

PBLD-Table #20

Don't Mess this Up! My Son is Finally Seizure Free on a Ketogenic Diet

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Goals:

- Comprehend the mechanism of a Ketogenic Diet
- Recognize the anesthetic implications associated with a Ketogenic diet
- Manage apprehensive and anxious parents in difficult clinical situations

Case Presentation:

A 4-year-old male, 14 kg with a complex medical history including intractable seizures treated with a ketogenic diet presented for an outpatient bilateral strabismus surgery. The patient had been seizure free for one year on this diet and had been successfully weaned off all anticonvulsants. Other medical conditions concerning were his history of myelodysplastic syndrome status post bone marrow transplant, congenital bicuspid aortic valve, elevated blood pressures, recurrent pulmonary infections, reactive airway disease controlled with albuterol, obstructive sleep apnea, poorly controlled reflux and global developmental delay including failure to thrive, all of which contributed greatly to the complexity of this patient's picture as well as to the anesthetic plan for this particularly outpatient procedure. The patient's mother was very concerned about maintaining the ketosis while under anesthesia as the patient had previous episodes of grand mal seizures after general anesthesia for various other procedures.

Questions:

What is a ketogenic diet and why is it used? What other studies or labs would be indicated? If the child were not cooperative, what would you use for premedication? Do you plan on a rapid sequence induction for his poorly controlled reflux?

Case history and physical examination (continued)

On exam, the patient was awake but appeared dehydrated and tired and agitated. His lungs were clear to auscultation and his cardiac exam was regular rate and rhythm without a murmur. His neuro exam was significant for developmental delay and generalized hypotonia. He was non-verbal. After careful consideration of the potential interactions with the ketogenic diet with some of the commonly used medications during general anesthesia and the type of procedure the patient was having done, it was decided to perform the procedure under general anesthesia with total intravenous anesthesia.

Questions:

How would you proceed with the anesthesia plan? How would you describe your anesthetic plan to a medically aware and apprehensive parent? What medications do you plan on administering or avoiding?

Case history (continued)

The patient was not premedicated per parental request. Instead parental presence for induction was requested. Mask induction with Sevoflurane was used and an IV placed after the patient was asleep. An attempt was made to insert a size 2.0 flexible LMA but was not successful. This was followed by placement of a size 1.5 AIR Q LMA, which also failed to sit properly, even with cuff inflation. The decision was made to secure the airway with a cuffed endotracheal tube size 4.5. It was a Grade 1 view easy intubation without any obvious anatomical abnormalities. After the airway was secured, the

Sevoflurane was turned off and a Propofol infusion was started at 250 mcg/kg/min and maintained throughout the procedure. Dexamethasone 0.05 mg/kg was also administered. IV fluid given for hydration was Normal Saline. Prophylactic Albuterol was given immediately after intubation as well as right before extubation because of the patient's history of reactive airway disease. The procedure proceeded uneventfully and there were no seizures observed. The patient had a straightforward emergence and was extubated and transported to PACU. During the patient's recovery in PACU, which was also uneventful, the patient's mother became increasingly upset when she found out that Dexamethasone was administered to the patient because she was concerned his blood glucose was going to be too elevated and that he would have a seizure. She refused to have the anesthesiologist at the bedside to check his blood glucose or even allow the nurse to check one. The pediatric neurologist was consulted. He felt that this low dose of Dexamethasone would not be a concern but the mother was not willing to accept the reassurances, ultimately displaying threatening behavior to the anesthesiologist.

Questions:

Does the administration of Dexamethasone in antiemetic doses increase blood glucose levels? What other medications do we routinely administer that could influence a patient's glucose level or alter the metabolic acidosis? How do you find this out at your institution? What should we do during a code or during treatment of anaphylaxis or if undergoing cardiac anesthesia where vasopressors containing dextrose would be needed? How confident are you that this patient won't have a seizure after they leave PACU and would you consider 12 or 24 hr. observation? How would you handle this frustrated and threatening parent?

Discussion:

Ketogenic Diet

Epilepsy is a disorder where unprovoked, recurrent seizures are caused by abnormal electrical discharges within the brain. Children are particularly susceptible to seizures and most epilepsy begins in childhood. Most seizures can be controlled by a single anti-epileptic medication, but sometimes seizures develop which are resistant to these medications or the side effects of the medications become intolerable. The ketogenic diet has been widely used to treat children with drug-resistant epilepsy since the 1920s. It is a high fat, low carbohydrate with adequate protein diet. While the exact mechanism of action is still unclear, it is hypothesized that the high fat and restricted carbohydrate content of the diet mimics what the body does under periods of starvation where ketone bodies become the main source of fuel for the brain's energy demand. Somehow this metabolic transition is associated with improved seizure control. The ketogenic diet is based on a ratio of fat: carbohydrate and protein of 3:1 or 4:1, which means 3 to 4 g of fat to 1 g of protein and carbohydrate combined. There are multiple forms of ketogenic diets available, from the classic ketogenic diet, a highly restrictive diet with a fat to protein and carbohydrate of 3-4:1 to Medium Chain Triglyceride Ketogenic diet (MCT)-KD to modified Atkins diet (MAD) and low glycemic index treatment (LGIT). The more liberal diets seem to be as effective in most of the seizure types but are much more palatable. If the seizures are successfully controlled or decreased in frequency then the diet is usually continued for about 2-3 yrs.

Children with baseline metabolic derangements and mitochondrial disorders may have their conditions exacerbated by the ketogenic diet. Conditions such as pyruvate carboxylase deficiency and similar conditions where energy metabolism is compromised could experience increased seizures on the ketogenic diet and metabolic decompensation especially during periods of severe illness. It's important for us to keep in mind that children on the diet can become acidotic.

Complications of the diet include potential for hypoglycemia, ketosis with associated lethargy, nausea, vomiting, even coma. It's possible to have platelet dysfunction while on the diet as well as cardiac complications such as prolonged Qtc and possible cardiomyopathy all of which could potentially be affected while under anesthesia. Children on a ketogenic diet do appear to be at risk for metabolic acidosis while under general anesthesia, especially during long surgical procedure.

In Summary: It is important for us as Anesthesia Providers to understand the metabolic changes that can occur in patients on a ketogenic diet and how our anesthetic managements of these patients must be carefully tailored to avoid seizures or the worsening of acidotic states. It is also important to review the

drugs before given to minimize the exposure of dextrose containing medications if suitable. Ideally one should have a hospital protocol for drugs to avoid for those patients to minimize mistakes.

Managing frustrated parents:

Always difficult – but a direct clear communication in a calm manner is one of the corner stones; to call for help while managing the upset parent is another, e.g. risk management, another anesthesia provider to help with following patients. A great summary about this topic ('When things go wrong' - responding to adverse events) is listed as reference.

References:

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