

Fontan's circulation with dextrocardia, recent pulmonary embolism, and inferior vena cava filter: Anesthetic challenges for urgent hysterectomy

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ABSTRACT

Fontan's circulation is a unique challenge for the anesthesiologist. Venous pressure is the only source of blood flow for the pulmonary circulation. Patients with such circulation are extremely sensitive to progression of cyanosis (decreased pulmonary blood flow) or circulatory failure. Any major venous compression can compromise the pulmonary blood flow worsening cyanosis; simultaneously, an increased afterload can precipitate circulatory failure. We present a rare patient of surgically corrected Ivemark syndrome with Fontan's physiology with dextrocardia who developed a large uterine fibroid compressing inferior vena cava (IVC). As a result of compression, not only the pulmonary circulation was compromised but she also developed stasis-induced venous thrombosis in the lower limbs that lead to pulmonary embolism (PE) (increased afterload). In addition to oral anticoagulation an IVC filter was inserted to prevent ongoing recurrent PE. Further, to prevent both circulatory compromise and deep venous thrombosis an urgent myomectomy/hysterectomy was planned. In the present case, we discuss the issues involved in the anesthetic management of such patients and highlight the lacunae in the present guidelines for managing perioperative anticoagulation these situations.

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INTRODUCTION

Fontan's repair since 1971 has been the primary surgical approach for complex congenital cardiac malformation not suitable for biventricular repair. Improved surgical techniques and experience had witnessed more than 90%, 10 years survival in these patients.^[1] As a result, many of these patients present for noncardiac surgery. They pose unique anesthetic challenges of altered physiology. The primary anesthetic aim in these patients is the maintenance of adequate venous pressure that propels the pulmonary flow. We present a rare patient with Fontan's circulation with large uterine fibroid compressing the inferior vena cava (IVC)

impending circulatory failure planned for hysterectomy at a young fertile age. In addition, we discuss the challenges related to management of anticoagulation in our patient

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who developed recent pulmonary embolism (PE) and was treated with IVC filter insertion.

A 32-year-old, married nulliparous female, post-Fontan's repair (15 years ago) presented to the hospital with increasing dyspnea. In the past Fontan's procedure had been performed 15 years ago because of congenital cyanotic heart disease "total anomalous pulmonary venous return with large unrestrictive ventricular septal defect (VSD) consistent" when she was also diagnosed to have dextrocardia with asplenia and diagnosis was consistent with Ivemark Syndrome.^[2] On investigation, she was found to have PE (diagnosed on computed tomography [CT] angiography) originating from deep vein thrombosis (DVT) of both the lower limbs. On further evaluation, she was found to have a massive uterine fibroid compressing upon the IVC that was not only leading to venous stasis but was also leading to impeding the failure of the Fontan's circulation. Eventually, with a possibility of DVT leading to further worsening of PE an IVC filter was placed on urgent basis and oral anticoagulation (warfarin) targeting international normalized ratio (INR) of 2–3 was also initiated. At the time of presentation, her effort tolerance was limited to 1–2 metabolic equivalent of tasks (METs), which subsequently improved to become >4 METs over next 10 days after IVC filter insertion. She did not have any features suggestive of failure and had maintained stable blood pressure despite increased ventricular afterload because of PE.

In view of huge size of the uterine fibroid, hysterectomy was planned in consent with the patient after discussing various possible options with the gynecologist. As the massive fibroid was compressing the IVC and to prevent failing of Fontan's circulation an urgent open hysterectomy was planned.

The anesthesia team examined her 3 days prior to the planned surgery for need of any further optimization. She was weakly built weighing 45 kg with a body mass index of 17.30 kg/m². After IVC filter, insertion dyspnea had disappeared and other than an ejection murmur chest examination was unremarkable. Both lower limbs showed signs of slight edema with persisting calf tenderness. Her room air pulse oximeter saturation was 86%. A room air arterial blood gas sample showed a partial pressure of oxygen and partial pressure of carbon-di-oxide of 56 and 32 mm of hg respectively with all other values being within normal range. Chest X-ray showed dextrocardia without obvious cardiac enlargement [Figure 1]. Echocardiography showed

dextrocardia, right isomerism, sub-aortic large VSD and an ejection fraction of 55% Biochemical investigations were within normal range, with hemoglobin of 14.6 m. She was on warfarin that had been stopped since last 5 days with present INR being 1.3 and she was receiving bridging anticoagulation with low molecular weight heparin enoxaparin, 40 mg subcutaneous twice daily. She was also receiving oral digoxin (0.125 mg once daily) and spironolactone (25 mg once daily), which were continued in perioperative period.

