

NUTRITIONAL CARE GUIDELINES FOR CHILDREN WITH SMA OR NEUROMUSCULAR DISORDERS DURING ACUTE ILLNESS OR FASTING

Revised 6/2006 by Kathryn J. Swoboda, M.D. University of Utah School of Medicine and Primary Children's Medical Center, Salt Lake City, Utah. Telephone 801-662-1000 for urgent questions; non-urgent questions call 801-585-1676.

Proactive Nutritional Management during illness or other catabolic states is critical! As a general guideline, do not allow fasting > 6 hours in SMA type 1 subjects, or > 8 to 12 hours in SMA type 2 or type 3 subjects or other neuromuscular patients when they are ill. SMA patients have a secondary fatty acid oxidation defect. In addition, many children have reflux and delayed gastric emptying which can worsen with illness.

In the event of decreased oral intake or refusal to take oral feeds, you can first try the following strategies:

-Oral re-hydration with a clear beverage that contains glucose as well as protein (hydrolyzed protein or amino acids): Resource Fruit Juice 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).

-If tolerated, advance to either an oral elemental or semi-elemental formula, like pediatric vivonex or tolerex (double-diluted). These formulas may be better tolerated during illness than regular formulas since as they have lower fat levels and can facilitate gastric emptying.

If your child can't or won't tolerate oral intake, or has recurrent vomiting, your next recourse is your local emergency room, where they can consider the following options:

1) Placement of an IV for hydration (and treatment to stop vomiting or treat underlying cause if necessary). This may help your child recover enough to allow reinstatement of oral feeds. If this strategy alone is ineffective, they may need to place a temporary nasogastric (NG) or nasojejunal (NJ) tube, and begin continuous feeds with a formula containing amino acids or protein, as described above.

2) If neither oral nor tube feeding are tolerated, supplementation with peripheral parenteral nutrition (PPN) can be instituted via an IV in order to help meet protein, vitamin and mineral requirements. This can help optimize recovery and avoid hyperglycemia, which often occurs when only IV glucose is used. The goal is to provide intake via whatever means necessary to reach estimated full caloric requirements within 4 to 6 hours of presentation, using a combination of the above strategies if necessary.

Guidelines for calorie, fluid and protein requirements, and fat intake:

Calories: Since children with SMA and neuromuscular disease often have reduced lean body mass, their metabolic needs may be somewhat less than other children their size or age. We estimate 9 - 11 calories per cm height as a daily requirement. If contractures preclude accurate

height measurement, arm span can be used as an alternative.

Fluids: Typical maintenance daily fluid requirements are 115 - 135 ml per kg body weight (may be higher with fever or dehydration, since many patients drink inadequately when ill).

Protein: 1.0 - 2.0 grams/kg/day total intake should be sufficient in most cases

Fat: Total fat from all sources for dietary supplementation during an illness should not exceed 15-20% of total calories, since these children have a secondary fatty acid oxidation problem.

Carnitine supplementation during illness may be helpful in improving energy metabolism in this setting: 50-100 mg/kg/day orally or IV.