
Nutritional Guidelines for SMA patients and their parents

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■ Current Recommendations:

- There is little published research data on diet and SMA for guidance
- General recommendations for a healthy diet:
 - 5 servings of fruits and vegetables a day
 - Fat < 30 % of calories
 - Include complex carbohydrates (whole grain breads, brown rice, beans and legumes, fruits, and vegetables, etc.)
 - Avoid simple sugars (soft drinks, candies, flavored drinks, etc.)

General Recommendations: Children 2-3 Years Old

Food	# servings	Portion size
Meat	2	1-2 oz
Milk or Dairy	4-5	½ cup
Fruits/Vegetables	5	
Cooked vegetables		2-3 TBSP
Raw vegetables		Few pieces
Raw fruit		½ to 1 small
Canned fruit		2-4 TBSP
Fruit juice		3-4 oz

■ General Recommendations:
Children 2-3 Years Old, Continued

Food	# servings	Portion size
Grains	3	
Whole grain breads		½ - 1 slice
Cooked Cereal		¼ - ½ cup
Dry Cereal		½ - 1 cup

Recommendations: Children 4-6 Years Old

Food	# of servings	Portion size
Meat	2	1-2 oz
Milk or Dairy	3-4	$\frac{1}{2}$ - $\frac{3}{4}$ cup
Fruits and Vegetables	5	
Cooked vegetables		3-4 TBSP
Raw vegetables		Few pieces

■ Recommendations:

Children 4-6 Years Old, continued

Raw fruit		½ -1 small
Canned fruit		4-6 TBSP
Fruit Juice		4 oz
Grains	3	
Whole Grain Breads		1 slice
Cooked Cereal		½ cup
Dry Cereal		1 cup

■ Energy Requirements:

- Energy needs are lower due to lower muscle mass, and decreased movement
- Estimation of caloric needs based upon an equation formulated for children with similar physical disabilities (spina bifida):
 - 9-11 kcal/cm for weight maintenance
 - 7 kcal/cm for weight loss

Pediatric Nutrition in Chronic Diseases and Developmental Disorders, Ekvall SW, Bandini L, Ekvall V, Oxford University Press, 1993;168

■ Protein Requirements:

- Patients may have higher protein requirement due to muscle atrophy, especially during catabolic states.
- The muscles normally provide significant reserves of glycogen, protein and minerals.
- During fasting, glycogen is broken down to provide glucose for energy use within the muscles, and protein is broken down to provide amino acids which are shuttled to the liver for gluconeogenesis to provide energy for the body.
- Estimated needs: At least 1 g/kg, possibly as much as 2 g/kg of body weight a day*

* Dr. Richard Kelley
Kennedy Krieger Institute, and
Department of Pediatrics, John Hopkins University School
of Medicine

■ Fat Requirements:

■ General recommendations:

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- Limit fat to < 30% of total calories. (Caution: infants and children under 2 years of age)
 - Limit Saturated Fat + Trans Fatty Acids to <10% of total calories.
 - Saturated Fat and Trans Fatty Acids are found in baked goods, cakes, cookies, crackers, processed foods, etc.
 - < 300 mg of dietary cholesterol a day
 - Up to 20 % of calories can come from Monounsaturated fats. Monounsaturated fats are found in Olive Oil, Nuts, Nut butters such as almond butter or peanut butter, Avocados, etc.

■ Fat Metabolism Abnormalities:

- Patients with more severe forms of SMA tend to have higher levels of medium chain fatty acids (C12) in the plasma, and higher concentrations of derivatives of in urine
- These abnormalities have been observed in both fasting and non fasting states in SMA children
- It is unknown at present to what extent diet has an influence on these parameters in SMA patients...but a catabolic state (fasting, ill, inadequate caloric intake) or high fat intake could worsen such abnormalities

■ Fiber Requirements:

■ General Recommendations:

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- Age + 5 (e.g. a child that is 3 years old needs about 8 g of fiber a day)
 - Foods high in fiber are fruits and vegetables, (such as carrots, apples, celery, oranges, etc), whole grain cereals, and whole grain breads.

■ Issues:

- Constipation: a frequent problem..can be regulated with dietary measures
- However, Miralax or other agents may be indicated
- Butyrate is produced in the large colon as a product of the digestion of fiber - unknown if this is of additional benefit in SMA patients

■ Vitamin Supplements:

- No substantiated benefit to consuming > RDA for any vitamins/minerals – ? theoretical benefit to increasing methylation capacity (i.e. B12 and folate ???)
- Excess supplementation can cause undue stress on the liver and kidneys; some vitamins can be toxic to nerves in excess: i.e. vitamin B6 (pyridoxine).
- Formulas (Ensure, Pediasure, Tolorex, Vivonex) contain vitamins and minerals, and can function as a multivitamin supplement.
- Carnitine levels (important in fatty acid oxidation) are sometimes low in SMA patients, and could contribute to muscle weakness. If meat intake is inadequate, levels should be monitored.

