Title: Anesthesia Considerations in Pediatric Patients with Glycogen Storage Disease Type IX

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Introduction: A total of nine types of this particular glycogen storage disease (GSD) reported in the pediatric literature all of which involve a defect in the synthesis and or breakdown of glycogen within muscle and liver cells.[1] Type 9 is due to a deficiency of phosphorylase kinase. The most common form is an X-linked liver phosphorylase kinase deficiency.[1] Ketosis occurs with fasting associated with mild hypoglycemia. This particular type of GSD can improve as the child matures and prolong fasting may occur without hypoglycemia and ketosis. [2]

Methods: We present a patient who was admitted to our pediatric hospital three times in a two week period for diagnostic studies and surgery under anesthesia. Because we are a tertiary referral center, our patient was followed by his pediatric gastroenterologist in another city and sent his patient to our hospital with orders for preoperative intravenous glucose the night prior to surgery. However when we questioned the mother about the patient home care, she stated that he slept through the night without the need for nighttime supplementation to his regular diet.

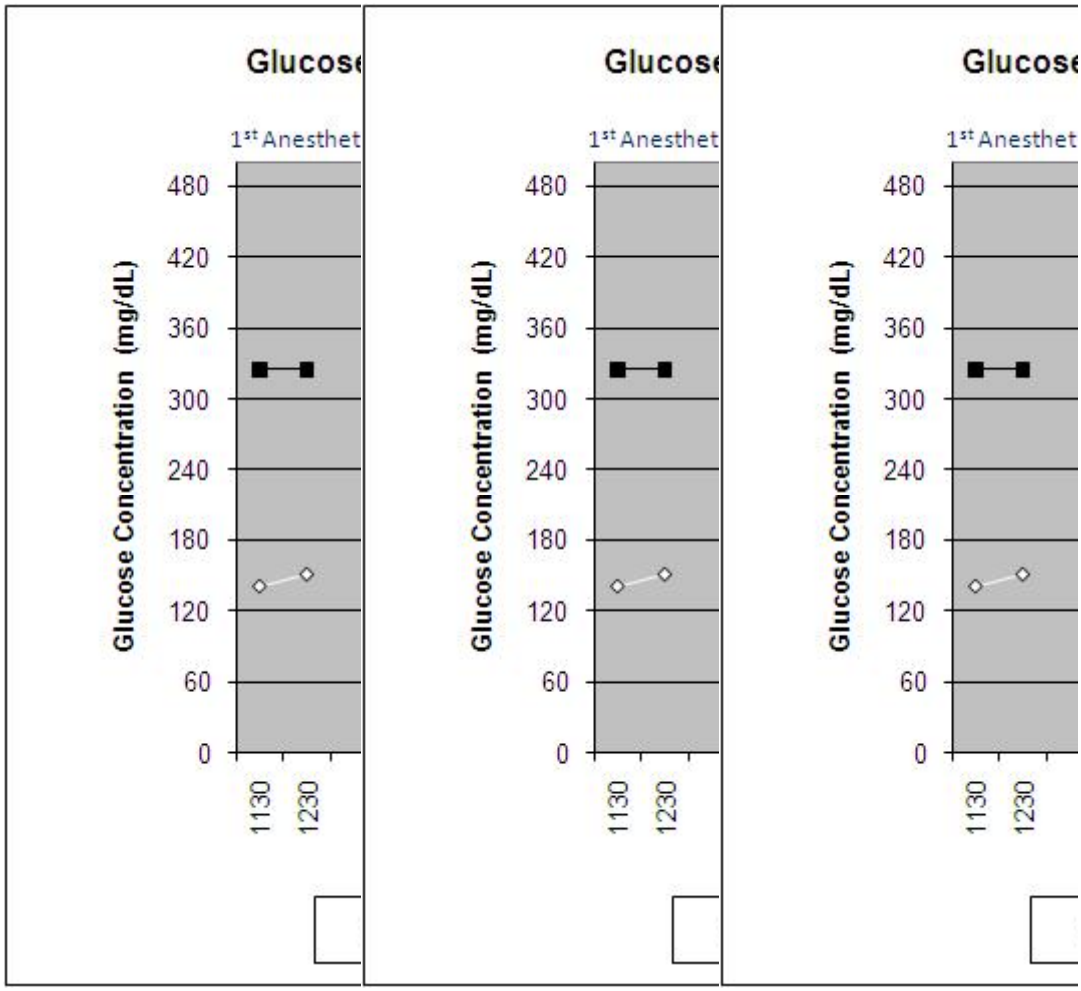
Past Surgical History included lumbar laminectomies. Past Medical History was significant for his type 9 GSD. He first presented for a MRI followed by a lumbar laminectomy for an epidermoid cyst and then followed by a repeat MRI postoperatively. His physician ordered that he receive a 10% dextrose infusion continuously while hospitalized. All his anesthetics were uneventful and he was discharged home without complications.

Results: Attached please find a flow sheet graph of intraoperative glucose levels and base excesses during the laminectomy and MRIs.

Discussion: We believe that a good history of the need for feeding at short intervals or as in our patient who had outgrown the need for frequent feedings during the night will help with the need to either admit the patient for intravenous glucose during long periods of fasting or not. However in those children who need glucose, following their intraoperative glucose is mandatory. We present a patient with GSD type 9 whose glucose was well controlled perioperatively.

Refs:

1. Tsong Y. et al., *Nelson Textbook of Pediatric* 2007.
2. Willems P. et al, Eur J Pediatr 1990.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