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Anesthesia Management of Scoliosis Correction in an Adolescent with Fontan Circulation Secondary to Hypoplastic Left Heart Syndrome: A Case Report

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ABSTRACT: Fontan Scoliosis Anesthetic Challenges For any child with a Fontan circulation, anesthetic challenges for correction of thoracic deformity, when combined with single-ventricle physiology and the potentially for compromising transpulmonary flow and cardiac output, can lead to increased complexity. We present the perioperative and anesthetic course for an 11-year-old girl with HLHS status post extracardiac, nonfenestrated Fontan, who underwent surgery for correction of adolescent idiopathic scoliosis. An 11-year-old girl with HLHS underwent correction of scoliosis using total intravenous anesthesia with propofol and fentanyl without paralysis, enabling intraoperative neuromonitoring; intraoperative ventilation, hemodynamic goals, and anesthetic management focused on maintaining adequate transpulmonary blood flow; the intraoperative 4-hour operation remained relatively bloodless and hemo-dynamically stable, the patient remained free of need for any pressor support, and was intubated and extubated prior to departure to pediatric intensive care. This patient and this report contribute to the sparse literature available concerning the anesthesia care for scoliosis correction in patients with Fontan physiology.

INTRODUCTION

Scoliosis is present with disproportionate frequency among children with congenital heart disease compared with the general pediatric population and occurs with an even greater prevalence among Fontan survivors with single-ventricle physiology.[1] The Fontan circulation is the terminal surgical palliative operation for a single-ventricle heart, which is constructed by passively rerouting systemic venous return directly to the pulmonary artery, obviating entirely a subpulmonary ventricle.[2] Since no ventricular pump drives blood through the pulmonary circulation, blood flow to the lungs - and, therefore, cardiac output - depends on low pulmonary vascular resistance (PVR) and sufficient preload to produce a favorable transpulmonary pressure gradient.[3].

Deforming spinal column Progressive scoliosis causes an intrinsic decline in the margin of safety in this already narrow physiologic field. Chest scoliosis contributes to a restrictive pattern of lung volume and low chest wall compliance over time, which results in an increasing PVR and reduced pulmonary blood flow and decreased ventricular preload and cardiac output.[4,5]. Several authors thus recommend that surgeons take into account whether or not curves are amenable to surgical correction at less than the typical 50-degree threshold for structurally normal patients, since correction of large curves is more difficult and leads to increased compromised cardiopulmonary reserve.[6] Nonetheless, posterior spinal fusion itself - which often necessitates prolonged operative time, the use of prone positioning, positive-pressure mechanical ventilation and a

potential risk of significant intraoperative bleeding - poses an additional threat to a precarious balance of Fontan physiology.[7] Herein, we report the approach of our institution to the perioperative and anesthetic management of a child with hypoplastic left heart syndrome and a Fontan circulation undergoing surgical correction of idiopathic scoliosis.

CASE REPORT

Patient Demographics and Diagnosis

Age	11 years
Sex	Female
Weight	40.7 kg
Height	137 cm
Primary cardiac diagnosis	Hypoplastic left heart syndrome, status post Fontan palliation
Comorbidities	Right-sided hemiplegia; adolescent idiopathic thoracolumbar scoliosis

Past Medical and Surgical History

The patient's had diagnosed this as hypoplastic left heart syndrome, palliated in a staged single-ventricle strategy, and this was closed with an extracardiac non-fenestrated Fontan procedure. The operative procedures are reviewed below

Date	Procedure
22 May 2014	Aortic arch augmentation, pulmonary artery banding (PAB), and patent ductus arteriosus (PDA) ligation
15 June 2015	Atrial septectomy, main pulmonary artery (MPA) ligation, and right bidirectional Glenn (superior cavopulmonary) anastomosis
04 April 2019	Pre-Fontan cardiac catheterization
11 April 2019	Extracardiac non-fenestrated Fontan completion with pulmonary vein exclusion

Her stroke on the 14 August 2015 resulted in an acquired right-sided hemiplegia. Adolescent idiopathic thoracolumbar scoliosis, was diagnosed which later needed correction.

