

What is the Amino Acid Diet?

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This information is based on opinions and experiences compiled from parents of children with SMA and should not necessarily be relied upon as an alternative to medical advice from professional healthcare providers.

ABOUT THE DIET

The Amino Acid Diet is a nutritional approach that many SMA families believe has had a positive impact on their child's quality of life. Using this dietary approach, milk and soy based formulas are eliminated and substituted with a low-fat, elemental free form amino acid formula. Elemental formulas are made from nutrients in their most broken down form, which require very little digestion. Tolorex and Pediatric Vivonex, from Nestle Nutrition, are the two formulas used most often. EO28 Splash, from Nutricia North America, is a ready-made, more palatable, amino acid formula sometimes preferred by children without g-tubes. Because the EO28 Splash contains a higher fat content, it may not be an appropriate formula choice for children with Type 1 SMA. These formulas contain vitamins, minerals, carbohydrates, protein and some fat. The protein is in the form of free amino acids. These amino acids are not derived from food sources. Most parents that implement this diet believe more benefit is realized with these elemental formulas, rather than hydrolyzed amino acid formulas, which contain broken down proteins from foods, such as dairy or soy. The free form amino acids in Pediatric Vivonex, Tolorex, and EO28 Splash, are in balanced amounts, and are less likely to trigger immune responses such as inflammation. They are easily digested and quickly absorbed into the bloodstream. When given frequently, in low amounts, these amino acids provide a steady source of accessible energy and may help prevent muscle breakdown.

Many children with SMA are very reactive to the protein and fat in dairy and soy formulas. Their reactions are often attributed to their SMA progression. However, when this protein and fat are replaced with free form amino acids and appropriately reduced amounts of fat, children with SMA experience improved respiratory health and fewer metabolic complications during illness. Most children with Type 1 SMA have an immediate reduction in airway secretions. Constipation, which can be a major complication, is more manageable or even eliminated. Some children have improvement in strength and regained function.

Parents using these formulas vary their child's particular diet based on tolerance, age, weight and length, and severity of SMA, but there are some basic guidelines that most children respond to favorably.

Children with SMA do not tolerate large amounts of amino acids or fat due to their reduced muscle mass. This reduction of muscle mass prohibits the proper absorption,

storage and utilization of amino acids and fat, which could result in toxic accumulation in the bloodstream.

- More than approximately 2 packets of either formula, over a 24 hour period, may cause increased heart rate, arrhythmia, and/or gastrointestinal distress.
- % of fat calories should be kept at the minimum requirement necessary for adequate essential fatty acid intake
- To avoid fat, protein, and calorie overload, these formulas should never be combined with soy or milk based formulas.
- Children should be monitored with the following laboratory testing: quantitative amino acid profile, essential fatty acid profile, complete metabolic profile

ADDITIONAL SUPPLEMENTS

Because these formulas aren't used to supply complete nutrition in SMA patients, additional calories, vitamins and minerals must be added to the diet to ensure that their daily requirements are met. Special attention should be given to calcium, magnesium, sodium, and potassium intake.

Commonly added supplements include:

- Multi-vitamin and mineral— NanoVM from Solacenutrition.com (may only require half-dosage)
- Additional Vitamin D3—this level should be monitored for adequate dose
- 1/8 to 1/4 teaspoon salt
- Probiotic—Floragen 3
- L-glutamine powder—(200-300mg/kg/day, not to exceed 10gm.)

Tolerex contains 1060mg/pack, Pediatric Vivonex contains 774mg/pack, EO28 Splash doesn't contain L-glutamine

Glutamine lab level should be approx. 500 on quantitative amino acid profile.

