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.

GAII has a variable presentation and age of onset. It is associated with birth defects including polycystic kidneys, developmental delay, cardiomyopathy, failure to thrive, and sudden death.

On the molecular level, these patients have mutations in electron transport flavoprotein and/or electron transfer flavoprotein dehydrogenase. These mutations disable the electron transport chain and metabolism of proteins and fats.

Diagnosis is by examining urine for the organic acids glutaric, ethylmalonic, adipic, and isovaleric acid.

Treatment of the disorder involves dietary manipulation with avoidance of fasting with or without protein restriction, riboflavin, and carnitine supplementation.

The goal is to replenish CoA supply, prevent neurological consequences, and amplification of physiological detoxification. Even minor illness requires aggressive maintenance of caloric intake either orally or via central line.

**CASE REPORT**

11 year old female born term and at 3 weeks of age had failure to thrive. She was admitted to the neonatal intensive care unit for 3 weeks. After this episode, genetic testing and workup that suggested glutaric aciduria type 2. She had developmental delay and started walking at age 6. She is easily fatigued and hypotonic. She was found to have neurogenic scoliosis and after orthopedic evaluation, she was scheduled for posterior spine fusion.

**PERIOPERATIVE MANAGEMENT**

**Preop**

She was cleared by cardiology, no reported cardiomyopathy. Due to her GAI, she was admitted the night before surgery for D10 IV fluid administration per recommendation of her metabolic specialist while she was NPO.

**Intraop**

She was taken to OR for a posterior spinal fusion of T6-L4 for treatment of scoliosis. The case started with a smooth IV induction and easy intubation. Standard ASA monitors, art line, PIV x2. Total intravenous anesthesia of Precedex, Ketamine, Propofol, Sufenta, and TKA were started transcranial motor evoked potential monitoring done by technician. D10 W at 100/hour was continued throughout case. She received a total of LR: 3000ml, PRBC: 2 units, Cell saver: 320ml 5% Albumin: 250ml Total Out: 1695ml - EBL: 1250ml UOP: 445ml. 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.

**Post-op**

She is transferred to the ICU at tertiary care center for further management. She was extubated postop day one. A multidisciplinary team was utilized to ensure adequate postop care with concomitant GAII.

**BIBLIOGRAPHY**


**CONCLUSIONS**

• Be aware of the stimulus and symptoms from the last GA II exacerbation. Preoperative and intraop D 10 infusions assist to maintain glucose levels and delay acidosis.

• These patients have variable presentation so note preoperative neurological, pulmonary, and cardiac exam.

• It’s essential to have metabolic team available for recommendations, consultant, and postop care.

• These patients require close intraop monitoring of fluid status, base deficit, glucose.

• A multidisciplinary approach is essential for positive outcome for GA II patients undergoing surgery.