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General Nutrition Guidelines for SMA Children

Nutrition is critically important for maintaining muscle mass, strength and clinical function in patients with SMA. An experienced nutritionist is vital in the management of patients with neuromuscular conditions or metabolic disorders.

Swallowing problems are sometimes overlooked in children with SMA, particularly among weaker children or in those with borderline nutritional status or who experience frequent respiratory illnesses. Swallowing studies should be performed on a regular basis to ensure that silent aspiration isn't contributing to respiratory problems.

Maintenance of appropriate nutrition is especially critical during illness. Children with SMA, particularly those with SMA type 1 or type 2, have diminished lean body mass and limited energy (glycogen) reserves in the setting of prolonged fasting. With prolonged fasting, the body will begin to break down muscle proteins; for children with SMA, this can result in the loss of up to 10% of muscle mass in the context of extreme fasts. Thus, when concerns regarding the ability to safely administer oral feeds develop, alternative forms of nutrition should be considered. Options include temporary nasogastric or nasojejunal feeds, or peripheral or total parenteral nutrition (PPN or TPN).

There is no established basis of caloric and nutritional needs specific to the SMA population. Based on experience, the following parameters are recommended:

Calories - 9-11 kcal per centimeter height (conversion from inches: $\text{cm} = \text{inches} \times 2.54$)

Ideally, caloric intake should be monitored and adjusted as needed to maintain weight for height ratio between 5th and 10th percentile. Based on data from older SMA patients with type 2 and 3 disease, children with BMI of 5% have median (50th percentile) fat mass. This is because SMA subjects have significantly reduced lean mass.

Protein

Recommended range is 1 - 2 grams per kilogram body weight (conversion from pounds: $\text{Kg} = \text{pounds} \text{ divided by } 2.2$). Protein in excess of 2 grams/kg/day over a long period of time could potentially result in kidney problems and negatively affect bone density.

Fat

30% of total calories. Children less than age 2 years require a critical minimum amount of fat in the form of essential fatty acids for normal brain development. Please consult a dietician if you have specific questions regarding such guidelines.

Fiber

A good general guideline is age plus 5 (a 3 year old would need $3 + 5 = 8$ grams of fiber a day)

Water

115 - 135 ml/kg (conversion is weight in pounds divided by 2.2). Children with fever may require more in some cases.

Nutritional Management for Infants and Children with SMA type 1

Formula Options:

At this time, it is unknown whether there is a particular benefit from usage of a specific brand or type of formula, or whether certain formulas are better tolerated in infants and children with SMA type 1. Even after gastrostomy tube (g-tube) placement, infants may continue to benefit from receiving breast milk, which is of particular benefit by bolstering the infant's immature immune system. If the child maintains the ability to safely feed by mouth (documented by a swallow study), he or she can sometimes still breast or bottle feed. Even in such situations, supplementation via gastrostomy is usually needed to maintain adequate weight gain and avoid the development of malnutrition due to insufficient nutritional intake. In such cases, weekly monitoring of weight gain can help guide appropriate g-tube supplementation.

Obviously, sputtering or choking during feeds is indicative of more severe swallowing dysfunction. Such infants are likely at risk of aspiration, and feeding should be restricted to g-tube only.

Elemental formulas may be easier to digest because they contain free amino acids rather than whole proteins; however, they taste poorly and are much more expensive than conventional formulas. While there is no evidence that elemental formulas are superior to regular formulas for those infants who tolerate them, many families of children with SMA feel passionately that this approach was effective for their children.

More controversially, some clinicians, and many families of children with SMA, advocate use of elemental formulas with low fat content, feeling that these formulas (vivonex, tolurex) may be better tolerated than those with higher fat content, such as Neocate. Children with SMA type 1 may have abnormal gastric emptying; in this context, a lower fat, elemental based formula such as Vivonex may improve motility, particularly among children demonstrating reflux or other difficulties tolerating feeds. There is, however, no objective data supporting the effectiveness of this approach. Given the potential harm of low fat dietary regimens as applied to infants and young children, it is particularly important that any such regimen be developed in coordination with a nutritionist experienced in the care of children with SMA.

Since babies with SMA have a lower lean body mass, lower activity, and thus (presumably) lower caloric needs, the quantity of formula should be adjusted as needed to meet the baby's growth requirements, with a goal of maintaining a weight of 5th to 10th percentile for age and gender.

Constipation is frequent in infants with SMA type I, and can add to abdominal bloating and discomfort and worsen reflux. Regulation of bowel movements with glycopyrrolate or a promotility agent such as metoclopramide may be indicated if they aren't having at least one or more bowel movements every day.

Feeding Strategies:

Bolus feeding may be poorly tolerated in children with SMA type 1; smaller, more frequent bolus feeds during the day may prove superior, with feeds every 3 hours or so during waking hours. This may become particularly important if the child does not receive a Nissen fundoplication during feeding tube placement. Nissen fundoplication, a procedure where the stomach is wrapped around the esophagus to increase pressure on the lower esophagus, is performed to reduce gastroesophageal reflux. Reflux is an especial problem in children with SMA, particularly for those with compromised ability to protect their airway. Reflux can, unfortunately, be worsened by proactive respiratory care using a cough assist machine or BiPAP; these approaches push pressurized air into the stomach (as well as the lungs), which can further aggravate reflux. It is important to vent the g-tube during treatments with the cough assist device, and when on BiPAP.

Additionally, the child's head and body should be elevated at a 30 degree angle from horizontal for at least 30-60 minutes following a bolus feed to limit reflux. A wedge under his/her mattress or play area may also work well, or a seat that is angled so that the stomach isn't compressed.

It is important to not assume that your child will not tolerate regular formula until you try it. Some infants tolerate regular formula without difficulty; regular formula, if tolerated, is to be preferred over alternative, elemental formulas (which may expose your child to potential dietary imbalances). If he or she fails to tolerate the regular formula with bolus feeds, a continuous feeding regimen using a pump may be helpful, or smaller bolus feeds during the day in combination with continuous nighttime feeds. Only if those strategies fail would we recommend substituting an elemental formula.

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

Proactive Nutritional Management during illness or other catabolic states is critical!

As a general guideline, do not allow fasting > 6 hours in SMA type I subjects or infants < 1 year of age with other neuromuscular disorders, or > 8 to 12 hours in SMA type II/III subjects or other neuromuscular patients when they are ill.

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):

1. 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).
2. Other comparable clear liquid products include Nestle Carnation Instant Breakfast Juice Drink (www.nestle-nutrition.com).

These clear beverages contain a significant amount of glucose or dextrose, and may need to be double diluted for use in some settings.

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: Children with SMA and neuromuscular disease have reduced lean body mass so metabolic needs may be somewhat less than other children their size or age. We estimate 9 - 11

calories per cm height as a typical daily requirement for maintenance of growth and weight gain. 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. Carnitine supplementation during illness may be helpful in improving energy metabolism in this setting: 50-100 mg/kg/day orally or IV.

PERI-OPERATIVE NUTRITIONAL CARE GUIDELINES FOR CHILDREN WITH SMA

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 I subjects or weak neuromuscular infants, and to no more than 8 hours in SMA type II or type III 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 should 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 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 gm/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. 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. Clear liquid beverages can also be used in this setting 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 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 no later than one to two days following the operation, or sooner if possible.