

## **Families of Spinal Muscular Atrophy**

### **Transcript of SMA Diet and Nutrition Chat, Wednesday, September 6, 2006**

This transcript has the proper question and answer put together for easier reading, and some of the questions and answers have been cleaned up, and some extra talk and some identifying information has been removed. Typos and errors may still exist although it has been reviewed by our expert.

*All answers posted are the professional opinion of Gail Wiebke, M.S., R.D.. Families of SMA neither supports nor rejects her advice and recommends that patients consult their own personal physicians and/or nutritionists before making changes to any existing diet or nutrition plans.*

*administrator* : Hi everyone and welcome to the FSMA chat room. Today's chat subject is SMA - Diet and Nutrition, and our expert is Research Dietician Gail Wiebke of Salt Lake City, Utah. Users are welcome to chat back and forth and offer their suggestions - just be sure to leave space for our expert to answer questions. If you miss anything, a transcript will be available later. If there are many questions, we ask everyone to ask just ONE question so everyone has a turn.

*GailW* : Thank you! I am glad to be here!

*B* : Hi Gail. Do you have any nutritional recommendations for Type 1 children with slow gastric emptying?

*GailW* : Slow gastric emptying is the kind of question I like to refer to the doctors!! It is common in kids with SMA and there is not always a dietary solution

*B* : Thank you.

*S* : Hello, My son is type 2, I just read the past chats and wanted to know if it true that folic acid has some benefits in SMA kids. What is the recommended amount of folic acid in a two year old?

*GailW* : S, what a great question! Folic acid is so interesting. Sure Folic acid has benefits for everyone, how much SMA kids need is less well known. They need to meet the RDA, but after that is tricky. Folic acid doesn't work alone so you can't give them high doses.

*D* : Hi Gail. I am a Brazilian student (nutrition course). Could you explain about energy requirements for SMA patient type I?

*GailW* : Energy requirements for SMA is another great question! You have to check their weight to really know what it is but you can start with 9 - 11 calories per cm height which is an amount based on research in a similar population, children with spina bifida.

*D* : Thank you

*P* : I've read that children with SMA should not lose weight, but what if you have a child that is significantly overweight? It hampers her mobility and puts her at risk of diabetes, which runs in my family. What do you recommend for an overweight child with SMA III?

*GailW* : P, next to kids who are underweight the biggest problem is kids who are overweight! Start with the recommendation for calories by height and then go from there. Learn to read labels carefully and keep track of what they are eating for a few days so you can record the calories. You don't need to have them lose weight, you need to keep them at the same weight and let them get taller!!

A: For a SMA1 child 5 years old, what are the clinical symptoms of kidney fatigue caused by an over-assumption of proteins? Which is the difference between formulas as Nutrini by Nutricia 1 ml= 1 cal and formulas like Tolerex or Vivonex?

*GailW* : A high protein diet can lead to kidney overload, especially in small children whose systems are less developed and who already are taking medication of some kind. Formulas differ in the source of the nutrient being provided. They have different sources of protein fat, etc. They are all required by law to provide certain necessary nutrients

S : Is it bad for SMA children (type 2) to take carnitine over the counter? What is the function of carnitine if taking alone?

*GailW* : S, Carnitine over the counter is not the same as by Rx, not as pure, possibly not as safe.

S : Thank you

H : Hi Gail. In regards to the Tolerex and Vivonex amino acid formulas, are you and Dr. Swaboda recommending them to SMA patients at this time or not? Also, when might we begin to see a nutritional study in regards to diet and these formulas in SMA patients?

*GailW* : Let me answer the amino acid question first. I neither recommend the AA diet nor doubt its benefits. The benefits reported by parents are: less secretions, less sweating and less suctioning. The cons: it should be used very carefully! It tastes terrible and separates what the SMA child eats from what the rest of the family is eating, it doesn't promote GI motility and can lead to loss of some GI function if the child has a normally functioning system due to a decrease in the amount of digestive enzymes produced .... so I am probably more con than pro but always happy to help parents who want to follow it!

B : I have heard that these children should stay at the 20% mark for weight, although my daughter is at the 50th. Should she lose weight or is this ok?

*GailW* : B, great question on the percentile. The 50th is a nice number - unless your child's weight has started to interfere with her functioning. In general I prefer the 20th so they have more "wiggle room" with growth and weight changes. Just keep a close eye on her weight and try to keep it on a curve that is normal for her.

P : My daughter is 13, weights close to 200 lbs, and has been 5 foot tall for the past two years. She seemed to stop growing taller when she stopped walking. When she was younger I had trouble finding pants that would stay up on her because she was so thin, but as her disease progressed she started gaining weight and we haven't been able to get it under control. We have tried Weight Watchers and the South Beach Diet, and while my husband and I lost several pounds, my daughter only lost 1. Her weight has become a big problem, making it difficult for her to turn over in bed, and for me to do transfers. I've read in Quest magazine that some people have had some success with some prescription weight loss drugs. Would you recommend them?

