KETOGENIC DIET NPO GUIDELINES: The goal of these guidelines is to maintain ketosis and prevent the effects of excessive ketosis which include hypoglycemia and acidosis while child is NPO.

Overview of ketogenic diet (KGD) therapy: The KGD is medical therapy for children with intractable epilepsy and for metabolic disorders including pyruvate dehydrogenase deficiency and glucose-1 transporter deficiency. This high fat diet regimen (70-90% of calories) forces the body into a dietary induced ketosis. The acidosis that occurs when the diet is first initiated corrects itself within days and is not sustained. The diet is manipulated with different ratios of fat to protein plus carbohydrate, much like the dosing of an anti-seizure medication, to achieve optimal seizure control. The literature is reporting that 60% of children placed on the diet experience a 50% or greater improvement in seizure control. The children with successful seizure control often have their anti-seizure medications reduced and in some cases completely discontinued.

Managing a child on the KGD who require NPO for procedures or surgery

1. **Fasting**: The KGD child can be fasted in the same fashion as other children. Clear liquids can include: "sugar free, caffeine free" such as diet caffeine- free sodas, sugar free gelatin, ice chips, and water. If fasting corresponds with vomiting, Pedialyte can be given in place of water.
2. **Medications**: Must be in lowest carbohydrate form such as crushed swallow tabs or I.V. DEXTROSE FREE form (no chew tabs, syrups, elixirs, reconstituted solutions). Check with the ketogenic diet team and/or the pharmacy before ordering new medications.
3. **Intravenous solutions:** Use 0.9 NaCl. Lactated ringers are also carbohydrate free. Dextrose solutions are to be avoided unless blood glucose levels are less than 50 mg%, when it is advisable to hang D2.5% to maintain a stable glucose level with goal 50-70 mg%.
4. **Glucose levels**: Check prior to surgery/initiation of NPO status, and then every 2 hours during surgery and after surgery until PO/feeds are able to be initiated. If less than 50 mg%, add D2.5% dextrose to maintain blood glucose between 50-70 mg%. Levels may increase due to stress response.
5. **Arterial pCO2:** Check prior to surgery and then every 2 hours during surgery and after surgery until PO/feeds are able to be initiated. Extended fasting may lead to excessive ketosis and acidosis. Intravenous bicarbonate should be administered to correct acidosis.
6. **Fluid volumes**: Children on the KGD follow a maintenance "fluid schedule" to ensure adequate hydration throughout the day. There have been reported cases of children on the KGD who have had breakthrough seizures after receiving large boluses of fluids at one time; therefore acute over-hydration needs to be avoided. Give fluids at maintenance unless discussed with KGD team
7. **Advancement of diet:** For Oral and enteral feeders, start with Pedialyte and then to half strength KGD prescription, and finally to full strength KGD prescription. When on half strength KGD prescription, blood glucose does not need to be checked and arterial pCO2 does not need to be checked. Urine ketones and USG checked q void per protocol.