Case Report

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Anaesthetic Management of a Patient with Failing Modified Fontan, Morbid Obesity, Atrial Flutter and OSA posted for RFA-A Case Report

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Abstract

The Classic Fontan has been abandoned these days due to occurrence of rapid and incessant complications. Presence of complications like, morbid obesity, obstructive apnoea, chronic atrial flutter, elevated Fontan pressures as seen in our patient, increases the risk of Anaesthesia manifold times. We report a case of 32 year old male patient with failing modified Fontan and aforementioned complications, posted for radiofrequency ablation under general Anaesthesia.

Key words: Failing Fontan Circulation; Remifenanil; Morbid obesity.

Key Messages: Anaesthetic management of patients with failing modified Fontan and its associated complications needs long standing experience , thorough understanding and proper selection of anaesthetic drugs to conduct a case safely without causing fatal acute decompensation.

Introduction

The Fontan procedure, first done in 1968, is a well-established surgical treatment for single ventricle congenital cardiac defects.[1] Several modifications have been made over the past decades, in particular, the atrial- pulmonary anastomosis has been replaced with a total cava-pulmonary artery anastomosis.[2] This has significantly improved prognosis and patient survival, with a lower occurrence of arrhythmias, delaying the onset of cardiac failure,as compared to patients with "classic Fontan"(atriopulmonary connections).[3-4]

Classic Fontan is totally abandoned these days hence its rare to encounter adult patients with this type of repair. Fontan circulation and its failing component implies highly complex physiologic and multiorgan considerations requiring a meticulous planning and execution of Anaesthesia in order to maintain homeostasis and avoid fatal complications and acute decompensation. Factors like morbid obesity, OSA, loss of A-V synchrony, rising pressure in Fontan Circuit can pose significant threat to life. Therefore managing this case perioperatively is a unique and rare challenge an anaesthesiologist would come around.

We present here Anaesthetic management of a case of failing Classic Fontan complicated with morbid obesity, chronic atrial flutter, Diabetes Mellitus type 2 and Obstructive sleep apnoea syndrome, posted for Radiofrequency ablation.

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Case History

A 32 year old male patient, weighing 132.6 kgs, BMI of 45, s/p Modified Fontan operation, had unbalanced AV canal defect, hypoplastic(rudimentary) right ventricle, severe Pulmonary stenosis. He had undergone staged surgeries on a single ventricle pathway and had a long history of atrial flutter with multiple interventions including DC cardioversions and RFA. Now presented with recurrentpalpitations, severe breathlessness NYHA-3, MET less than 4, orthopnoea, desaturations to mid 80s and persistent cyanosis. His latest catheter data correlated with failing Fontan, pressure in Fontan circuit 20mm Hg, high TPG, moderate ventricular dysfunction, moderate AV valve regurgitation, massively dilated IVC, RSVC and Coronary sinus draining into hugely dilated right atrium draining into Main pulmonary artery. Dilated Fontan circuit had stasis and energy losses. Had history of acute decompensation after RFA in the past and now was on sotalol, warfarin and lisinopril.Lab data wise had polycythemia and elevated hepatic enzymes.

He had severe anxiety disorder particularly needle phobia and refused for sedation in spite of counselling for benefits of maintaining spontaneous breathing, hence, we planned GA for him. We anticipated difficult intubation and mask ventilation.

On the day of procedure, no oral premedication was given in view of his sleep apnoea. Monitors were connected and 18G IV cannula secured. Preinduction fluid 500mls Hartmann solution given. Anaesthetic induction done with Remifentanil infusion on TCI with effect site target of 2ng/ml initially later increased to 4ng/ml for intubation, supplemented with Inj Midazolam 2mg, Inj Propofol 100mg and Inj Atracurium 100mg. Intubation was done in single attempt with McCoy blade followed by arterial and central line. Hehad mild hypotension and bradycardia, BP was in low 80s and heart rate low 50s after induction drugs, which was countered with another 500mls of volume, Inj Phenylephrine boluses and Inj Glycopyrollate. Thereafter his hemodynamics stabilized to acceptable values. He was Ventilated on volume control, peak airwaypressures limited to 20, PEEP of 6, RR to maintain $ETCO_2$ less than 40mmHg. We could not monitor his SVV, PPV or cardiac output due to his atrial flutter rhythm.

Maintenance of Anaesthesia was done with Remifentanil @ 2ng/ml on TCI and Desflurane with a MAC of 0.7 to 0.8, no further boluses of muscle relaxant were given. BIS was maintained between 40-50.. His Fontan pressure was 20 to begin with and later increased to 24. Started on Phenylephrine infusion in low dose to support his pressure which maintained a pressure above 90 systolic. ABG showed normal values. He received a total of 2.5 litres of volume throughout the procedure. Urine output improved to 1ml/kg after first hour and later was increased to 1.5ml/kg.

Patient underwent procedure uneventfully which lasted for 4 hours. On conclusion his Anaesthetics were stopped, he was reversed and extubated in sitting position after he was fully awake. He was shifted to ward after 30mins. Post operative course was uneventful.

Discussion

The hallmark of the Fontan circulation is a sustained, abnormally elevated central venous pressure acting as driving force for pulmonary circulation in the absence of subpulmonary ventricle, combined with decreased cardiac output, especially during periods of increased demands, resulting in a cascade of physiologicalconsequences(5-6). Any alterations caused during Anaesthetic management can trip off the balance of such precariously maintained circulation.

In our patient the problem was compounded by extremely high pressures in Fontan circuit(sign of failing Fontan), morbid obesity, OSA and loss of AV Sequential synchrony, specially in a very high risk substrate of "classic Fontan" circuit.(9, 10,11,12) By contributing to alterations in pulmonary functionand increased systemic vascular resistance, overweight contributes rapidly to Fontan failureand physiological changes associated with OSA, hypercarbia, hypoxia, and hypoxic pulmonary vasoconstriction (HPV) worsens the PVR.[7]

Other Clinical hazards faced by these patients include progressive fatigue, heart failure, arrhythmias, and end-organ complications such as liver disease, plastic bronchitis, protein loosing enteropathy, desaturations, thromboembolic complications, multiple surgical and non surgical interventions, in addition to anxiety and concern about their condition and future.[13,14] Our patient had almost all the problems except plastic bronchitis and PLE.

Induction of general endotracheal anesthesia with administration of cardiac depressant medications and conversion to positive pressure ventilation often results in decreased contractility and decreased pulmonary blood flow secondary to increased mean airway pressure leading to acute decompensation.

Choosing Remifentanil and Desflurane kept his hemodynamics stable and led to full and quick recovery from GA.[8] Choice of drugs was of utmost importance to maintain his physiology.

Conclusion

Patients with Fontan,are in a state of chronic low cardiac output and elevated systemic venous pressure. Risks and challenges of anaesthesia are increased due to presence of various complications specially in a classic modified Fontan.

Therefore we believe that complete understanding of Fontan circuit, factors affecting flow through this circuit, effect of long term complications, current catheter data and most important is the experience to deal with this highly complex substrate of patients is the key to managing these patients successfully.

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