

The Full Article on the Amino Acid Diet

Over the years, I have had many parents ask me about the diet I use for Hannah. I will state over and over that I learned this diet from other moms who have had great success with it. I hesitate a bit to write this diet information. All of the children with SMA are unique and different individuals just as we are all as individuals unique and different. There are some basic fundamental principles that should be followed if you decide to use this diet for your child in my opinion.

Elemental amino acid formula

Double dilution or more of the formula

Vitamins/Minerals

No meats

No dairy

No junk food (candy, soda, ice cream etc. etc)

Acidophilous

Because the children are so different in where their strengths and weaknesses lie, their formula recipe needs to be adjusted for accordingly.

I emphatically do not believe you must have a medical background or a college degree to feed a SMA child this diet. All that is needed is hope for your child and willingness to learn. I firmly believe this diet helps slow the progression of muscle loss and or stops the progression.

Typically children with SMA have inborn errors of fatty acid and protein metabolism. Most SMA children I know of who are not on the diet present with symptoms of malabsorption and Kwashiorkor . The children look bloated. Some children have fatty livers (as noted upon autopsy). One common mistake is for SMA children to be put on high caloric-high high-protein diets. This type of diet makes the children weaker. They can not handle the protein and tend to become fat from all the calories. They also become sick, get recurrent infections and usually end up dying. Unfortunately this happens all too frequently with SMA type one children. This diet is not just for type 1 SMA but all types.

Children with SMA have a lot of allergies. I am not sure why when tested lab results turn up negative to the many things they seem to show reactions to. The source of all allergens is protein. Dust, animal dander, plants etc. Allergens are caused by antigens floating around in the bloodstream. Dairy and milk proteins seem to be a major allergen for children with SMA. Other allergens are whey, soy, and oats. Side effects noticed are copious secretions, flushing, high fevers, bloating, weakness, general malaise, bronchitis, pneumonia and constipation and ear infections.

The beauty of the elemental amino acid diet is that you are giving protein in a broken down simplistic form. Whole proteins tend to be the problem. These proteins are not easily digested

or are not utilized somehow in SMA. This is also why when utilizing the amino acid diet you also must eat only fruits and vegetables to get the most benefit.

I have been asked who designed the diet for Hannah? I did! I began with the Tolorex and added one ingredient after another over a period of time to make sure no adverse side effects were noted. I learned the basics of the diet from other families of type 1 children who were using the diet and had success. Their children were living. I noticed the children who were on formula such as Similac, Pediasure, Progestimel, Scandi Shake etc. were not doing as well and becoming sick and / or dying.

Tolorex and/or Pediatric Vivonex does not give all the necessary vitamins, minerals or necessary fat according to the RDA for all age groups. This is why I add additional vitamins, minerals, evening primrose oil, safflower oil, vit e, calcium, magnesium, phosphorous, potassium, co Q 10, etc to Hannah's diet. I try to give most of the vitamins and minerals in amounts close to or a bit above the RDA. All one needs is a nutritional almanac to research this. I usually recommend the one written by Gayla and John Kirschmann. There are many books on the market. This book in particular is easy to read and readily available in most bookstores. The FSMA website has a few articles you can refer to under nutrition.

My opinion

In SMA you do not need a lot of protein for protein synthesis. A normal healthy child is recommended to get 2 grams per kilogram of protein a day. In SMA it has been my experience that less is better. 1 to 1 ½ grams per kilogram are sufficient. **SMA persons do not have the lean muscle mass that "normal" healthy persons do. This is why they do so much better when fed less protein. Due to the lessened muscle mass, this can cause problems. Vitamins and minerals can not be stored. The stores that they have are not stored long because they have nowhere to stay.**

To begin

Use 1 to 1 ½ packets of tolerex. Double dilute per package instructions. This is essential. You can dilute with juice and or water. I add even more water but you must double dilute otherwise the formula is too strong. Then you feed this mixture over the course of a day. Not to be given all at once. Add baby food fruits and veggies at meal times. No whole proteins like chicken or veal. No dairy. You can add vitamins and minerals. You can add safflower oil. Safflower oil is an essential fatty acid.

I mix the Tolorex in a blender with baby foods. This consistency is not to thick to place through a g-tube. It works quite well. The key is dilution. You must flush the g-tube after every feeding. If a continuous feed is given via pump then additional water can be bolused or added in over a 24-hour period of time. I have used both methods and both ways work well.

The danger of most physicians and dieticians who try to prescribe this diet is that they overload these children on amino acids. Four to five packets daily are prescribed. This is just way too

much and is dangerous! It can kill these children. SMA patients can not handle this much amino acids and or protein! I do not know of anyone who uses more then 2 packets a day. This is for children that I know of up to age 13. These children do not have the muscle mass to handle this much protein. You can not treat these children as if they are " normal". Another danger is that physicians and dieticians prescribe way to many calories. A normal healthy active moving child needs more calories. Typically 70-80 kcal/kg. In SMA, a good range is 60-70 kcal/kg. As long as the child grows and is gaining weight then they are doing well. Children do grow when on this diet and do gain weight. You can not base their growth on the charts. As you recall, the growth charts are for formula fed children not breastfed or Tolorex fed children. The main factor is they grow and gain weight. When they are fed to many calories they get fat and can not breathe or move as well.

