

GLYCOGEN STORAGE DISEASE

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Background Information: The Pediatric PACU nurses were presented with a challenge of caring for patients that have Glycogen Storage Disease (GSD). Glycogen storage disease type I is an inherited disorder caused by the buildup of a complex sugar called glycogen in the body's cells. The accumulation of glycogen in certain organs and tissues, especially the liver, kidneys, and small intestines, impairs their ability to function normally. The nurses were not familiar with GSD and an educational session was required. These patients could not adhere to our NPO guidelines because they need to ingest cornstarch to maintain a normal blood sugar. This alone posed a problem. A new plan of care was needed for these patients

Objectives of Project: The Pediatric PACU nurses needed to know accurate NPO guidelines for these children. Can they have their cornstarch mixture prior to surgery? How many hours prior to surgery can they ingest the cornstarch? How often should we be checking blood sugars? Do these patients need to go to the PICU?

Process of Implementation: The patients could not follow the established protocol of being admitted overnight and having an IV of dextrose solution infusing to maintain blood sugar. These patients had poor vascular access and needed to be put to sleep to have an IV started. Pediatric Anesthesia, Nurse Managers, and Educator met with GSD MD to discuss a protocol to care for this population of patients in the pre/post setting.

Statement of Successful Practice: After the discussion with specialist on GSD, anesthesia, managers, and educator it was decided that the PACU nurses needed education on GSD. The GSD MD provided an in service for the staff on children with GSD. A new routine care protocol was developed by the physician and anesthesia.

Implications for Advancing the Practice of Perianesthesia Nursing: The Pediatric PACU nurses are beginning to pre-op and post-op this unique population of patients. Routine protocols in pre-op and post-op care have been altered to meet the needs of these patients. Specifically, changes in NPO guidelines. The new protocol allows the nurses to deliver safe, quality, evidence-based patient care.

Reference

Glycogen storage disease type I (2015). Genetics Home Reference: Your guide to understanding genetics. Retrieved from: <http://ghr.nlm.nih.gov/condition/glycogen-storage-disease-type-i>