Case report

Increased blood loss during posterior spinal fusion for idiopathic scoliosis in an adolescent with Fontan physiology

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Summary
A 15-year-old patient with Fontan physiology experienced major blood loss during posterior spinal fusion for idiopathic scoliosis. Contributing factors for the increased blood loss and potentially useful measures to limit blood loss in patients with Fontan physiology are discussed.

Keywords: scoliosis; Fontan physiology; spinal fusion; blood loss; hemostasis; child

Introduction
Idiopathic scoliosis occurs in 2–4% of the general population and spinal fusion with instrumentation is used to stop progression of the deformity (1). The surgery carries a small risk of neurological injury and may lead to major blood loss with need for transfusion of blood products (2). Advances in the management of complex congenital heart disease are giving rise to a growing population of patients with palliated heart lesions. The Fontan procedure is the final stage to palliate a variety of complex congenital defects with functional single ventricles. We report on an adolescent with Fontan physiology who underwent posterior spinal fusion for idiopathic scoliosis and who experienced excessive intraoperative blood loss because of markedly elevated venous pressures in the prone position despite use of a Jackson table. A chronically elevated prothrombin time (PT), which is not uncommon in patients with Fontan physiology may have contributed to the magnitude of the blood loss. We discuss additional measures that may allow lesser blood loss in scoliosis patients with this challenging physiology.

Case report
A 15-year-old male with Fontan physiology presented for posterior spinal fusion (T4-L1) with instrumentation. His scoliosis was diagnosed at 8 years and managed conservatively with a rigid thoracolumbosacral brace. At 14 years of age during the pubertal growth spurt he experienced rapid progression of the scoliosis curvature (see Figures 1 and 2) and was advised to undergo posterior spinal
fusion with instrumentation. His medical history was significant for complex congenital heart disease with D-transposition of great vessels, tricuspid and pulmonary atresia and single left ventricle equivalent. The patient underwent staged single ventricle palliation with a final Fontan operation with an intraatrial venous conduit at 5\(\frac{1}{2}\) years of age (see Figure 3). His medications were captopril 12.5 mg twice a day and 81 mg acetylsalicylic acid once daily. The latter was stopped 10 days prior to surgery. The patient had not received thromboprophylaxis with coumadin for many years. On preoperative anesthesia examination he was 168 cm tall and weighed 56 kg. His oxygen saturation in room air was 91%. Preoperative laboratory tests showed hemoglobin 15.1 g\(\text{dl}^{-1}\), hematocrit 44.5%, platelets \(195 \times 10^9 \text{ l}^{-1}\), PT 15.1 s, international

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Figure 1
A scoliosis survey shows a curvature measuring 54° to the right in the thoracic spine and 36° to the left in the lower thoracic spine. Sternotomy wires are visible from cardiac surgery.

Figure 2
The PA scoliosis survey shows the postoperative correction and hardware in place.
normalized ratio (INR) 1.63, partial thromboplastin time (PTT) 29.7 s and bleeding time 11.5 min. The PT corrected to 12.4 s (normal) with a 1 : 1 normal plasma dilution. After 2 weeks, a repeat coagulation profile on the morning of surgery was essentially unchanged except that with cessation of the acetylsalicylic acid the bleeding time had normalized to 7.5 min. The preoperative transthoracic echocardiogram showed excellent function of the left ventricle and patent Fontan channels. Pulmonary function tests showed a forced vital capacity (FVC) of 2.21 l (60% of predicted), forced expiratory volume in 1 s (FEV) of 1.72 l (51% of predicted) and a FEV/FVC ratio of 78% suggestive of moderately severe