Current Medications

Medication	Dose	Route/Frequency
Warfarin	5 mg	PO, once daily
Lisinopril	2.5 mg	PO, once daily
Furosemide (Lasix)	20 mg	PO, once daily

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Preoperative Cardiac Evaluation

Transthoracic echocardiography done on 12/03/25 shows patent Fontan and patent Glenn with flow within normal limits (unobstructed) and good interatrial communication. There is mild regurgitation of the right atrioventricular valve and mild aortic regurgitation, with a mild velocity across the left ventricular outflow tract, with no significant gradient. The systemic outflow tract and aortic arch were un obstructued.

There was mild to moderately reduced ventricular systolic function and no pericardial effussion.

A 12-lead electrocardiogram obtained on 10/12/25, preoperatively during surveillance of patient's cardiac status. The 12-lead electrocardiogram (ECG) demonstrated normal sinus rhythm with a heart rate of 88 beats per minute. Notable findings included signs of right ventricular hypertrophy (RVH) with right axis deviation, which is commonly anticipated in patients with palliated hypoplastic left heart syndrome (HLHS). There were no acute ST-segment or T-wave abnormalities, and the PR and QTc intervals were within normal limits for her age

Reason for admission

The patient is being admitted preoperatively to the pediatric service, to address and optimize her cardiopulmonary system in preparation for her scoliosis repair.

The surgical team wishes for her to have surgery in the early part of next week.

Patient has been treated with medication and the medical management has been maximized with appropriate regard to Fontan physiology, and she is stable.

It is not desired that her scoliosis progresses further due to impact on her lung capacity and Fontan circulation. The risk for worsening on Fontan circulation associated with thoracic deformity decreases pulmonary compliance and can increase pulmonary vascular resistance, which in turn leads to decreased pulmonary blood flow and, eventually, lower output. In patients with pulmonary arterial resistance above an upper limit of 25 wood units on invasive hemodynamic monitoring at rest with reduced cerebral oxygenation (indicated by abnormal transcutaneous oximetry), even curves less than 50 degrees represent a valid indication for surgery as correction improves these parameters by optimizing lung volumes.

Cobb Angle and Indication for Surgical Management Standing spine x-ray images taken on 10/07/25 show the thoracic curve has an estimated Cobb angle of 48 degrees (thoracolumbar curvature).