*GailW* : P, no, no weight loss drugs and no diets! How strange for a dietician to say! But I would rather have them eat the food they like, in small enough portions to keep their weight on the low side. It's so much harder to do than to say and as you noted takes the whole family. Carefully monitor the amount of calories she gets. This may be very different from what you and your husband need

S : Is there some food that we have to avoid for SMA type 2? I'm not just talking about sugars and calories, but about too much protein, how much is too much?

*GailW* : S, right now I recommend using 1 gram of protein per kg body weight per day up to 2 grams

per kg bw. That should meet their needs and not be too much.

*K* : What is your opinion on combining AA diet with other formulas - milk or soy based?

*GailW* : *K*, about combining AA diet with other formulas, be careful not to go over on total protein. Also, purists would say if you don't follow the diet exactly you might not see the whole benefit.

*S* : I'm sorry but how do I measure protein in a glass of milk? or eggs? My son weighs 14 kg.

*GailW* : *S*, check the labels but milk has about 7-8 grams in 8 fl ounces and 1 egg also 7-8 grams. That would about do it for a day for him and there is protein in everything but fruit.

*S* : Thank you very much for your answers - I just went to read the label on the milk gallon!

*M* : Hi Gail: so you are saying that Dr. Swoboda is not having a trial on the amino acid diet and its benefits. The amino acid diet is used by a lot of Type 1 and some Type 2 Children but there has been no clinical studies done on it.

*GailW* : *M*, thank you for your question about the AA diet and possible trials. Right now, no, there are no trials ready to go on the diet but for a very good reason! One, if a child is on a diet trial they would not be eligible to participate in a trial that might be more beneficial, namely a drug trial. That would not be ethical. Also, diet studies require a lot of participants, out of the total pool of SMA patients we would be taking away from kids who could be in drug trials, It's a very tricky business deciding which trials are the most important to do and hotly debated among researchers. Remember I am speaking for myself only here, not Dr. Swoboda.

*M* : Thank you.

*D* : An about high triglycerides levels in Type I?

*GailW* : *D*, children with SMA do often have unusual tg levels or lipid levels, exactly what to do about it is the thing. We know their metabolism is different; probably how they use fats is different which is why I recommend a healthy diet level of 30% of calories from fat and not more. In the future researchers hope to be able to say more about which type of fat to provide or whether 30% is too high but it not for sure yet. They need glucose for their muscles to work not fat. Stay tuned to upcoming research

*D* : Thank you

*B* : What are the fluid requirements for children with SMA?

*GailW* : *B*, fluid requirements, another great question! Enough so their urine is pale yellow. I start with 115 - 135 ml per kg bw, but that can be a lot of urine and a lot of work for parents. Give them small amounts frequently so they have time to use it before they excrete it!

*P* : Thanks for your help. I have one more question... You said no diets... Since my daughter's insulin levels are elevated her doctor has recommended a diabetic diet to help bring it back in line. When you say no diets, I assume you are concerned that it might affect the progression of the SMA. Is it safe for her to follow a diabetic diet? Are there warning signs that I should watch for? Thanks again for all your help.

*GailW* : *P*, what I meant by no diets was that I don't like to be put on a diet with unusual foods or strict guidelines so I can't recommend it for kids. I'm a picky eater. It is absolutely safe to follow a diabetic diet! Whole grains, whole vegetables, whole fruits, excellent! You should also know that while I don't follow a diet, I don't eat fast food and don't recommend anyone else does either. No hot dogs ever. And only one donut every 5 years. No I'm not kidding. That's how you limit calories

P : Gail, thanks for all your help.

B : Regarding body fat, you say 30% of calories. What is the calculation to determine grams?

GailW : **B, there are 9 calories in every gram of fat!**

B : Thank you.

D : Gail, can you give more information (articles, etc) about the use of equations for estimation energy requirements for SMA patient?

A: As D, I would like to read some articles

GailW : **Here's the reference I use for a place to start with caloric requirements for children with SMA.**

**Ekvall SW, Bandini L, Ekvall V.: Obesity. In Ekvall SW (ed): Pediatric Nutrition in Chronic Diseases and Developmental Disorders, Oxford University Press, 1993; 168.**

**Children with SMA are similar to children with spina bifida in that there is a loss of muscle control so after much searching and discussing with colleagues decided it would be the best place to start with estimating how many calories might be needed. Then calories can be adjusted according to weight gain or loss.**

*Rounds of thanks from questioners*

*administrator* : Thank you so much, Gail, for answering our questions!

*GailW* : **Everyone, you're welcome! and good luck!**

**The following questions were submitted by people who were unable to attend the chat.**

W : I had been a faithful milk drinker from as far back as I can remember (since birth). I drank a minimum of one glass a day (6-8oz approx) up until my mid 30's. At the age of 39 I was prescribed Mestion to aid in giving me some of my "mojo" back. I was weakening and lost the ability to bend my left forearm inward and a few other moves I took for granted. Now in the last two weeks, along with my Peptinex by Novartis I am drinking (almost forcing myself) one glass of 2% milk again per day. What I am feeling is more confidence when moving not strength or regained movement.

My question is could milk "maintain" motor neurons anyway? I wonder if I had kept up my one glass a day would I still be faced with the same loss of abilities? I don't eat meat or eggs or fish or cheese, milk was my sole source of protein. Just how important is milk to a woman's diet who has SMA II and is 41 years old? I guess it won't hurt me eh?