Figure 3
The diagram shows the typical sequence of palliation for patients with hypoplastic right heart syndrome secondary to tricuspid atresia. The final Fontan palliation consists of an intracardiac (3) or extracardiac (4) inferior vena cava to pulmonary artery connection. Our patient had undergone three previous cardiac palliative surgeries. However, as his tricuspid atresia was associated with transposition of the great arteries and aortic coarctation his first surgery was not a Blalock Taussig shunt (as shown in 1) but a coarctation repair and pulmonary artery banding, because his pulmonary blood flow was not restrictive but excessive (not shown). Subsequently, he had a superior vena cava to pulmonary artery (bidirectional Glenn) connection (2) and finally an intraatrial tunneled inferior vena cava to pulmonary artery (later tunnel Fontan) connection (3). In Fontan palliation the systemic veins from the upper and lower body are directly connected to the pulmonary circulation without the benefit of a pulmonary ventricle. Central venous pressure thus equals pulmonary arterial pressure and is chronically elevated leading to a number of pathophysiological alterations as discussed (diagrams courtesy of Elliot May, Chief Cardiovascular Surgery PA, Children’s Hospital of Wisconsin).
pulmonary restrictive disease which was entirely consistent with his significant scoliosis.

On the morning of surgery general anesthesia was induced intravenously with propofol (140 mg), rocuronium (50 mg) and sufentanil (5 μg). Monitoring included invasive arterial blood pressure, central venous pressure (CVP) (is equal to pulmonary artery pressure in Fontan patients) via a right internal jugular 5F catheter, somato-sensory and motor-evoked potentials and urine output. Anesthesia was maintained with isoflurane (0.4–0.8%) in an oxygen–air mixture, sufentanil infusion (0.1–0.3 μg·kg⁻¹·h⁻¹) and rocuronium. The patient was turned prone on a Jackson table and care was taken to allow for unimpaired abdominal movement (see Figure 4). The initial arterial blood gas in the prone position on fraction of inspired oxygen (FiO₂) of 50% showed a pH 7.49, PCO₂ 3.6 kPa (28 mmHg) PO₂ 15.7 kPa (121 mmHg), HCO₃ 21.6 mmol·l⁻¹, SaO₂ 98%, base deficit −0.4. Minute ventilation was adjusted to normocapnia. Soon after incision steady bleeding and poor clot formation was noted, which was managed with two adult units (500 ml) of fresh frozen plasma. Cell saver volume and blood loss within the first hour exceeded 700 ml and transfusion with whole blood was started. The initial supine CVP was elevated at 15–18 mmHg and increased further after prone positioning to the 20–25 mmHg range where it had to be kept to avoid arterial hypotension. Bleeding was continuous and increased during the bone decortications requiring ongoing replacement with blood products and cell saver blood. Oxygenation on FiO₂ of 0.4–0.5 remained in the partial pressure of arterial oxygen (PaO₂) range of 10–15 kPa (82–121 mmHg). Total intraoperative blood loss was estimated in excess of 3.5 l (62 ml·kg⁻¹ body weight). The intraoperative transfusions included two adult units of whole blood (900 ml), nine adult units of packed red blood cells (2250 ml), 2800 ml of cell saver blood, eight adult units of fresh frozen plasma (1600 ml) and two full doses of single donor pheresed platelets (which contain at least 1.1 × 10¹¹ platelets). Intraoperative coagulation studies showed a platelet count of 148 × 10⁹ ·l⁻¹, PT 15.6 s, INR 1.76, PTT 32.9 s, fibrinogen 148 mg·dl⁻¹ and D-dimer 0.2–0.4 μg·ml⁻¹. Despite ongoing transfusion bleeding did not slow down until the wound was closed and the patient was turned supine which resulted in a decrease of the venous pressures to the mid-teens mmHg range. During surgery the patient remained hemodynamically stable and normothermic. No dysrhythmias were noted and he remained in normal sinus rhythm throughout the perioperative period. He was awakened and extubated in the operating room with a temperature of 37°C.

The postoperative course was remarkably stable. Total wound drain output in the first 12 h was only 150 and 130 ml in the subsequent 24 h. The drains were discontinued after 48 h. On postoperative day 5 the patient was stable with a hemoglobin of 9.7 g·dl⁻¹, hematocrit of 27.8% and a platelet count of 130 × 10⁹ ·l⁻¹. He was transfused with two adult
units of packed red blood cells (500 ml) and discharged home on postoperative day 7.