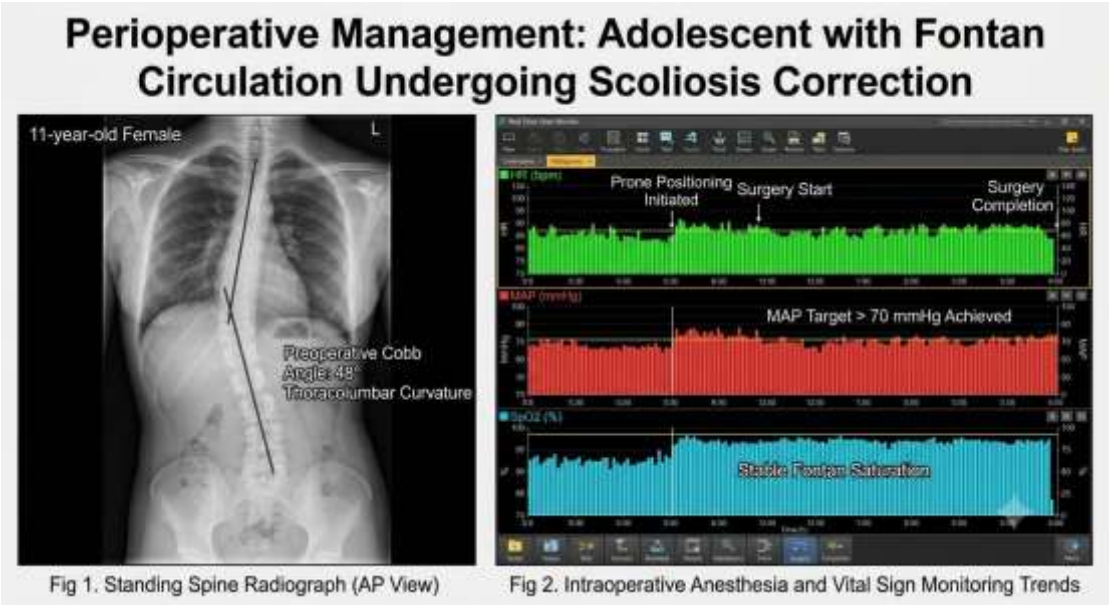


Fig 1. Standing Spine Radiograph (AP View)

Fig 2. Intraoperative Anesthesia and Vital Sign Monitoring Trends

Correction surgery is recommended to avoid potential decline inFontan physiology due to effects of severe chest wall deformities on pulmonary function. Even with mild degrees of curvature below 50%, the potential for progression and

progressive cardiopulmonary decline is thought to be a valid indication for surgical correction in patients with complex single ventricle anatomy such as this.



Fig 3.The goal of the posterior spinal fusion extending from the fourth thoracic vertebra (T4) to the first lumbar vertebra (L1) was to prevent any further deterioration in pulmonary function and compromise to the Fontan circulation.

Preoperative Investigations

Preoperative workup Completed blood count, coagulation panel, and chemistry was obtained The preoperative laboratory findings were as follows:

Complete Blood Count (CBC): Hemoglobin: 14.1 g/dL, Hematocrit: 42%, White Blood Cell (WBC) count: 6.2 \times 10^9/\text{L}, Platelets: 185 \times 10^9/\text{L}.

Coagulation Profile: International Normalized Ratio (INR): 1.2 (on day of surgery, post-reversal), Prothrombin Time (PT): 13.8 seconds, Activated Partial Thromboplastin Time (aPTT): 34.2 seconds.

Basic Metabolic Panel (Chemistry): Sodium: 138 mEq/L, Potassium: 4.1 mEq/L, Chloride: 101 mEq/L, Blood Urea Nitrogen (BUN): 14 mg/dL, Creatinine: 0.52 mg/dL.

The patient's Fontan management and concomitant anticoagulation with warfarin has been reviewed with cardiology as she is requiring operative intervention. Following consultation with pediatric cardiology, a structured anticoagulation bridging protocol was implemented: Warfarin was discontinued 5 days prior to surgery. Once her INR fell below 2.0, bridging therapy was initiated with Low-Molecular-Weight Heparin (LMWH, Enoxaparin) at a therapeutic dose of 1 mg/kg subcutaneously every 12 hours. The last dose of LMWH was held 24 hours prior to the scheduled surgery. Oral Vitamin K (1 mg) was administered 24 hours preoperatively to ensure the INR was optimized to a target of < 1.3 on the morning of surgery. Postoperatively, LMWH bridging is planned to resume 24 to 48 hours after surgical hemostasis is secured, with gradual re-introduction of Warfarin. Chest x-ray was performed. The preoperative chest radiograph revealed mild cardiomegaly with stable, chronic postoperative changes of the sternum from previous sternotomies. Pulmonary vascular markings were within normal limits, and there was no evidence of acute pulmonary congestion, active parenchymal disease, pleural effusion, or atelectasis. The trachea was midline. Preoperative vital signs taken on the morning of 12/06/2025 were Heart Rate (HR) of 82 beats per minute, Blood Pressure (BP) of 106/64 mmHg, Respiratory Rate (RR) of 16 breaths per minute, Temperature of 36.8°C, and Oxygen Saturation (SpO_2) of 94% on room air (which is expected for her non-fenestrated Fontan physiology).