■ Drug – Nutrient Interactions:

- Valproic Acid can interfere with the absorption and utilization of certain nutrients.
- If taking valproic acid, it is important to ensure that your child is receiving the RDA of vitamin D, vitamin K, vitamin B-6, vitamin B-12, folate, and calcium.
- Valproic acid depletes carnitine; any SMA patient on this medication for any purpose should receive carnitine of 50 mg/kg/day up to 1 gm/day, with regular monitoring of levels

■ Recommended Daily Allowances for Selected Nutrients

Age	Vit D	Vit K	Vit B-6	Vit B-12	Folate	Calcium
1-3	5 mcg	30 mcg	0.5 mg	0.9 mcg	150 mcg	500 mg
4-8	5 mcg	55 mcg	0.6 mg	1.2 mcg	200 mcg	800 mg

- Gastroesophageal Reflux Disease:

Frequent in SMA patients: SMA 1 > 2 > 3

- To ameliorate reflux:

- Eat multiple small meals throughout the day (6 or more)
- Eat a low fat diet. High fat diets tend to relax the lower esophageal sphincter, and make reflux more likely
- Avoid lying down after eating, or elevate the head of the bed/seat to at least 30 degrees
- Avoid acidic foods (soft drinks, citrus, tomatoes, etc)
- Avoid spicy foods, chocolate, peppermint

Gastroesophageal Reflux Disease:

- Reglan increases speed of gastric emptying and gastrointestinal movement
- Acid blocking agents are commonly utilized to prevent esophageal damage and food aversion
- Nissan (surgery) is sometimes needed, especially in type 1 children
- A g-tube and Nissan don't necessarily have to be done together, but newer laparoscopic techniques make this feasible if indicated, with faster recovery time

■ Elemental Amino Acid Diet:

- There is no *proven* benefit to taking formulas high in elemental amino acids for SMA patients
- Adult patients with gastrointestinal diseases **on parenteral or enteral nutrition** have shown a reduction in bacterial infections, and a healthier gastrointestinal tract when taking supplemental glutamine (nonessential amino acid)
- Glutamine is the preferred fuel for the cells in the gut, and helps maintain gut integrity
- Adults have shown no adverse affect with doses up to 20 g a day. However for children, a reasonable amount is 200 to 300 mg/kg/day, no more than 10 g/day

■ Abnormal fatty acid metabolism:

- Patients with more severe forms of SMA tend to exhibit abnormal fatty acid metabolism.
- Abnormalities are present in both catabolic (associated with fasting or illness) and non-catabolic states, manifest as an elevated C12:C14 ratio.
- Higher C12:C14 ratios may be associated with a NAIP deletion???
- Patients with SMA may have reduced mitochondrial DNA, contributing to a defect in energy metabolism

Crawford T, et al, *Abnormal fatty acid metabolism in childhood spinal muscular atrophy*. Ann Neurol 1999 Mar;45(3):337-43

Berger A, et al, *Severe Depletion of mitochondrial DNA in spinal muscular atrophy*. Acta Neuropathol 2003,105: 245-251

■ Bone Health

- Special issues for SMA patients include increased risk of osteoporosis, fractures, and complications re: scoliosis surgery
 - Weight bearing, from a young age, via whatever means possible, is very important and can delay scoliosis
 - Regular exercise
 - Adequate intake of calcium and vitamin D
 - ? Role of medications to increase bone density: should they be standard of care for SMA patients? Definitely should be considered prior to scoliosis surgery, or in light of history of fractures

■ Obesity is a serious health concern

- Greatly increases burden of care
- Decreases quality of life and self image
- Makes physical activity more challenging
- Increases risk of diabetes and hypertension
- Increases pain and associated complications with hips, back
- The time to ensure a lifetime of good dietary habits is as early as possible

■ Management of diet during illness

- SMA children frequently demonstrate significant loss of strength during illnesses. Problems include: significant weight loss, loss of muscle mass, metabolic acidosis, and rapid progression of contractures
- Tendency for getting in trouble is clearly related to a child's reserves..ie severity of muscular atrophy increases risk of serious problems and regression of functional abilities with illness

- Management of diet during illness....
empiric guidelines

- If illness is gastrointestinal, with vomiting or diarrhea, early presentation for IV fluids is indicated (?? 8 to 12 hours maximum). SMA 1 babies and weak SMA 2 children stand to benefit the most from rapid intervention.
- Amino acid supplementation (TPN) as opposed to just glucose, is indicated in any circumstance in which fasting or inadequate caloric intake is expected to continue for any period of time. At most hospitals, it takes up to 4 to 6 hours to fill a TPN order.
- Children with SMA who are ill should not fast more than 12 hours without supplementation in some fashion

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