**Discussion**

The average intraoperative blood loss during spinal fusion surgery for idiopathic scoliosis in children and adolescents is 800–1200 ml, i.e. less than half a blood volume (3). Factors correlating with increased blood loss in adolescents include surgical technique, duration of surgery, number of vertebral levels fused, site of autologous bone graft harvest, mean arterial pressure, and patient position (3). Bleeding in scoliosis surgery is thought to be mainly venous (4,5). The preoperative indicators for increased intraoperative red cell transfusion include low preoperative hemoglobin, history of pulmonary disease, surgery for tumors, large number of vertebral levels to be fused and positioning, specifically not using a Jackson table (6). The Jackson table helps to minimize intraabdominal pressure and thus venous pressure in the epidural veins during the prone position (see Figure 4).

A recent study has shown that intraoperative use of ε-aminocaproic acid significantly reduces perioperative blood loss in patients with idiopathic scoliosis undergoing posterior spinal fusion and instrumentation (3). Our orthopedic surgeons have a very busy scoliosis practice but do not favor the routine use of antifibrinolytics. In addition our cardiologists are concerned about inducing a hypercoagulable state in Fontan patients with antifibrinolytic agents in noncardiac settings (see below).

Although experience with scoliosis surgery in Fontan patients is still too limited to assess whether Fontan physiology is another risk factor for excessive blood loss, the elevated venous pressures as a result of largely passive flow of blood through the pulmonary circulation are likely to significantly contribute to bleeding during bone decortications. The Fontan operation has become the accepted final stage of palliation for patients with single ventricles and establishes in series circulation between the systemic and pulmonary vascular beds (see Figure 3). However, the Fontan physiology comes at a considerable cost. In order to maintain adequate single ventricle preload in the 5–8 mmHg range CVP is chronically elevated and typically in the 12–15 mmHg range or even higher as in our patient, reflecting lack of a pulmonary ventricle and subtle elevations in pulmonary vascular resistance because of lack of pulsatility. Despite the elevated CVP the mean pulmonary artery pressures are typically lower than in the two-ventricle situation (known as Fontan paradox), because of the absence of a ventricle providing pulsatile flow in the pulmonary circulation. The flow through the central veins and Fontan circuit can be sluggish and places the patient at increased risk of thromboembolism (7). Impaired liver and bowel function as a result of elevated lower body venous pressures in patients with Fontan physiology often contribute to abnormalities in the coagulation profile including decreased levels of factor 7 (8–11).

Multiple factors related to the limitations of Fontan physiology likely contributed to the excessive blood loss in our patient although markedly elevated venous pressures in the prone position aggravated by positive pressure ventilation were likely the main cause in our patient (see below). In addition, the preoperative coagulation tests showed a repeatedly abnormal PT >15 s (normal range: 11.0–13.3 s) reflecting an INR of >1.6, which corrected with a 1:1 dilution with normal plasma. This translates into a significant decrease in multiple hepatic coagulation factors of <50% of normal (12). Therefore a moderate coagulation factor deficiency was assumed and fresh frozen plasma to normalize this clotting factor deficiency was an essential part of our blood replacement strategy early on.

We frequently encounter mildly elevated PT in our Fontan patients. Many Fontan patients are chronically anticoagulated with coumadin to keep the INR at about 2 to minimize thrombotic risks. In patients on coumadin we routinely discontinue anticoagulation 5–7 days before elective surgery and use fresh frozen plasma as needed to control bleeding. Our cardiologists are reluctant to use vitamin K for coumadin reversal, because they are concerned that this may lead to a hypercoagulable state. However, our patient had not received coumadin for many years and his chronically elevated PT was thought to be secondary to the limitations of the Fontan circulation related to venous hypertension and stasis. Preoperative platelet counts were normal but the initial bleeding time of 11.5 min suggests platelet dysfunction that normalized with discontinuation of acetalsalicylic acid.

