

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. This diet has not been studied by the scientific community and is not endorsed by Families of SMA.

ABOUT THE DIET

The Amino Acid Diet (AAD) is a nutritional approach that many SMA families believe has had a positive impact on their child's wellness and stability. Using this dietary approach, milk, soy, and most other food proteins are eliminated and substituted with an elemental free form amino acid formula. Elemental formulas are made from nutrients in their most broken down form. This form requires very little digestion. Tolorex and Pediatric Vivonex, manufactured by Novartis Nutrition, are the two formulas that are used most often. Both formulas contain vitamins, minerals, carbohydrates, protein and a small amount of fat. The protein is in the form of free amino acids. A free amino acid is not derived from a food source and is in its simplest and purest form. Parents that use this diet believe more benefit is seen with free form amino acids, rather than hydrolyzed amino acids, which are broken down proteins from foods, such as dairy or soy. The free form amino acids in Pediatric Vivonex and Tolorex are in balanced amounts, and will not trigger immune responses such as inflammation. They are easily digested and quickly absorbed into the bloodstream.

Many parents believe their children with SMA are extraordinarily reactive to the proteins in dairy and soy products. Their symptoms are commonly attributed to their SMA progression, yet when these proteins are eliminated and substituted with free form amino acids, most children, with all types of SMA, have experienced improved respiratory health and fewer complications from illness. Most children with Type 1 SMA have an immediate reduction in airway secretions. In addition, constipation, which can be a major complication, is more manageable or even eliminated. Some children have improved strength and regained function.

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

- Although these formulas can be used to provide complete nutrition, many SMA families use approximately 1/3-1/2 of this amount in order to avoid amino acid overload. *Children with SMA do not tolerate large amounts of amino acids due to their reduced muscle mass. This reduction of muscle mass prohibits the proper absorption and storage of amino acids resulting in possible toxic levels in the bloodstream.*
- More than 2 packets of either formula, over a 24 hour period, can cause an increase in heart rate, other cardiac problems and/or gastrointestinal distress.
- To avoid protein and calorie overload, these formulas should not be given to a child still on soy or milk based formulas.

The idea is to replace those food proteins with the free form amino acid protein found in the Tolorex or Pediatric Vivonex.

ADDITIONAL SUPPLEMENTS

Because these formulas do not provide complete nutrition, additional vitamins and minerals must be added to the child's diet to ensure that the RDA's are met. Special attention should be given to calcium, magnesium, and potassium intake. Other commonly well-tolerated added supplements (*by most children*): vitamins and minerals, acidophilus (dairy free), L-glutamine powder (dosage is 200-300mg/kg/day), (the formulas contain 1060mg's in one pack of Tolerex and 774mg's in one pack of Pediatric Vivonex), L-Carnitine, and Coenzyme Q10. Sometimes, small amounts of additional essential amino acids, in free form, are used by older children.

Although children with SMA do not require as many calories as active children without SMA, the amount of formula used is inadequate to meet their caloric need. If using Tolerex, parents sometimes add very small amounts of extra fat to ensure that adequate essential fats are given. A commonly used fat supplement is 1/4 to 1 teaspoon of safflower oil added to the daily formula mixture. Some children do well with 1-2 grams of evening primrose oil per day. Any fat supplements should be added gradually over a period of days, as some of the most affected children may experience slow stomach emptying and gastric distress. The Pediatric Vivonex contains ample fat for most children.

INSTRUCTIONS ON ADMINISTERING DIET

The Formula:

These formulas should be diluted as much as possible (more than the directions call for) with water, juices or enriched rice milk. The benefits of using juices or rice milk include not just the improved palatability, but the additional calories, potassium, and other nutrients they provide. A well-tolerated dilution is approximately 12-20 ounces of liquid for each packet of formula, depending on whether the child is G-tube fed or drinking the formula. To ensure adequate hydration, extra fluids should be given.

Children that are exclusively G-tube fed should have the formula administered by a continuous drip or given in small boluses. Some children without G-tubes drink the formula throughout the day. It should be diluted as much as possible, and divided into at least 3-4 servings.

Additional Food:

In addition to their Tolerex or Pediatric Vivonex, SMA children should consume fruits, vegetables, and those with types 2 or 3 SMA usually tolerate grains. Parents should use vitamins, minerals, additional supplements (discussed earlier), juices, and baby foods (fruit and vegetable) to meet their child's nutritional requirements. The formula, fluids, and any added supplements and baby foods should be mixed in a blender and refrigerated. . Since children with Type 1 SMA are often the most reactive to changes in their diet, foods and supplements should be added one at a time to check for tolerance.

Children receive the most benefit from the diet by greatly **reducing or avoiding** animal protein foods. This includes dairy products, eggs, and meat. A vegetarian diet is highly-recommended to see the most benefit. Remember, the children 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. For some children, this can interfere with improvements they may gain from this diet.

CLOSING

Personally, I have had success with this diet for my daughter Krista, a happy, vibrant 15-year old who has SMA Type 2. Through research, trial-and-error, we placed Krista on this diet over 10 years ago and have been very pleased with her wellness and stability.

Although children are affected by their SMA differently, the recommendations stated earlier seem to be the safest and most effective way to implement this diet. Parents must take responsibility for monitoring and problem solving when placing their child on this diet. Unfortunately, due to the lack of study this diet has received, most medical professionals are unfamiliar with this dietary approach and often overload our children with these formulas.

I encourage all parents to research and read the many informative nutrition books available at bookstores and libraries. In addition, networking with other SMA families that have successfully used this diet can also be an invaluable source for information on the benefits and administration of this regimen.

ADDITIONAL INFORMATION

Pediatric Vivonex and Tolerex are formulas manufactured by Novartis Nutrition. Ph# 1-800-333-3785. With a prescription, they are often paid for by insurance or Medicaid. Information on their home delivery program can be found at www.resource.walgreens.com.