

Getting the Most Out of What You Eat!

Nutrition for Oral Feeders

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Make today a breakthrough.



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SMA and Nutrition

- There is little published research on optimal diet for SMA.
- Knowledge based on:
 - professional experience
 - extrapolation from other diseases
 - listening to parents/caregivers/patients.
- Every person with SMA is different!

SMA and Nutrition

- Developing a nutrition care plan: what goes into the pot?



Why is Good Nutrition Important?

- Enables growth in children or weight maintenance for adults
- Optimizes motor and pulmonary function
- Prevents illness and/or speeds recovery
- Improves quality of life
 - Prevention/healing of pressure sores
 - Energy
 - Healthy hair, bones, nails
 - Enjoyment!

A dietitian who is knowledgeable about the special issues that people with SMA face can help optimize an individual nutrition plan – because one diet does not fit all.

Your SMA Dietitian Will Evaluate...

- Growth (for children); weight (for adults)
- Dietary intake and quality
- Intake of supplements
- Nutrient deficiencies/excesses
- Nutritional lab values
- Medications
 - drug nutrient considerations
 - those that affect nutrition

Recommended Nutrition Labs

- Complete blood count (CBC) & iron
- Liver function
- Vitamin D
- Essential fatty acids, if following a low-fat diet
- Protein status (amino acid profile, albumin, pre-albumin)
- Minerals (magnesium, sodium, potassium, phosphorus)

Basics of Good Nutrition

- Calories
- Protein
- Fat
- Carbohydrate
- Vitamins/Minerals
- Fluid

Calories

- Not too many, not too few
- People with SMA need less
 - less muscle mass
 - decreased movement
- Very individualized and can change over time
 - Physical therapy, adaptive sports, surgery, illness, assistance with breathing
 - Spinraza may increase needs
- Sitters and walkers are at risk for overweight as they get older.
- Weight loss can be difficult – prevention is the best strategy!

Protein

- How much is enough?
 - Goal daily grams of protein~ ½ to 1 gram for each pound of body weight
 - Example: 30 lb. child needs about 15 – 30 grams of protein
- Good sources of protein are:
 - Eggs – 6 grams in 1 large egg
 - Low-fat dairy products – 8 grams in 8 oz. milk
 - Lean meats, poultry, and fish – 21 grams in a piece the size of a deck of cards
 - Even grain products and vegetables contain protein!
- Adequate protein will promote growth and help to minimize muscle breakdown and loss
- It will not build more muscle and make people with SMA stronger.

Protein

- Some people choose to follow a vegan (plant based) diet and get protein from:
 - Nuts, nut butters
 - Beans and legumes
 - Vegetables
 - High protein grains (quinoa, bulgur)
 - Special formulas
- If this is a diet you want to try, please work with your dietitian to make sure you/your child is getting what they need for vitamins and minerals and for growth.

Fat

- A moderate fat diet can:
 - Improve stomach emptying
 - Reduce reflux => reduce risk of aspiration
- Limit high fat foods such as:
 - Deep-fried foods
 - Fatty cuts of meat (harder to chew anyway)
 - Full-fat dairy products**
- Choose healthful fats to meet essential fatty acid needs such as:
 - Fatty fish – salmon, halibut, tuna
 - Avocado (healthy source of fat but not high in the essential fatty acids)
 - Hemp milk
 - Plant-based oils – walnut, canola, flaxseed, etc.

**Many young kids with SMA who eat orally struggle to gain weight. It may be necessary for your child to eat full fat dairy like whole milk, cheese, cottage cheese, and whole milk yogurt until weight gain improves.

Carbohydrate

- Choose complex, high fiber carbs like whole grain breads, cereals and pastas, brown rice, beans and legumes, fruits and vegetables.
- Limit processed and refined sugars (soft drinks, candies, desserts, etc.)
- Watch portion sizes – ½ cup of fruit or vegetable and 1/3-1/2 cup starch per meal (even less for a toddler)

For some people it looks like this...



Fluid

- Amount needed depends on body weight, age, condition
 - Adults: ~ ½ oz. per pound body weight.
 - Children: ~ 1 ½ oz. per pound of body weight.
- May need more if ill or has increased secretions.
- Water, flavored waters are best.
- Less than 1 cup of juice per day.
- Avoid soda, juice drinks, and other sugar-sweetened beverages (these are empty calories that don't help with fullness).

Vitamins/Minerals

- Best source is from healthful foods!
- Your dietitian can help you identify any nutrient deficiencies and make suggestions for supplements, if needed.
- Excess supplementation can cause undue stress on the body.
- Commercial formulas contain vitamins and minerals and must be factored into daily total.

Supplements

- Be sure to tell your healthcare provider about any supplements you give your child:
 - multivitamin and mineral supplements
 - carnitine
 - omega 3 fatty acids
 - probiotics
 - herbal and homeopathic supplements
 - antioxidants
 - anything else that you give your child....