Anesthesia Preoperative Orders Preoperative laboratory workups to include complete blood cell count, coagulation profile and comprehensive metabolic panel.

Preoperative chest radiograph to evaluate for any thoracic disease. Preoperative electrocardiogram to ensure there is no ongoing or acute process of cardiac ischemia/infarction. Preoperative TTE was performed on 12/03/2025 showing a normal, well functioning Fontan circulation.

It is essential that the medical optimization of this patient's physiology continues into her admission. The cardiologists on the case were consulted on management of anticoagulation due to the patients use of warfarin to maintain patency of Fontan

circulation. It is crucial that during her surgical intervention there is no intraoperative loss of consciousness that leads to a significant period of hypotention which can decrease cerebral perfusion and risk stroke in a patient with single ventricle.

Furthermore the anesthesiologist on the case has developed specific plans to maintain cerebral blood flow throughout the duration of her surgery as well as strategies for management of the patient's anticoagulation throughout the course of her admission, per cardiology.

Anesthetic Management

Anesthesia Management After informed consent was obtained, the patient was brought to the operating room. General anesthesia was induced using intravenous propofol 60 mg and fenta nyl 100 mcg. Tracheal intubation was achieved with a size 6.0 cuffed endotracheal tube. An intravenous line was placed peripherally and a right radial arterial line was placed to monitor arterial pressure.

Intraoperative neuromonitoring equipment was placed on the patient, who was then positioned prone.

Total intravenous anesthesia was utilized throughout the procedure, maintaining anesthesia with infusions of propofol at 150 mcg/kg/min and fentanyl at 2 mcg/kg/hr. Muscle relaxants were not utilized intraoperatively to allow continuous neuromonitoring. The antifibrinolytic agent tranexamic acid was administered intraoperatively at a rate of 10 mg/kg/hr to minimize blood loss. To minimize the risk of perioperative cerebrovascular accidents in the setting of her prior stroke, cerebral oximetry via near-infrared spectroscopy (NIRS) was monitored continuously, successfully maintaining regional oxygen saturation (rSO2) within 10% of baseline throughout the procedure. Surgical correction was achieved via a posterior spinal fusion extending from T4 to L1, optimizing spinal alignment while mitigating long-term pulmonary compromise.

Prophylactic intravenous cefazolin was administered 1000 mg preincision. Mean arterial pressure was maintained above 70 mmHg, without use of vasopressors or inotropes. Maintenance fluids were administered at 200 mL/hr of Ringer's lactate, with two 100 mL boluses provided as necessary for intraoperative fluid optimization.

A low tidal volume (5 mL/kg) and a low level of PEEP (3 cmH2O) were used throughout mechanical ventilation, avoiding hypercapnia due to its potential to elevate pulmonary vascular resistance.



Fig.4: vital sign monitoring trends

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The goals for the anesthetic case are listed in table 1.

Table 1. Intraoperative anesthetic goals applied in this Fontan patient.

Domain	Intraoperative goal / action
Airway/ventilation	Low tidal volume (5 mL/kg) with low PEEP (3 cmH2O); avoidance of hypercapnia to limit any rise in pulmonary vascular resistance (PVR)
Hemodynamics	MAP target >70 mmHg; no vasopressor or inotropic support required intraoperatively
Fluid management	Ringer's lactate maintenance at 200 mL/hr, with two 100 mL boluses(total fluid administered: 1000 mL) for optimal preload and volume optimization
Coagulation	Tranexamic acid infusion at 10 mg/kg/hr as an antifibrinolytic to limit surgical blood loss
Neuromonitoring	No muscle relaxant used at induction or during maintenance to preserve motor-evoked potential monitoring
Infection prophylaxis	Cefazolin 1000 mg IV administered prior to surgical incision
Emergence	Fully awake extubation at the end of the procedure

The duration of surgery was roughly 4 hours with a small total amount of blood loss. Postoperative hemoglobin was 137 g/L. At the end of surgery, the patient was fully awakened and extubated to 2 L/min supplemental oxygen by nasal cannula and was transferred to the pediatric intensive care unit and was transferred to the pediatric intensive care unit (PICU). The patient remained in the PICU for 3 days for hemodynamic monitoring and pain management. She was successfully weaned to room air on postoperative day 1. Importantly, the patient did not develop any Fontan-specific complications, such as pleural or pericardial effusions, or chylothorax during her hospital stay. The patient was discharged home in stable condition on postoperative day 7.

