[NM-290] Anesthetic Management of a Patient with Fontan Physiology Being Evaluated for Liver Transplant

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AE is a 6-year-old female with single ventricle, S/P Fontan procedure and agenesis of portal vein (Abernathy malformation) resulting in hepatopulmonary syndrome. She is being considered for liver transplant and presented for MRA abdomen.

Patient was born with complex congenital heart disease- double outlet right ventricle, D-malposed great arteries, hypoplastic pulmonary arteries and interrupted IVC. She had BT shunt placement shortly after birth followed by Glen/Kawashima procedure with BT shunt take down at 6 months of age. Subsequently she had Fontan completion when she was two years and four months of age. Preoperative evaluation was significant for persistent desaturation, room air SpO2 being 83-89% in sitting position. Her weight was 19.5 kg and there was cyanosis of the lips. For medications she was taking enalapril, lasix, spironolactone and aspirin. Echocardiography was showing unobstructed right hepatic vein pathway through intracardiac Fontan. Extracardiac Fontan was seen unobstructed. ECG was ectopic atrial rhythm. Recent cardiac catheterization confirmed pulmonary arterio-venous malformations. There was an area of mild narrowing of the intracardiac lateral tunnel that was balloon dilated and stented.

For MRA abdomen, MRI compatible noninvasive monitoring including ECG, pulse oximeter, blood pressure and ETCO2 were applied. Anesthesia was induced with sevoflurane and nitrous oxide. LMA was placed and anesthesia was maintained with propofol infusion. Patient was breathing spontaneously during the procedure at FiO2 0.4. SpO2 and hemodynamics remained stable during the procedure. PACU course was uneventful and patient was discharged home the same day. Currently the patient is awaiting liver transplant and is also being considered for a possible combined heart and liver transplant.

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. [1] Thus, the main determinants of the Fontan circulation are systemic venous pressure and volume, pulmonary vascular resistance, cardiac rhythm and left ventricular function. Hence, during perioperative period we ensured maintenance of adequate intravascular volume, minimized high dose volatile agents which could depress myocardial contractility and avoided hypoxia and hypercarbia. For relatively short procedures, as in this situation, Fontan patients are probably better off breathing spontaneously, as long as severe hypercarbia is avoided. [2] Potential disadvantages of mechanical ventilation relate to subsequent decrease in venous return and cardiac output.

References

1. McClain CD, McGowan FX, Kovatsis PG. Laparoscopic surgery in a patient with fontan physiology. Anesth Analg. 2006; 103(4): 856–8.
2. Nayak S, Booker PD. The Fontan circulation. Contin Educ Anaesth Crit Care Pain. 2008; 8(1): 26–30.