Common Nutrition Concerns

- Growth
- Feeding Concerns
- Fasting and Illness
- Drug-Nutrient Considerations
- Constipation
- Reflux

Growth

- Weight
 - There are no growth charts specific to SMA.
 - We often rely on a physical exam and look at patterns on the growth chart.
 - Rate of growth and goals depend on severity of disease.
 - Due to reduced muscle mass, weight may be at lower percentiles on the growth charts, especially at a younger age.
 - Concern for obesity with aging.
 - The more strength a person has, the more similar growth will be to those without SMA

Feeding Concerns

- Swallowing problems
 - Signs: coughing, choking, pocketing, and noisy breathing with feedings
 - Some people have trouble swallowing thin, clear liquids
 - Video swallow study can assess for aspiration
- Chewing problems
 - Hard to chew foods:
 - Large chunks of meat and/or tough meats
 - Raw fruits and veggies
 - Sticky foods like peanut butter and thick cheese spread.
 - Rather than eliminating foods, modify food and beverage textures:
 - Smoothies with fruits and veggies
 - Cooked or pureed fruits and veggies
 - Meats cooked in a crock pot

Feeding Concerns

- Meal Fatigue and Early Satiety
 - Small, frequent meals can help
- Self-Feeding
 - Use adaptive utensils or plastic ware
 - Offer finger foods
 - Cut foods into small pieces
 - Straw cups or Camelbacks

Fasting and Illness

- Limit fasting during illness
 - Usually 8-12 hours for oral feeders
- The stronger the person is, the longer fasting can be.
- If well, under most circumstances, waking to feed overnight is unnecessary for children over age 1.
- We often recommend a snack with protein and carbohydrate before bed.
- During illness, please contact your medical care team.

Drug/Nutrient Considerations

- Keep your health care team up-to-date on medications :
 - Valproic acid (may cause weight gain)
 - Antibiotics (may cause diarrhea and/or GI distress)
 - Bowel regulating medications
 - Gut motility medications
 - Antacids/proton pump inhibitors
 - Vitamin/mineral supplements (separate calcium and iron)
 - Herbal supplements

Constipation

- Weak intestinal muscles → poor motility.
- Symptoms:
 - abdominal pain & distention
 - fullness and lack of appetite
- Manage with diet
 - increase fluid, fruits & vegetables & whole grains
- May need medicine.

Special Diets

- Many people with SMA do well eating orally and eat regular food.
- However, for some people, tube feedings may be needed to help them meet their nutrition needs.
- Different formulas can be used successfully.
- Is the amino acid diet beneficial for someone who is able to eat?

Amino Acid Based Diet

- Eliminates all protein from meat, dairy, and soy in favor of an amino acid based formula (Tolerex®, Vivonex® family).
- Formula is combined with juice, water, pureed fruits, pureed vegetables, oils, and vitamin and mineral supplements.
- Get other calories from oral vegan diet of grains, beans/legumes, fruits, vegetables.

Amino Acid Diet

– Pros:

- Incorporates fruits and vegetables which may help with constipation.
- Often lower in fat so may reduce reflux.
- Fat source is usually healthful oils rich in essential fatty acids.
- Some patients and caregivers feel it improves health and strength.

– Cons:

- May be unnecessarily restrictive by eliminating favorite foods
- Formula does not taste good – may cause oral aversion & weight loss
- May result in nutrient deficiencies if not monitored closely.
- Expensive if not covered by insurance.

Generally unnecessary for someone who can digest protein foods as amino acids are just fully digested protein.

More Research Needed

- What is the optimal oral diet for people with SMA?
- How will the new drug therapies affect nutrient needs?
- Preventing overweight/obesity in SMA.

Simple Rules for Good Nutrition

- 3 small meals and 2-3 small snacks per day
- Choose foods that have ingredients that you recognize and can pronounce
- Better yet, choose foods without an ingredient list!
- Make it a family affair
- Unless you need to gain weight, don't drink your calories
- Move as much as your ability allows

And finally...

- If your child doesn't like the healthy foods you serve...
 - try again tomorrow...
 - and again the next day
 - or present them in different ways
 - or give them cool names – be creative!
- Most importantly, don't give up and don't back down

Breakout Discussion

- Adults with SMA - Rebecca Hurst Davis
- Children with SMA – Erin Seffrood and Stacey Tarrant

SMA Conference Survey

Please complete your conference survey at this link:

<https://www.surveymonkey.com/r/2017SMAAnnualConference>

Or fill out the paper survey in your conference folder.

- **All participants who complete a survey by 10:30 am on Sunday July 2nd, will have their name entered into a raffle for a brand new iPad!**
- **The winners will be drawn and announced on Sunday, July 2nd at the Closing General Session/It's a Wonderful Life.**