Case Report

# Anaesthesia for a minor procedure in a patient with fontan physiology

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## ABSTRACT

Fontan procedure is a palliative surgery done for patients born with single ventricle physiology. An understanding of the hemodynamic alterations in such a patient is important for successful perioperative management. We have discussed the anaesthetic considerations in a 12 year-old girl with complex congenital heart disease ultimately palliated by a Fontan operation, who was posted for Botox injections for upper limb spasticity under general anaesthesia.

**Key words:** Congenital heart disease, fontan, general anesthesia

## INTRODUCTION

In 1971, Fontan and Baudet first described a palliative surgery for patients with tricuspid atresia.[1] It revolutionised the management of patients with complex congenital heart disease characterized by a single functional ventricle. We describe the anaesthetic management of a 12 year-old girl who had Fontan physiology and right hemiparesis, for botox injections, managed under general anaesthesia.

## CASE REPORT

A 12 year-old girl weighing 41kg was posted for Botox (botulinum) injections and casting for spasticity of the right upper limb. She gave history of having some heart disease since birth for which multiple operations had been done. On going through her past medical records, it was found that she was born with complex congenital heart disease, having dextro-transposition of great arteries,double outlet right ventricle,pulmonary stenosis, and a large ventricular septal defect. She underwent a right subclavian to right pulmonary artery shunt shortly after birth, a Glenn operation at 4 months of age, and an extracardiac Fontan operation (total cavopulmonary conduit) at 5 years of age. She was on chronic therapy with warfarin. At age 10, she was hospitalized with a cerebrovascular accident for a month. Computed tomography (CT) brain showed left middle cerebral artery (MCA) thrombosis due to which she had developed right hemiparesis. The hemiparesis improved gradually and she developed spasticity of the right upper and lower limbs. She had developed pedal edema a few months earlier, due to an element of protein losing enteropathy, which was treated with diuretics and high protein diet. She gave history of no cardiovascular (CVS) complaints at the time of admission, and was regular in her studies and daily activities. She was on treatment with tablet warfarin 5mg and tablet furosemide 20mg once a day.

On examination, she was conscious, oriented, well nourished. She was afebrile, pulse was 82 beats/minute and regular, blood pressure was 102/66mm Hg. A median sternotomy scar was present. CVS examination showed a regular heart rate, and a grade 2 systolic murmur. She had spasticity of the right upper limb more than the lower limb. Rest

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of the systemic examination was unremarkable. Investigations revealed haemoglobin of 11.2g/dL, total and differential white blood cell counts were normal. Renal function tests, serum electrolytes, liver function tests including serum proteins were within normal limits. Prothrombin time was 22s, with an INR of 2.1. Electrocardiogram (ECG) showed sinus rhythm. Echo showed patent functioning cavopulmonary shunts, mild ventricular systolic dysfunction, no thrombi/ vegetations.

A cardiology opinion was taken. She was advised to withhold warfarin and switch over to a low-molecular weight (LMW) heparin 5 days prior to surgery. She was started on enoxaparin 40 mg subcutaneous twice a day. Infective endocarditis prophylaxis was also advised. Enoxaparin was withheld 12 hours prior to surgery.

On the night prior to surgery, patient was given tablet alprazolam 0.25mg. Infective endocarditis prophylaxis was given with IV ceftriaxone 1g 1 hour prior to the procedure. It was decided to give general anaesthesia since the patient was very apprehensive and insisted on it. Patient was taken to the operation theatre, monitors were attached (ECG, pulse oximeter, capnograph, non-invasive blood pressure monitor). An infusion of lactated Ringer's solution was started and she was preloaded with around 200ml. Patient was premedicated with IV ondansetron 4mg. Preoxygenation was done for 5 minutes and anaesthesia was induced with IV fentanyl 80 mcg and propofol 80mg IV slowly and maintained with sevoflurane 0.6-0.8% in 50% nitrous oxide in oxygen, on spontaneous ventilation with intermittent assist. Blood pressure was maintained within 20% of the baseline value. Fluids were given to maintain blood pressure. End-tidal CO2 was maintained between 30 and 35mm Hg. Patient received 300ml Ringer's lactate intraoperatively, and the procedure lasted 30minutes. At the end of surgery, a 50mg diclofenac per rectal suppository was given for post op analgesia. Once awake, responding to oral commands and stable, patient was shifted to the recovery room for monitoring. Patient was comfortable, and her vitals were stable. She was shifted to the ward after an hour and had an uneventful post operative period. She was discharged the next day.

## DISCUSSION

Prior to the development of the Fontan procedure, pulmonary blood flow in patients with single ventricle and pulmonary stenosis was surgically augmented by means of systemic to pulmonary artery shunts. These shunts improved life expectancy remarkably in the short term, but survival past the second decade remained unusual.[2] In 1971, Fontan and Baudet,[1] and in 1973, Kreutzer *et al.*[3] independently described a right atrial to pulmonary artery shunt procedure for tricuspid atresia. It involved diverting systemic venous blood from the right atrium to the pulmonary arteries, thus bypassing the right ventricle. It was then used for treating a number of complex congenital heart lesions with a single effective ventricle.

In Fontan physiology, systemic venous blood from the great veins passively enters the pulmonary artery. Oxygenated blood then drains into the left atrium and then into the single ventricle that empties into the systemic circulation. The difference between central venous pressure and systemic ventricular end-diastolic pressure (termed the "transpulmonary gradient") is the primary force promoting pulmonary blood flow and, more importantly, cardiac output.[4] Since intravascular volume is the main determinant of central venous pressure, hypovolemia is poorly tolerated.

Thus, the main determinants of the Fontan circulation are systemic venous pressure and volume, pulmonary vascular resistance, cardiac rhythm and left ventricular function. A disturbance in any of these compromises the cardiac output.[4]

Complications in post-Fontan surgery patients include arrythmias,[5] thromboembolism,[6] protein losing enteropathy,[7] and ventricular dysfunction.[8]

Preoperatively, the functional capacity of the patient must be assessed. Relevant biochemical investigations should be carried out, including the coagulation profile. ECG and echocardiogram will give valuable information on the patient's cardiac status. Infective endocarditis prophylaxis should be considered.

Invasive monitoring has to be considered for major surgeries. For induction, one should avoid drugs that depress myocardial contractility like thiopentone. Propofol, with its transient systemic vasodilatation, is usually less problematic, as long as normovolemia is maintained.Etomidate, with its cardiostable property would be the best drug for induction is these patients.

High concentration of volatile agents should be avoided since they cause myocardial depression. Hypercarbia, hypoxia, inadequate analgesia, and acidosis should be avoided as they will lead to an increase in pulmonary vascular resistance, decreased pulmonary blood flow and thus decreased cardiac output. For short procedures, spontaneous ventilation is better, as long as hypercarbia is avoided.[9] Controlled mechanical ventilation leads to increase in intrathoracic pressure which decreases venous return, in turn causing decreases pulmonary blood flow, and hence, decreases cardiac output.

Regional anaesthesia can also be employed depending on the surgery. Epidural anaesthesia has been successfully employed in such patients.[10-12]

Postoperatively, good analgesia has to be ensured. For more painful surgeries, continuous catheter techniques, epidural analgesia[10,11] and patient controlled analgesia[12] are options provided that any coagulopathies are taken into account. Continuous monitoring, including oxygen saturation is a must.

## CONCLUSION

Fontan patients have a unique physiology which needs to be addressed during anaesthesia. Normovolemia needs to be maintained, and hypercarbia, hypoxia and acidosis should be avoided. Minor procedures can be safely performed on a day care basis.

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