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The excessive intraoperative bleeding was probably related to the even more markedly elevated venous pressures after prone positioning. Low venous pressures are very helpful in controlling bleeding during scoliosis surgery but our attempts to let venous pressures drift below 20 mmHg in the prone position repeatedly resulted in arterial hypotension. We considered it unwise to use deliberate hypotension considering the limitations of single ventricle physiology and in view of the uncertain effectiveness of arterial hypotension on blood loss in scoliosis surgery. There was no evidence of ventricular dysfunction of the single ventricle on preoperative transthoracic echocardiography and the CVP trends proved to be a very reliable predictor of hypotension. We considered the use of transesophageal echocardiography (TEE), as was described in one of the published case reports. However, we decided against routine TEE because assessment of ventricular function in single ventricle patients cannot be easily quantified because of geometric limitations and Doppler indices such as the myocardial performance index are not adequately validated in single ventricle patients. TEE performed by an expert pediatric echocardiographer was available as backup option in case of hemodynamic instability, i.e. if unexplained hypotension or hemodynamically significant dysrhythmias had occurred.

Finally, the extent of the surgery (10 vertebral levels) and long duration of the operation (7 h and 44 min) also likely contributed to the magnitude of bleeding. As is common in these protracted operations bleeding is best controlled by wound closure and resumption of the supine position which decreases venous pressures in two ventricles as well as single ventricle physiology. The modest postoperative blood loss in our patient suggests that markedly elevated venous pressures in the prone position during positive pressure ventilation, which partially normalized in the supine position, especially after resumption of spontaneous respiration, were indeed a major cause for the increased intraoperative bleeding.

A number of interventions might have been beneficial to curb intraoperative bleeding. Both prophylactic perioperative ε-aminocaproic acid and aprotinin have been shown to decrease blood loss in similar high-risk procedures (3,5,13). Excessive intraoperative bleeding may also dramatically and promptly respond to single or repeated doses of activated factor VII, a coagulation factor known to be deficient in Fontan patients (9,14,15). The senior author (E.S.) is a pediatric cardiac anesthesiologist with over 10 years experience in the routine use of prophylactic aprotinin and over 2 years experience with the emergency use of activated factor VII in congenital heart surgery. However, because of concerns about the thrombotic risks of these therapies in a circulation already prone to venous stasis and thrombosis we opted against the prophylactic use of these agents. The intraoperative course in our patient suggests that the benefit of these agents may outweigh any thrombotic risk, although the amount of postoperative bleeding after wound closure, repositioning and resumption of spontaneous respiration was very limited. Also because of concerns about thromboembolism acetylsalicylic acid was restarted after removal of the wound drains before hospital discharge.

A review of the anesthetic and the surgical literature lists only two other anesthetic case reports of scoliosis surgery in patients with Fontan physiology(16,17). A 15 year and a 14-year-old boy also had single left ventricle equivalents. In both cases CVP was elevated in the prone position but to a lesser extent than in our patient (at about 20 mmHg) and had to be kept elevated to provide stable hemodynamics. Surprisingly no excessive blood loss was reported in either case.

In summary, we describe the third case in the world literature of an adolescent with Fontan physiology who underwent scoliosis correction. Our patient experienced blood loss in excess of one blood volume that was likely caused by a combination of markedly elevated CVP during prone positioning and moderate derangements in coagulation related to Fontan physiology. With increasing use of Fontan palliation major non-cardiac surgery in this population will become more frequent and the anesthesiologist needs to be proactive. We suggest that prophylactic use of aprotinin or ε-aminocaproic acid may be routinely warranted in this population and that coagulation abnormalities should be aggressively corrected early on with appropriate blood products. If intraoperative blood loss continues to be excessive, a bolus of activated factor VII should be considered. Positioning should be optimized and the Jackson
table seems most suitable for Fontan patients as it keeps the abdomen free allowing unimpaired diaphragmatic excursion but CVP will likely be quite high despite optimal positioning efforts in Fontan patients until spontaneous respiration resumes. If at all possible the extent and the duration of the surgery should be limited as much as possible.

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