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## Background

Glutaric Aciduria type II(GAII) is an autosomal recessive metabolic disorder that usually presents as acidosis, hypoglycemia, heart failure, and a characteristic "sweaty feet" odor. On the molecular level, these patients have mutations in electron transport transfer flavoprotein(1). These mutations disable the electron transport chain and metabolism of proteins and fats. Proteins and fats are metabolized incompletely and it can lead to profound non-anion gap metabolic acidosis. Diagnosis of GAII is by examining urine for organic acids. Treatment of the disorder includes avoidance of fasting, protein restriction, and vitamin supplementation. The goal is to replenish CoA supply, prevent neurological consequences, and detoxification of organic acids. Even minor illness requires aggressive maintenance of caloric intake either orally or via central line(2).

## Patient Overview

Our patient is a 11 year old female born full term. At 3 weeks of age, she had FTT and was admitted to the NICU for 3 weeks. After this episode, genetic testing was done that suggested glutaric aciduria. She has developmental delay and did not walk until age 6. She is easily fatigued and hypotonic.

## Case Report

She was seen by cardiology with an echo remarkable for an ASD that closed without intervention and no cardiomyopathy. Due to her GAII, she was admitted the night before surgery for D10 IV fluid administration per recommendation of her metabolic specialist while she was NPO. She was taken to the OR for a posterior spinal fusion of T6-L4 for treatment of scoliosis. Total intravenous anesthesia of Precedex, Ketamine, Propofol, and Sufenta were infused. Transcranial motor evoked potential monitoring was done throughout the case by a technician. D10 W at 100ml/hour was continued throughout case. Blood glucose was monitored hourly and with intermittent arterial blood gases. The patient slowly developed a metabolic acidosis and the metabolic specialist was called to coordinate care for and transfer to outside facility. She was transferred to the ICU for further management. She was extubated postop day one. A multidisciplinary team was utilized to ensure adequate postop care with concomitant GAII.

## Considerations

GA II has a varied presentation so the anesthesiologist must be aware of the stimulus and symptoms from the last GA II flare. Perioperative D10 infusions assist to maintain glucose levels and delay acidosis. Use short acting medications that encourage fast return to oral feeds. It is essential to have a metabolic specialist available for recommendations, consultation, and postop care. These patients require close intraop monitoring of fluid status, base deficit, glucose. The level of preop and postop care may change depending on risk or type of surgery. These patients can have neurological impairment so note preoperative neurological exam. In conclusion, a multidisciplinary approach is essential for positive outcome for GA II patients undergoing surgery.

## References:

1. Gordon N. Glutaric aciduria types I and II. *Brain and Development*.28(2006)136-140.

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