[NM-207] Anesthetic management of a patient with GLUT1 Deficiency Syndrome

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Introduction: GLUT1 deficiency syndrome is a rare encephalopathy resulting from abnormal glucose transport into the brain. Patients usually present with early-onset seizures, developmental delay, and movement disorder. The hallmark biochemical feature is low glucose concentration in the CSF in the setting of normoglycemia (1). Treatment of GLUT1 deficiency syndrome revolves around a ketogenic diet in order to supply the brain with an alternative fuel source. Here we present the first description of patient with GLUT1 deficiency syndrome undergoing anesthesia and discuss the anesthetic implications of this rare disorder.

Case: A 22 month old girl with GLUT1 deficiency syndrome presented for bilateral myringotomy and insertion of ear tubes. Her initial presentation at 7 months of age was with seizures and developmental delay. Neurologic workup revealed a low glucose CSF concentration and genetic testing confirmed a GLUT1 gene defect. The patient was scheduled as a first case start in order to minimize NPO time. She underwent inhalational induction with sevoflurane and was maintained with mask anesthesia. Intranasal fentanyl was given for analgesia. Point of care glucose testing was performed and determined to be within normal limits. The patient tolerated the procedure well, was taking PO in PACU, and resumed her ketogenic diet. She was discharged home in stable condition.

Discussion: GLUT1 deficiency syndrome is very rare with fewer than 100 cases being reported in the literature prior to 2007 (1). Nonetheless, this disorder presents unique features that the practicing pediatric anesthesiologist should consider. These patients are frequently maintained on a ketogenic diet for seizure control. Accordingly, standard fasting guidelines are sufficient except clear liquids should be free of carbohydrates (2). Blood glucose should be monitored during the perioperative period and normoglycemia is desired. It is prudent to avoid glucose-containing solutions so as not to disrupt the patient’s ketotic state. Breakthrough seizures may occur and should be treated with anti-epileptics. The effects of general anesthetics on these patients have not been completely elucidated. However, there is some evidence that barbiturates further inhibit glucose transport in patients with GLUT1 deficiency and their use might put these patients at increased risk (3). In summary, GLUT1 deficiency syndrome is a rare disorder that warrants certain anesthetic considerations in this patient population.

References:
1. Klepper J and Leiendecker B. GLUT1 deficiency syndrome – 2007 update. Developmental Medicine & Child Neurology. 2007; 49: 707–716
2. McNeely JK. Perioperative management of a paediatric patient on the ketogenic diet. Paediatric Anaesthesia. 2000; 10: 103–106
3. Klepper J, et al. GLUT1-Deficiency: Barbiturates Potentiate Haploinsufficiency in Vitro. Pediatric Research. 1999; 46: 677–677