For those who are breastfeeding. Keep it up. I still give Hannah breastmilk. I am a firm believer this is another reason she does so well. You can mix together the Tolorex and the breastmilk. (Do not mix breastmilk together in a blender). If a mom were still nursing I would encourage this. I recently had a baby. I am again pumping my milk for Hannah. When I was no longer able to breastfeed prior to the birth of my daughter, I got breastmilk from a Milkbank in North Carolina. (www.hmbana.org) There are six milkbanks in the USA. A doctor who researches SMA and nutrition has stated to me that when children with SMA who breastfeed stop nursing they go downhill and rather quickly. My belief is that it is because they are put on formula with dairy. Also, <http://www.hmbana.com/wsnAC28.html> is the website address of the milkbank I used for 4 years.

When beginning I recommend you start with just the Tolorex. The next ingredient to add would be glutamine. Start with 500 milligrams. Glutamine is the most abundant amino acid in the body! Typically SMA children are low in glutamine. A very conservative doctor who studies SMA and nutrition recommends 200 - 300 mg/kg glutamine. I have used additional glutamine with Hannah since she was one year old and have never had a problem.

Signs to look for are increased heartrate and vomiting. These are classic signs of protein toxicity (to many amino acids). I have not ever had this problem. Begin slowly and work up the amounts of amino acids. You should be OK. Now that I have said this, I give no 100 percent guarantee. I just believe you will not have any problems if you do it this way.

The next ingredients to add would be up to the mom. Usually calcium and the vitamins are a good place to start.

I also highly recommend you begin giving acidophilus to counteract the yeast problems so many SMA persons have. I have never ever heard of any problems from giving acidophilus only great results. I use Natures Way Primadophilous for children. It is found in the refrigerator at the health food store here. Read the labels. A lot of the acidophilus has dairy proteins in it, especially, the liquid ones.

In SMA, there is a lot of loss of water via breathing fast, open mouthed, and sweating. I highly recommend you give lots of water for hydration. Secretions should be thin not tacky. The

recommendation for a 1 - 3 year old is 115-125 cc/kg of water. I give Hannah the amount a younger child should get. Hannah almost age five gets 53 or more ounces a day of liquids in her diet consisting of juice, water and / or breastmilk. On days where she is hot and sweats, I give her more water.

It has been proven that children on this diet do not get scoliosis like other SMA children. In some instances, there has been a correction of the degree of curvature of the spine. I know of one example where a little girl with type 1 went from a 33-degree curve to a 21-degree curve with early detection and proper bracing! Some SMA patients have begun this non-dairy elemental amino acid diet when having been on a dairy or whey or soy formula diet and actually had dramatic improvements in curvature of the spine as well.

Currently I add the following to Hannah's diet in addition to the Tolerex.....

L-glutamine
L-valine
N-acetyl-cysteine
L-isoleucine
L-leucine
L-phenylalanine
L-arginine
L-carnitine
Co enzyme Q10
Creatine
B12 (oral and b-12 sublingual drops)
Folic acid
Copper
Folinic acid
Methylcobalamin
Vitamin E
Vitamin C
Phosphorous
Calcium
Magnesium
Potassium
Evening Primrose oil
Multivitamin

Other things I have tried.....

I have given Hannah flax seed oil. I thought it would be great. She had almost immediate respiratory problems, became cyanotic (blue lipped) and had secretions the rest of the day even though she only had one feeding of this. Same thing happened with fish oil. This was my experience.

If you really are going to use this diet, I recommend you have labwork drawn regularly (fasting

quantitative plasma amino acids, basic metabolic panels, mg, zinc, phos, creatine). When fasting the children get their morning meal and then fast 6 hours. The lab results always come back for Hannah saying within normal limits. However, if a biochemist reviews the results it is noted otherwise. The biggest problem most often seen is metabolic acidosis!

I send the lab results to a biochemist in Canada at Nutrichem (www.nutrichem.com). I speak to Kent MacLeod. He mixes more amino acids to add to the tolerex food mixture I give Hannah based upon HANNAHS bloodwork. I just feel this works best for me. I am not good at guessing and want to try to do what I think is best for Hannah. Most of the families who use the amino acid diet with their children just guess and add more amino acids and supplements. I do not feel comfortable doing things this way. Again, my medical background is a strong factor in this area. I do it this way. I am the only mom I know who does this. All the other moms just guess. This is ok for them. They add glutamine, arginine, creatine, some branch chain amino acids (leucine, isoleucine and valine) All this "stuff" is what I give Hannah extra of from my mix made in Canada. What can I say? It still is a guessing game however. Noone truly knows all the ins and outs of SMA. I usually follow the recommendations of the biochemist in Canada. He however is also no "expert" in regards to SMA. I have found Hannah gets weaker sometimes with the mix I give her so I slowly work up the mixture of which I give her until I am at the recommended dosage of the biochemist. To much protein? Over time Hannahs body adjusts and she is actually **stronger** then weaker. All the children are different and their individual diet has to be made for them. The basics that are important are the elemental amino acids, essential vitamins and minerals, no dairy, whey, soy or dairy derivatives like casein, casienate etc. These all cause weakness in SMA.

Over the years I have known many parents from all over the world and have talked with them regarding this diet. Some have used this diet and some have not. There are families who would give their left arm to know all this information I have just told you. Those that do say they wish they had known of it earlier. It is best to begin this diet when the child is young so that muscle wasting can be spared. In my opinion why not try the diet? You have everything to gain and nothing to lose.

Unfortunately, diet alone is not enough when it comes to SMA. Aggressive respiratory care is needed as well. I again refer you to Dr. John Bach. The children who follow this diet and his respiratory care protocol tend to live longer and healthier then any other SMA patients in this country!

If you would like I can refer you to parents who use this diet and will tell you the dramatic differences they have seen in their children since beginning this diet.