated with lower venous pressures which may reduce effusion burden.

Cardiac catheterisation in the 6-month follow-up after Fontan operation was as high as 26% in the pre-pathway group with over a third of those indicated for recurrent pleural effusion. There were no patients in the post-pathway group requiring catheterisation in that time frame. Catheter interventions in the post-operative Fontan population have previously been reported with intervention for physiologic indications being associated with prolonged pleural drainage.<sup>16</sup> Our protocol does incorporate therapies aimed at the reduction of central venous pressure and improved diuresis in the immediate post-operative period which may contribute to decreased need for catheterisation due to recurrent effusions.

The results of these practice changes affect the early post-operative care, but also have significant implications for long-term outcomes. Longer ICU/hospital length of stay and chest tube duration have been associated with long-term mortality and increased reintervention in this population.<sup>17</sup> Another long-term study by Downing et al. also demonstrated that ICU stay >1 week was associated with increased risk factors for post-Fontan death, transplant, or takedown both at <1 year and >1 year after operation on univariate and multi-variable analysis.<sup>16</sup> This is hypothesised to be due to suboptimal haemodynamics, as increased pulmonary artery pressures over 15 mmHg were the strongest predictor of early morbidity.

The introduction of quality improvement efforts in health care has helped provide better care for patients and reduce costs. Efforts

in quality improvement across the care for children with CHD have become essential, aimed to drive process change in delivery of specialised care.<sup>18</sup> Our incorporation of the quality improvement process allowed for adherence and consistency in the review process of the change in practice at our institution and can serve as blueprint for large multicentre studies to address these challenges in the Fontan population.

The financial implications and cost savings evidenced from this study show a modest total cost savings per patient observed during the post-pathway period for the initial Fontan hospitalisation and cost for readmission for effusion within 30 days of discharge. The Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) reports an average hospital length of stay following Fontan of 12.7 [11.3–14.9] days. In our post-pathway group, there was a median length of stay of 9.5 [7.5–13.5] days, reflecting a decrease in overall hospital costs. Our cost data however were only reported as total combined hospital costs and not broken down into costs for hospital stay, medications, or interventions. The cost reduction for readmissions within 30 days for effusion may also be a product of the ability to treat recurrent effusions with diuresis as opposed to the need for chest tube insertion at time of readmission. The most significant source of cost savings between the pre- and post-pathway cohorts was the decrease in cardiac catheterisations needed within 6 months of Fontan completion to evaluate haemodynamic concerns in patients with recurrent effusions. Continued studies to improve post-operative Fontan management will help short- and long-term outcomes in these complex patients.

### Limitations

This study was conducted as a single-centre, historically controlled study, with retrospective chart review of a pre-intervention cohort and comparison to a non-randomised prospective intervention cohort and may incur selective bias. All retrospective information gathered was obtained by chart review and gaps in documentation or variation in echocardiogram or catheterisation interpretation can contribute to error. Patients undergoing Fontan palliation for single ventricle physiology are heterogenous and matching cohorts for evaluation is difficult. The differences in pre-operative age and weight were likely attributable to delays in Fontan palliation due to the COVID-19 pandemic as there were restrictions to performing elective surgical cases during our study period. Our study period also reflects a change at our centre of primary surgeon performing most operations. Surgical technique varies between surgeons and degree of pulmonary artery augmentation at the time of the Fontan was not controlled for. In addition, early extubation practices by anaesthesia in the post-pathway group is a significant limitation of our study.

### Conclusions

Prolonged pleural effusion and readmissions for effusion after Fontan palliation continue to be a significant post-operative problem. Implementation of a post-Fontan management pathway, from the immediate post-operative period to discharge, resulted in significant reductions in chest tube duration, chest tube output, and readmission rates for effusion in the perioperative period.

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**Competing interests.** None.

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