Preoperatively, she was prescribed anti-aspiration prophylaxis. However, sedation was avoided to prevent any possibility of CO₂ retention due to hypoventilation. A 16G intravenous (IV) line was started and maintenance fluid during the fasting period was advised. We advised to skip a preoperative night and following morning dose of enoxaparin.

In the operating room, standard ASA recommended monitoring was connected that included continuous 5 lead electrocardiogram (ECG), pulse oximetry, blood pressure monitoring. The ECG showed an inverted QRS complex due to dextrocardia. An arterial line for monitoring invasive blood pressure was placed in the right radial artery prior to induction using local anesthesia. After preoxygenation with high flow 100% oxygen, anesthesia was induced with 3 µg/kg fentanyl, titrated dose of etomidate and 4 mg of vecuronium. During mask ventilation, high positive pressure was avoided. Post-induction a 20G peripherally inserted central catheter (PICC) was placed in left cubital fossa vein allowing us to measure pulmonary artery pressures. Inotropic drugs such as milrinone and dobutamine (additional pulmonary vasodilation action) and vasopressors such as mephentramine and ephedrine (with preferential vasoconstrictive action) were kept at hand.

Volume control ventilation maintaining peak pressure <20 cm of water without any positive end-expiratory pressure was used. Any further need to increase the ventilation was planned to be managed by increasing the ventilatory frequency rather than tidal volume. Marginally low EtCO₂ (30–35 mm of Hg) was maintained using a tidal volume of 300 ml (roughly 7 ml/kg) with a rate of 20/min in oxygen air mixture (FiO₂ = 80%) to avoid hypoxic pulmonary vasoconstriction. Pulmonary artery pressure (via PICC) was continuously measured and was targeted to remain higher than peak airway pressures throughout the procedure. With this ventilatory strategy we were able to maintain a SpO₂ of 91–96% (PO₂

measured intraoperatively being 64) throughout the procedure [Figure 2]. In addition to 1 g paracetamol, fentanyl infusion at 20 µg/h was started after an initial bolus of 40 µg for intraoperative analgesia. As pulmonary artery pressure in these patients is directly dependent on the venous pressure, any possible hypovolemia was avoided with vigilant fluid administration chasing blood loss. Simultaneously, hypervolemia leading to circulatory failure was also kept in mind, and calculated amount of fluids were administered. Unfortunately, no noninvasive method of estimating fluid loss (cardiac output estimation) is presently validated for single ventricle physiology, so clinical judgment has to be relied upon. Intraoperative blood loss was estimated to be around 500 ml (200 ml blood loss + 300 ml in the removed uterus with huge fibroid) [Figure 3], which was supplemented with 500 ml of colloid and 1000 ml of crystalloid. At the end of surgery neuromuscular block was reversed and trachea was extubated. Patient was shifted to Intensive Care Unit (ICU) where, 20–30 µg/h fentanyl infusion was used for postoperative analgesia. Patient was able to maintain a saturation of 92–94% on facemask with an oxygen flow of 6 L (PO₂ being 58 mm

of Hg and PCO₂ of 36 mm of Hg) and was eventually discharged from the ICU next morning.

DISCUSSION

Anesthetic management of patients with Fontan's procedure is a challenging experience. Our patient was further complicated by additional dextrocardia, recent DVT followed by PE, IVC filter placement and systemic anticoagulation. The present case is extremely unique as it poses many ethical, technical and management based questions - some of which still remain unanswered due to gray areas or the limitation in the available literature. Despite extensive literature and guidelines on perioperative management of patients on anticoagulation the present practical situation remains almost untouched. Although IVC filters are now days only indicated where contraindication to anticoagulants exist, in our case, it was added to the anticoagulation to prevent PE and heart failure. IVC filter would immediately prevent any further pulmonary flow compromise by preventing ongoing PE.

