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**Summary:** We present the case of an infant with citrullinemia scheduled for a broviac catheter insertion. We discuss the anesthetic implications and management of this rare inborn error of metabolism.

**Case Report:** A 14-day old full-term female was admitted with lethargy, minimal urine output and several episodes of vomiting. In the ED she was noted to be hypotonic with twitching of the upper extremity. A metabolic panel revealed an ammonia level of 427  $\mu\text{g/dl}$  (newborns: 90-150  $\mu\text{g/dl}$ ); EEG showed global cerebral dysfunction. Emergent hemodialysis was performed. Metabolic workup revealed elevated citrulline levels. A diagnosis of citrullinemia was made. The patient was started on ammonul, arginine, levocarnitine and phenobarbital. Ammonia levels subsequently decreased to the range of 32-64  $\mu\text{g/dl}$ .

A broviac catheter was planned for long-term venous access. Preoperative evaluation revealed a normal-appearing baby girl with mild hypotonia and no evidence of tremor or seizure activity. Ammonia was 30  $\mu\text{g/dl}$ ; other labs were unremarkable.

Standard ASA monitors were applied. Baseline vital signs were normal. After preoxygenation the patient received atropine, fentanyl, propofol and rocuronium to facilitate endotracheal intubation. A 3.5cm uncuffed ETT was placed and secured. Anesthesia was maintained with sevoflurane in 50% oxygen. A femoral broviac catheter was placed. Neuromuscular blockade was reversed and spontaneous respiration resumed with adequate tidal volumes and end tidal CO<sub>2</sub> of 46. The child was extubated and transported to PICU.

**Discussion:** Citrullinemia is a rare autosomal recessive genetic disorder with an incidence of ~1:250,000 people. Two forms of citrullinemia exist and are caused by mutations in different genes. Type 1 citrullinemia is caused by a defect in arginosuccinate synthetase located on chromosome 9 and is the most common form of the disease. Arginosuccinate synthetase is a urea cycle enzyme that catalyzes the synthesis of arginosuccinate from citrulline and aspartate. Type 1 citrullinemia inhibits the ability of the liver to convert ammonia to urea, causing ammonia and citrulline to accumulate. Patients become symptomatic 24-48 hours after birth and exhibit poor feeding, vomiting, lethargy, and hypotonia, which can progress to seizures and coma. Signs of increased intracranial pressure and cerebral edema are the direct result of hyperammonemia on the CNS.

Treatment of patients with citrullinemia includes a low protein diet to minimize production of ammonia and essential amino acid supplementation. Sodium benzoate and sodium phenylacetate can help remove ammonia from blood by providing an alternative pathway for nitrogen elimination. Dialysis may be used to remove ammonia from the blood when it reaches critical levels.

The major anesthetic goal in the management of patients with citrullinemia is to minimize the adverse effects that can result from worsening of hyperammonemia. Fasting, trauma, surgical stress, GI hemorrhage and drug interactions all have the potential to precipitate a hyperammonemia crisis perioperatively. Although mortality has decreased with these interventions, morbidity remains high in patients with this disease given the associated growth and mental retardation.

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