*GailW* : **Milk can only maintain motor neurons in the same way that all foods do, you need food to stay healthy and maintain body functions and growth. You would most likely have faced the same loss of abilities whether you kept drinking milk or not, because SMA is not caused by dietary changes. Protein is provided not just in milk, eggs, meat and fish, it's also in vegetables, grains, nuts, soy, beans, any food that isn't a fruit, has protein in it. As humans age, many of them can not digest milk as well as when they were young due to a decrease in the production of lactase (the enzyme that breaks down milk sugar). This is normal aging. The symptoms of lactose intolerance are diarrhea, gas, bloating and abdominal pain. So, milk isn't quite so important as you age, but it sounds like it's very comforting to you. Continue to drink it unless it's causing you problems!**

*R* : I am writing from Poland so I am unable to participate in chat due to time difference. I am a mother of 10 month boy with SMA type I. So far we have managed to avoid a gastrostomy as he is still able to swallow and suck.

Right now we are working on the diet topic. As it is little known in my country, we try to adapt knowledge from others. I have noticed that a lot of experts mention nutrition in the context of feeding SMA I babies via gastrostomy. Almost no one (incl. Dr J. R. Bach) wants to talk about SMA type I diet for babies without PEG. Therefore I would like to ask:

1. What about the babies who are taking food orally? Do any special nutrition rules apply (e.g. elemental amino acid formulas? or extra vitamins or minerals?)
2. What would you suggest for 10 month baby that is able to eat in a natural way? Last month I stopped breast feeding and right now we are struggling with slight loss of weight. We use formula (in composition similar to natural mother's milk) and typical baby food (fruits, vegetables) + L-carnitine + B12, D3 vitamins. Do you recommend any of high calories nutrient to stop loss of weight? My baby now is 30 inches tall and 236 ounces (6700 grams) weight and eats approx 80 kcal per kg of weight per day?

Please advise me. Thanks and regards.

*GailW* : I sympathize with your frustration in being able to find information on diet and SMA, and what you do find is often conflicting. First of all a Type I baby needs to have a swallow test to determine what kind of liquids they can handle. Even when it appears they can swallow fine, they still may be choking a little and it's very important to get that test done. From there you can go on and feed them according to the test results. You can use a regular formula, like Enfamil Lipil AR and then move on to Pedisure with fiber when they are older. These are just formulas to try, all formulas have to be nutritionally complete so if those don't work, try some others. There are many good products on the market. SMA babies don't NEED an elemental amino acid formula, but it can be used. Make sure you carefully research that topic before you put your baby on it. Right now there is not a recommendation for specific nutrients. Carnitine is often low in these babies so a supplement might help. The B-12 might be OK, it's interaction with other nutrients like folate is complex, so I don't know what levels would be "safe" to take. D3 vitamins? If you mean Vit D, be careful, that's the most toxic vitamin. Check the recommended intakes for both those nutrients. If you need to boost calories, use a carbohydrate source, like cereal, fruit or vegetables, rather than protein or fat. If feeding and weight loss become a problem, here are the signs that it might be time to consider a PEG: they are only getting ~50% of the calories they need, feeding time is more than 4-6 hours a day, weight is below their growth curve and dropping. Even if they get a PEG, they can still continue to take food orally!!

*D* : Our son is 4 years old and has Type 2 SMA. I have three questions:

1. He refuses to eat fruit and, even though we only know a few SMA families here in Australia, we find many other SMA sufferers won't eat fruit either. Is this usual in SMA children?
2. He only will eat 2 pureed eggs for breakfast each day - sometimes with a little toasted bread. Is this need for high protein in his diet a common thing? (Though he will not eat pureed meat readily).
3. Do you believe that when he is congested with phlegm in his upper bronchia, he should not drink milk or eat dairy products?

Thank you so much for this opportunity to answer a few questions we find so difficult to find answers for in Australia. Unfortunately, we find it impossible to participate in the online chat with your experts.

*GailW*: Not eating fruit is a problem for 4 year olds the world over! It can be an issue of wanting control over what they eat (and if you want them to eat it, they won't) but most likely it's because their taste buds are growing and changing. Most kids will eat one fruit or another at any given time, but young kids don't like a lot of variety in their food in general. About the time you stock up on one, they decide never to eat that fruit again! You can try "dipping." Kids this age love to dip their food in something. For fruit, try yogurt, sugar, non-dairy lite whipped cream, chocolate, etc. Make it fun. You can do the same thing with vegetables. Things taste differently to kids than they do to adults so you won't always know what they are going to like. The protein in 2 eggs a day might be all right but the cholesterol and fat concern me more. Right now I recommend 1-2 grams of protein per kg body weight, it can go higher if they have a growth spurt or are ill, up to 4 grams per kg body weight. Protein adds up fast so at the end of the day, the total can be quite a bit higher than 2 grams per kg of bw. I do think that when children (or adults) are congested that consumption of dairy foods should be limited. If your child can tolerate thinner liquids, they are better to drink.