

## **PBLD—Table #7**

### **One last MRI in a recently adopted infant with hypotonia and seizures... Sweet! Or maybe not?**

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#### **Objectives:**

1. Discuss the differential diagnosis of hypotonia in infancy and the associated anesthetic implications
2. Discuss the differential diagnosis of hypoglycemia
3. Summarize the pathophysiology and management of congenital hyperinsulinism

#### **Case History:**

A 14-month-old girl recently adopted from India presented for magnetic resonance imaging (MRI) of the brain under anesthesia to evaluate seizures and hypotonia. Very little past medical history was known other than that the seizures had persisted for six months despite initiation of two antiepileptic medications (Phenobarbital and Phenytoin).

#### **Questions:**

What is the differential diagnosis of hypotonia in infancy? How do the anesthetic considerations differ for neuromuscular diseases versus mitochondrial disorders? How do you develop an anesthetic plan for a diagnostic procedure for a hypotonic child?

#### **Case History:**

Upon meeting the patient, she was remarkably lethargic. Her father reported that she had been seen in the emergency department (ED) four days prior for seizures and was incidentally noted to have a right-sided groin abscess that was drained during the visit. She was initially started on Clindamycin and was changed to Trimethoprim-Sulfamethoxazole after bacterial sensitivity was identified. Her father stated that her appetite had lessened and she had been more listless since their visit and wondered if this could be an effect of the antibiotic.

#### **Questions:**

Does the patient's lethargy concern you? What could be the cause?

### **Physical Examination and Vital Signs:**

Physical examination revealed a small, nondysmorphic, notably lethargic girl lying on a stretcher. Her airway was externally normal, heart rate and rhythm regular with no murmur, and lungs clear to auscultation bilaterally. The patient's weight was 8.5 kg, heart rate 130, respiratory rate 26, blood pressure 112/79, and oxygen saturation 93% on room air. She was afebrile.

### **Questions:**

How would you proceed? Would you request further evaluation? Would you anesthetize the child? How?

### **Case Progression:**

After discussion with the patient's father, the decision was made to proceed with a general anesthetic involving an inhalation induction, placement of an intravenous (IV) catheter, propofol infusion, and supplemental oxygen administration and end-tidal carbon dioxide monitoring with a nasal cannula. The plan included checking the patient's glucose, electrolytes, and hemoglobin with an i-STAT after induction to evaluate any treatable causes of her remarkable lethargy.

### **Intraoperative Care:**

The patient underwent a smooth inhalation induction and had an IV catheter placed easily. Blood was drawn before IV fluids were initiated and i-STAT test results were as follows:

- pH 7.35
- pCO<sub>2</sub> 49
- pO<sub>2</sub> 38
- HCO<sub>3</sub> 27
- Total CO<sub>2</sub> 28
- Base excess 1
- Ionized calcium 1.25
- Glucose <20
- Potassium 4.3
- Sodium 139
- Hemoglobin 14.6

### **Questions:**

How would you proceed with this information? What are possible causes of profound hypoglycemia in this patient? Could this glucose level be explained by fasting? Infection? Something else?

### **Intraoperative and Postoperative Care:**

The patient was given 1.5 ml/kg of D50 and an infusion of D5 was started. Her blood glucose was 351 mg/dl and the decision was made to proceed with the MRI. Blood glucose at the end of the MRI was 246 mg/dl. Upon discontinuation of the anesthetic, the patient awoke and was alert, interactive, and strong. She drank a bottle and her blood glucose was 136 mg/dl.

### **Questions:**

What do you think is the appropriate next step for this child? Would you contact the pediatrician? Discharge home? What would you tell her father?

### **Postoperative Course:**

After discussion with her father regarding her hypoglycemia, the patient was allowed to return home and her father was asked to make her pediatrician aware of this incident and promptly return to the hospital if she were to become lethargic again.

The next day, the child's father brought her back to the hospital as she was again lethargic. Blood glucose was measured and was 22 mg/dl. Critical hypoglycemia labs were drawn and showed an inappropriately high level of insulin as well as inappropriately low levels of ketones and free fatty acids, all of which are consistent with the rare disorder of congenital hyperinsulinism (CHI).

The patient was admitted and IV fluids were administered with a glucose infusion rate of 13.3 mg/kg/minute. She was subsequently started on diazoxide, found to be responsive, and ultimately weaned from IV glucose and discharged home. She has since undergone genetic testing but a definitive cause for her CHI has not been identified. She has been weaned from her antiepileptic medications and has had no further seizures.

### **Discussion:**

Infants with hypotonia of an unknown cause can present a dilemma when they require anesthesia for diagnostic procedures. The causes of hypotonia in infancy include neuromuscular diseases and mitochondrial disorders and these two classes of pathologies have different anesthetic implications. With some neuromuscular diseases, including Duchenne muscular dystrophy, intravenous techniques are recommended. On the other hand, intravenous techniques involving propofol can cause metabolic decompensation in patients with mitochondrial disorders. In patients presenting with hypotonia of unknown etiology, anesthetic technique is based on the patient's presumptive diagnosis given the presenting signs.

In patients who are fasting, physiologically normal glucose levels range from 70-100 mg/dl. A lower level to prompt consideration of pathologic hypoglycemia and urge further evaluation have not been agreed upon, although some sources cite a range of 45-55 mg/dl or less. Hypoglycemia in infancy can be caused by hypermetabolic states (including infection), pituitary hormone deficiency, disorders of fatty acid oxidation or gluconeogenesis, insufficient glycogen stores, and CHI.

CHI, although rare, is the most common cause of severe persistent hypoglycemia in infancy. Comprised by a group of various genetic disorders, the common finding is hypoglycemia as a result of hypersecretion of insulin by pancreatic  $\beta$ -cells. Symptoms vary based on the severity of hypoglycemia and the age of the individual. Neonates often present with hypoglycemic seizures. Recurrent, severe hypoglycemia can lead to irreversible neurologic damage.

Immediate treatment of hypoglycemia secondary to CHI is an enteral or parenteral glucose load. Subsequent medical management may consist of glucagon (releases sugar from the liver), diazoxide (suppresses insulin release), or octreotide (decreases insulin secretion). Surgical options may also be available depending on the form of CHI (focal or diffuse) and an individual's response to medical management.

#### **References:**

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