DISCUSSION

Several principles consistent with the limited literature regarding scoliosis surgery in the child with a Fontan circulation are illustrated in this case. Firstly, as there is no right-sided pump to actively drive blood through the lungs and rely is placed on the transmural gradient to do so, any factor that increases pulmonary vascular resistance can significantly decrease pulmonary blood flow and cardiac output, these include hypoxia, hypercapnia, acidosis, hypothermia, and positive end expiratory pressure (PEEP). Similar concerns have been highlighted for posterior spinal fusion in a single-ventricle patient with heterotaxy syndrome and a Fontan conduit with emphasis on hyperventilation pre- incision, avoidance of hypothermia, and management of any of the triggers listed for increased PVR using a total intravenous anesthetic technique with maintenance of muscle tone in order to facilitate neuromonitoring.

Secondly, maintenance of preload and a sufficient central venous pressure is crucial to maintain trans pulmonary flow as placing the patient in the prone position or any use of positive pressure can compromise venous return to the circulation by reducing this pressure gradient. In an adolescent patient undergoing posterior spinal fusion for scoliosis, intra-operative hypotension was encountered shortly after placing the patient prone, this resolved after volume resuscitation and short-term administration of adrenaline. Maintenance of a fixed rate fluid infusion, supplemented with boluses in this patient as required allowed preservation of stable hemodynamics with no requirement for any inotropes or vasopressors.

Thirdly, Minimizing the duration of surgery and total blood loss are repeatedly identified as strategies to limit peri-operative complications in the child undergoing spinal fusion for scoliosis in the setting of CHD. Tranexamic acid, an antifibrinolytic agent (used in this patient), is a commonly used strategy to minimise blood loss in pediatric spine surgery and has been utilised in the management of patients with Fontan physiology.

Fourthly, a rapid (ideally fully awake) extubation post-surgery is preferred as pulmonary blood flow is increased during spontaneous breathing, as spontaneous breathing has a suckling action on the pulmonary circulation in relation to positive pressure breathing, as was demonstrated in this case at the end of surgery with excellent post-operative hemodynamics.

Finally, a specific issue arises with anticoagulation in children with a Fontan circulation and those on chronic warfarin medication, as the risk of thromboembolism through the Fontan circuit needs to be balanced with the risk of surgical bleeding, necessitating close pre-operative liaisons between the cardiology and surgical teams to achieve safe per-operative anticoagulation. Other pre- operative optimization in similar complex cases such as adequate cardiovascular, pulmonary and hematological assessment is highlighted to promote uneventful post operative recovery.

Overall this case provides support for feasibility of performing posterior spinal fusion in a child with a single-ventricle Fontan physiology when an anesthetic plan is developed that proactively addresses potential issues of elevated pulmonary vascular resistance, preload, maintaining hemodynamics and facilitates early, awake extubation.

CONCLUSION

The anesthetic management of the child undergoing posterior spinal fusion with a Fontan physiology must be designed to accommodate the unique characteristics of passive, PVR and preload driven blood flow through the pulmonary circulation. In this 11-year-old patient with hypoplastic left heart syndrome and Fontan palliation, a TIVA anesthetic without muscle relaxation, lung protective ventilation without the development of hypercapnea, appropriate fluid resuscitation, an anti-fibrinolytic regimen and rapid extubation allowed a smooth and hemodynamically stable intraoperative course and an uncomplicated post-operative transition to recovery.

Declarations

Ethics:

Funding:

Conflicts of interest:

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