Classically anesthetic management of Fontan's physiology revolves around maintaining the Central venous pressure (CVP) dependent pulmonary blood flow.^[3] It is, therefore, extremely important to maintain optimal intravascular volume in these patients. Thus in the present case, we advised the patient to take plenty of salt-rich fluids and allowed water orally until 2 h prior to shifting for surgery in addition to overnight maintenance fluid during the fasting period. Hypercarbia can further decrease pulmonary blood flow by increase in pulmonary vascular resistance, so we avoided preoperative sedation. We preferred general anesthesia to a sub-arachnoid block as sudden loss of sympathetic tone, increased venous capacitance and bradycardia are tolerated poorly compromising the pulmonary blood flow.^[4] Case reports have described

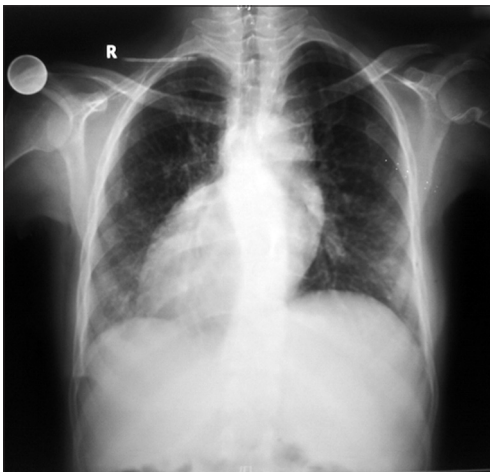


Figure 1: Chest roentogram showing – dextrocardia



Figure 2: Intraoperative patient monitor critical to keep airway pressures (left) lower than the Pulmonary artery pressure represented as central venous pressure (right)



Figure 3: Uterus showing massive fibroid (Huge size of fibroid in relation to the uterus lead to hysterectomy rather than fibroid removal)

the successful use of epidural analgesia in these patients^[4,5] we avoided the same due to issues related to anticoagulation in our patient. A low transverse incision was planned for hysterectomy so IV opioids and nonsteroidal anti-inflammatory drugs sufficed for postoperative analgesia allowing us to avoid epidural analgesia. For induction of anesthesia we used etomidate, which is known to have least vasodilator and cardiac depressant effect. During induction and maintenance of anesthesia we used fentanyl adequately as that helped us to reduce the requirement of inhalation agent (sevoflurane); all of which are known to be vasodilators that eventually increase venous capacitance and lower venous pressure.

CVP in Fontan's, unlike in most patients reflects the pulmonary artery pressure as superior vena cava is in direct communication with the pulmonary artery and provides the driving pressure for the pulmonary circulation. The most literature recommends avoiding or using central venous catheters for an extremely short time in these patients due to associated potential complications like thrombosis of the Fontan pathway, paradoxical emboli, and infection. Under spontaneous ventilation, the venous pressure remains higher than alveolar pressure but under positive pressure

ventilation, measuring pulmonary artery pressure can guide to maintain safe airway pressure without compromising pulmonary flow.^[6] In anticipation of need for vasopressors or inotropes, PICC is also preferable to a peripheral IV cannula. Hence, we used a PICC line for the intraoperative period and removed it within 2 h of surgery in the ICU. Fontan's patients have higher propensity to show venous ooze in perioperative period due to elevated systemic venous pressure, presences of collateral vessels, coagulation factor abnormalities, and antithrombotic therapy, thus we made sure that adequate amount of blood/products were available in the operating room during the procedure. Milrinone infusion was prepared because of its lusitropic and pulmonary vasodilatory properties, it is well-suited for the Fontan physiology.^[7]

Unique, unanswered question for the management of our patient was the management of anticoagulation. Classically IVC filters are inserted in patients for PE prophylaxis where contraindications to anticoagulations exist.^[8] Our patient had recurrent PE (diagnosed using CT angiography, on being symptomatic) that not only worsened chronic hypoxia but also could cause ventricular failure.^[9] Since she had no contraindication to anticoagulation, she was started on warfarin targeting an INR of 2–3. With the history of DVT being only 2 weeks, she was extremely high risk for stopping anticoagulation.^[10] In addition, a high hematocrit and thrombogenic IVC filter further added to risks of increasing DVT and its complications.^[11] Present guidelines do not suggest any management directives for patients with recent DVT with IVC filters,^[12] although the available literature does classify them into very high-risk group. Prophylactic use of mechanical DVT prophylaxis in patients with recent history of DVT is also questionable. Keeping the above conditions in mind and in view of low risk of bleeding in the present surgery we started enoxaparin on the evening of the day of surgery after ensuring minimal drain output.

Thus we conclude that if prime anesthetic goals of low ventilatory pulmonary airway pressure and adequate volume preload are met, general anesthesia can be safely used for high-risk patients with Fontan's circulation.

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Conflicts of interest

There are no conflicts of interest.

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