

Nutritional Guidelines for Caregivers and Hospital Staff SMA/Neuromuscular Patient with Acute Illness

Proactive Nutritional Management

- Individuals with reduced muscle mass are at increased risk for metabolic acidosis, blood sugar and electrolyte disturbances in the setting of acute illness – this is especially true for infants and young children.
- Reflux and delayed gastric emptying can become evident for the first time or dramatically worsen with illness, increasing the risk of vomiting and aspiration, which can result in serious respiratory complications.
- The guidelines below are meant to be general in scope. Please consult with your primary care provider at the earliest sign of illness for specific guidance.

For those unable to tolerate usual oral or gastrostomy tube feeds during illness

- A clear or low-residue formula that contains a sugar source, electrolytes, vitamins and minerals, and protein (isolated whey protein, hydrolyzed protein, or amino acids) can be substituted for usual formula or food during an acute illness. These formulas may be better tolerated since they contain little or no fat and a more easily digestible form of protein. A variety of products are commercially available and can be purchased at your local pharmacy (e.g. Pediasure Clear, Ensure Clear). Please consult a dietitian to pick the best alternative for your situation. Since these formulas contain significant amounts of sugar, diluting them 1:1 with Pedialyte or water may be helpful in the setting of diarrhea.
- Alternatively, dilute their regular formula 1:1 with Pedialyte or similar product. Advance as tolerated to their regular formula or breastmilk (infants), a full strength clear protein-containing beverage or formulas (child > 1 year), or a low-fat elemental or semi-elemental formula (e.g. Pediatric vivonex or Tolorex).

For those who can't tolerate oral or continuous tube feeds, or have recurrent vomiting, seek emergency care within 6-12 hours

- An IV for hydration (+/- medication to treat nausea/vomiting) may buy some time to allow your care team to rule out more serious conditions. In some cases, resuming oral/tube feeds in the ER may be possible. In those without a gastrostomy tube, temporary placement of a nasogastric (NG) or nasojejunal (NJ) tube can be used to provide temporary nutrition with a clear formula containing amino acids or protein.
- Supplementation with peripheral or total parenteral nutrition (PPN or TPN) may be indicated to meet ideal protein, electrolyte, carbohydrate, vitamin and mineral requirements. The ideal nutrition goal is to achieve full caloric replacement within 6-12 hours of presentation.

Guidelines for calorie, fluid, protein and fat intake during acute illness

- **Calories:** Metabolic needs are often reduced for age. Maintenance of growth and weight gain in the child with neuromuscular disease is 7-11 cal/cm height (arm span or ulnar length can be used to estimate height if needed).
- **Fluids:** Typical maintenance daily fluid requirements are 115 - 135 ml per kg body weight (or higher with fever or dehydration).
- **Protein:** 1.0 - 2.0 grams/kg/day total intake is ideal in most cases
- **Fat:** Total fat from all sources for dietary supplementation during an illness should not exceed 20% of total calories (except for infants < 1 year). Carnitine supplementation may help to improve energy metabolism: 50-100 mg/kg/day orally or intravenously.