Anesthetic Management of Biventricular Device Explantation in a Child with LCHAD

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Introduction

Long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency is a very rare condition that prevents the body from converting certain fats to energy, particularly during periods of fasting.

The incidence of LCHAD deficiency is unknown.

Our literature review revealed minimal reports of anesthetic management for BiVAD explantation in the pediatric population, specifically pediatric patients with LCHAD deficiency.

For more details about Triheptanoin (C7) trial, please visit: https://clinicaltrials.gov/ct2/show/NCT01886378

Case Presentation

• An eight year old male child weighing 25.5 kg with LCHAD deficiency developed a metabolic crisis in the setting of a viral gastrointestinal illness. He progressively developed cardiomyopathy requiring extracorporeal membrane oxygenation (ECMO) support and eventual conversion to biventricular assist device (BiVAD) support.

• After resolution of his viral gastroenteritis he was placed on the appropriate nutrition required for LCHAD management as well as triheptanoin and levocarnitine. After three weeks his cardiac function recovered as shown by serial echocardiograms to the point where explantation of the BiVAD was discussed as opposed to heart transplantation.

• He was brought to the cardiac catheterization lab where his data showed favorable end-diastolic pressures in both ventricles with normal pulmonary and systemic pressures. He tolerated the BiVAD clamp trial well with minimal pressor support. Therefore, based on these findings, he was subsequently taken to the operating room for explantation of the BiVAD.

• The patient arrived to the operating room with a preexisting cuffed endotracheal tube, left radial arterial line and right upper arm peripherally inserted central catheter. Anesthesia was induced with intravenous midazolam, fentanyl and rocuronium. A central line was placed in the left internal jugular vein. Anesthesia was maintained with a mixture of oxygen and air, isoflurane and infusions of dexmedetomidine and sufentanil.

• In order to maintain adequate nutrition for his metabolic requirements as per recommendation from the genetics and endocrine services, we were required to administer a dextrose 25% in water infusion, which also required an insulin infusion to maintain control of his blood glucose levels. (See figure 1)

• He was started on epinephrine and milrinone infusions to prepare for explantation of the BiVAD. In addition, he also underwent primary repair of a left ventricular aneurysm and patch repair of his right atrium and pulmonary artery. He tolerated the procedure well with good biventricular function postoperatively.

Discussion

This case presented challenges from a cardiorespiratory and hematologic perspective for the explantation of the BiVAD which was made more complex from the metabolic requirements to prevent a crisis in light of his diagnosis of LCHAD.

This case reflects the importance of perioperative communication between the different medical services involved in the patient care (anesthesiology, cardiothoracic surgery, cardiology, endocrinology, genetics and intensive care medicine) for patients with complex medical issues undergoing major surgery.

References


Figure 1. Intraoperative glucose levels


Figure 1.

Intraoperative glucose levels

Glucose, Whole Blood