

PERI-OPERATIVE NUTRITIONAL CARE GUIDELINES FOR CHILDREN WITH SMA OR NEUROMUSCULAR DISORDERS

Nutritional Management during catabolic states is important to minimize fatigue and prevent respiratory failure for children with neuromuscular disease undergoing elective surgical procedures. Their diminished lean body mass limits mobilization of amino acids during prolonged fasting or illness. In the peri-operative setting, it is common to restrict feeding after midnight the day prior to the procedure. In the case of a prolonged procedure, such as scoliosis surgery, this can mean that children will have restricted oral intake for 24 -36 hours. This is subsequently followed by the slow introduction of nutrition, starting with clear liquids. Some children may experience nausea due to the medications they've received during the procedure or afterwards to treat pain related to the procedure. Under usual circumstances, children typically receive intravenous fluid with sugar during the recovery period. However, the amount of calories received is limited, and children with neuromuscular disease appear to be more likely to develop hyperglycemia (higher blood sugars) in this setting. Finally, many neuromuscular patients have abnormal gastric motility and reflux which can be exacerbated in the post-surgical setting.

Prolonged fasting or inadequate caloric intake can negatively impact recovery by enhancing fatigue and making it more difficult for them to wean off ventilatory support. Under the most ideal circumstances, pre-operative fasting should be limited to no more than 6 hours in SMA type 1 subjects or weak neuromuscular infants, and to no more than 8 hours in SMA type 2 or type 3 subjects or other neuromuscular patients.

Recommended NPO times

6-8 Hours before procedure (depending on child's age): As per anesthesia guidelines – No solids, formula or non-breast milk allowed. Clear liquids and breast milk are still acceptable.

4 hours before procedure:-As per anesthesia guidelines-Clear liquids only from this point forward. Children be encouraged to drink fluid up until the 2 hour limit. Clear liquid could be a beverage that contains glucose as well as amino acids.(see below). Other acceptable but clear liquids include water, pedialyte, apple juice, Gatorade, or clear soda such as Seven-Up or Sprite.

2 hours before procedure- As per anesthesia guidelines-No more intake prior to surgery after this time point.

Clear Liquid Alternative for Neuromuscular Patients:

Resource Fruit beverage or Boost Breeze are two options (parents can go to www.resource.walgreens.com and look under before/after surgery medical nutrition tab). Other comparable clear liquid products include Nestle Carnation Instant Breakfast Juice Drink www.nestle-nutrition.com

During Operative Procedure

An intravenous line (I.V.) will be placed prior to the procedure, sometimes in the pre-operative waiting area and sometimes in the operating room. The doctor or nurse may check a blood sugar

level and other laboratory studies at this time. Depending on the length of the procedure, intravenous fluids with or without sugar will be administered to maintain hydration.

Post-Operative Management

Once your child has been taken to the recovery room, peripheral parenteral nutrition (PPN) can be started through the I.V.. The intravenous solution used will include dextrose 10-15%, 1.5 gms/kg amino acids per 24 hour period and standard vitamins, minerals and electrolytes. This will provide a source of sugar and amino acids to help maintain your child's energy level, and keep blood sugar levels stable. If oral intake is restricted for more than 48 hours, the dietician may recommend adding a 10% intralipid infusion. However, since SMA patients have a secondary defect in fatty acid oxidation, total fat from all sources should not exceed 15-20% of total calories (all intralipid infusions available in the U.S. currently contain long-chain fatty acids). Peripheral parenteral nutrition (PPN) will need to be ordered in advance the morning of the procedure in order to be ready during the post-operative period. Most pharmacies require several hours notice to prepare these solutions. Total peripheral nutrition, or TPN, requires a larger IV, and allows even more sugar and fat to be administered to help boost calories. However, this is usually not necessary in an uncomplicated peri-operative setting.

Recommendations for oral re-feeding:

Once a child begins taking clear liquids, PPN or TPN can be discontinued. However, if nausea or decreased appetite limit intake, PPN can be used for several days to enhance the recovery process. The clear liquid beverages which contain hydrolyzed protein can also be used in this setting to provide an additional protein source if the child is unable or unwilling to advance to regular feeds. If the child has a g-tube, he or she can begin receiving an elemental or semi-elemental formula, like pediatric vivonex, or tolerex (double-diluted), if regular formulas are not tolerated. These formulas are sometimes better tolerated than regular formulas like pediasure, as they have lower fat levels, and help facilitate more rapid gastric emptying.

The goal should be to advance oral supplementation to estimated full caloric requirements based on body weight/length for neuromuscular patients (see document entitled: general nutritional guidelines for neuromuscular patients) no later than one to two days post-operatively, or sooner if possible.

For difficulties in weaning from mechanical ventilation, see "CARE GUIDELINES FOR SMA CHILDREN WITH ACUTE RESPIRATORY ILLNESS".