Pre/Post Operative Guidelines for Hospital Staff
SMA and Neuromuscular Weakness

The goal of this document is to standardize the peri-operative care management of vulnerable neuromuscular patients undergoing elective surgical procedures. These children have a increased risk for respiratory failure, gastrointestinal dysmotility and increased weakness in the setting of a catabolic state. Prolonged inadequate caloric intake can negatively impact recovery by enhancing fatigue and making it more difficult to wean off ventilatory support. Further, medications used for pain management can aggravate underlying gastrointestinal dysmotility and increase risk for postoperative respiratory failure or pneumonia. Under ideal circumstances, **pre-operative fasting should be ideally limited to no more than 6 hours** in SMA type I or II subjects or weak neuromuscular infants.

1. **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: Clear liquids only from this point forward. Children be encouraged to drink fluid up until the 2 hour limit.
   - 2 hours before procedure- As per anesthesia guidelines-No more intake prior to surgery after this time point.
   - **Peripheral parenteral nutrition (PPN) should be ordered pre-operatively and ideally, begun either when the child is made NPO or shortly following the procedure.** The basic recipe includes dextrose 10-12%, 1.5 gms/kg amino acids per 24 hour period/standard vitamins, minerals and electrolytes. Lactated ringer's may be substituted during the actual procedure.

2. **POST-OPERATIVE MANAGEMENT**
   - PPN should be resumed within first 1- 2 hours post-op and ideally prior to extubation. A nutrition consult is mandatory, along with daily weights.
   - PPN should continue until full baseline caloric intake is resumed. PPN can be tapered as increased GI feeding is tolerated. If their usual formula is not tolerated, starting with half-strength or substituting an elemental or semi-elemental formula can be considered to help facilitate gastric emptying in the immediate post-op period.

3. **RESPIRATORY CARE POST-OP**
   - For <u>TYPE I SMA patients post Nissen-G-tube surgery</u>, **extubation to bipap to be performed in the PICU** at staff discretion when the patient is:
     - afebrile and minimal requirement for supplemental O2
     - on minimum respiratory depressants/narcotics
     - airway suctioning is 1 time/hour or less
   - Extubate to continuous nasal mask BIPAP ventilation and wean as tolerated
     - IPAP 15-20 and EPAP 3-6 using the spontaneous timed mode with a backup rate: <1yr: 30/min; 1yr-18 months: 25/min; > 18 months 20/min
   - **Lower Cough Assist pressures may be indicated post-extubation following Nissen-G-tube surgery**: +25 for 1 second, -25 for 1 second, and 1 second pause: 5 sets/5 reps at least every six hours or as needed. Resume pre-op pressures within 72 hours post-op.
   - Wean cough assist regimen and BIPAP to pre-op baseline as tolerated.

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