- Based on lab reports, the amino acids L-carnitine and L-arginine are often supplemented.
- Additional essential amino acids should be added as indicated by a quantitative amino acid profile.
- CoEnzymeQ10

Although most children seem to tolerate Tolerex best, parents must add LOW amounts of essential fat to this formula. To accomplish this, there are several different options successfully used by parents. Over the last couple of years, it has become increasingly common for parents of children with Type 1 SMA to add breast milk to their children's Tolerex. For many, this has made a tremendous difference in their children's growth and wellness, especially their gastrointestinal health. Based on their essential fatty acid profile lab reports, approx. 8-10 oz/day of additional breast milk meets the children's essential

fatty acid requirement. More breast milk is sometimes given when an extremely reactive child cannot tolerate other calorie sources, such as baby foods and juices. If a child's mother can't supply breast milk, many families report a positive experience acquiring it from local donors, signed off as healthy by their doctors, or donor milk banks. For more information go to milkshare.com and [eats on feets on facebook](https://www.facebook.com/eats.on.feets). There are many SMA caregivers willing to share information on acquiring breast milk from donors. Questions can be posted on SMA family on facebook, SMASpace.com or SMAsupport.com.

If breast milk is not an option, a commonly used fat supplement is 1/4 to 1/2 teaspoon of safflower oil added to the Tolorex mixture. Some children also tolerate 1-2 grams of evening primrose oil and/or 1-2 grams of an Omega 3 supplement. Some parents find that a combination of Tolorex and Pediatric Vivonex works well for their child. *It can't be stressed enough that all fat supplements should be added slowly, in very low amounts, to check for tolerance.* When used exclusively, Pediatric Vivonex and EO28 Splash should contain ample fat for most children.

Almost all dietary fat is used by muscle. Because SMA patients have very reduced muscle mass they have an extremely low tolerance to dietary fat. When the percentage of calories given as fat exceeds 10%, many children with SMA experience flushing, high heart rate, increased secretions and delayed stomach emptying with reflux and vomiting. *Because our children experience improved health and quality of life on a VERY LOW FAT diet, it is necessary to verify that their essential fatty acid requirement is adequately supplied. This should be monitored with an essential fatty acid profile laboratory test.*

INSTRUCTIONS ON ADMINISTERING DIET

These formulas should be diluted as much as possible (more than directed) with water and other fluids, such as breast milk, juices, rice milk, almond milk, and fruit and vegetable baby foods. These will also increase calories, potassium, and other nutrients. 16 ounces of fluid for each packet of formula is a well tolerated dilution for most g-tube fed children. Those drinking their AA formula should dilute as much as possible with water and juices. Adequate hydration is extremely important, and recommended daily intakes should be met.

Additional Food:

In addition to the elemental formula mixtures, fruits and vegetables should be consumed. Those with Type 2 or 3 SMA usually tolerate some grains. The formula, fluids, supplements, and fruit and vegetable baby foods should be mixed in a blender and refrigerated. Because children with Type 1 SMA are often extremely reactive to changes in their diet, foods and supplements should be added gradually, one at a time, to check for tolerance.

Children receive the most benefit from this diet by greatly reducing or avoiding animal protein foods. Children following the AA diet correctly are receiving adequate protein from the amino acids in their elemental formula, and can easily be overloaded if they are

also eating high-protein foods. Most children tolerate 1gm/kg to 1.5gm/kg of amino acid protein/day. They should never be fed high amounts of amino acids over a short period of time.

CLOSING

Although children are affected by their SMA differently, the recommendations stated earlier have proven to be the safest and most effective way to implement the AA diet. Parents must take responsibility for monitoring and problem solving when placing their child on this diet. Unfortunately, due to the lack of study on nutrition in SMA, many medical professionals are unfamiliar with this dietary approach and often overload our children with fat and/or amino acids. Networking with other SMA families that are successfully using the AA diet and following the above guidelines can be an invaluable source for information on the benefits and proper administration of this diet.

Personally, I have had success with this diet for my daughter Krista, a lovely 21-year old, who was diagnosed with SMA Type 2 when she was 8 months old. We placed Krista on this diet over 16 years ago and have been very pleased with her wellness and stability. There are many other children with SMA that have also experienced more quality and length of life from following the AA diet as stated above. The feedback from their caregivers has played a significant role in our ability to adjust and improve the AA diet. I would like to thank all of you who have contributed by sharing your experiences and knowledge over the last 16 years. It has been a joint effort! A special thank you to Jeanna Huetter, who contributed greatly to the revision of this paper.