

January 15, 2004

Dear Dr. Bach:

I hope this finds you and your family well. The girls must be so big by now! My son Kevin says hello to Gavric.

I wanted to discuss with you your current recommendations for SMA type 2 children, and along with several other type 2 parents, share with you some interesting results we have seen with our children.

Brief history on Erinne: DOB 5-16-97 Currently 6.8 yrs old, type 2 diagnosed at 14 mos. via DNA testing. Hx of 11 pneumonias, May 2001 surgery to release hip flexors & IT bands. Post surgical complications, went into ARDS (5/01 – 9/01, intubated 85 days, 3 failed extubations (successful extubation after NIV protocol was finally followed.) Weight at discharge (9-1-01) 26 lbs and 40 in long. Extremely weak. Unable to support head. Surgeries right after ARDS: Placation of the left hemi diaphragm (due to frozen diaphragm probably resulting from multiple chest tubes) and GJ tube placed August of 2001. T & A April 2000. Nissen done November of 2003 (possible history of micro aspiration pneumonia and reflux.) Changed over to a Mic-key button at that time.

January 2002: Amino Acid diet (2 packets of Vivonex per day (double dilution), plus added antioxidants, Co-Q 10, acidophilus, L-Glutamine, Creatine) instituted. Also began daily use of nocturnal Bi-Pap (20/2), daily In/E x with pressures of +40/-40 (for chest expansion).

Presently, (24 months later) Erinne now weighs 60 lbs and is 51 in long. That is a gain of 34 lbs and 11 inches in just 24 months. I attribute this to correct nutrition as well as Bi-Pap use during sleep (10 hrs a night) and daily use of the In/E x. Her chest is symmetrical. Erinne has no restrictions of any food and no history of choking. She continues to eat as she wishes (mostly snacking). She receives approximately 600 – 800 ml/day of the Vivonex mixture, the higher number on days when she does not eat as much. Bracing: TLSO, AFOs, and KAFOs.

E rinne has shown significant and steady improvement and strength gains while on this diet and NIV regime. All of E rinne's doctors and therapists have said they have never seen a SMA child with steady, sustained improvement such as seen in E rinne. She is seldom tired and goes at full speed from the time she gets up until the time she goes to bed. She attends first grade all day and participates fully in all activities. (Prior to implementation of the diet and protocol, she fatigued easily, woke up tired in the morning, and had very little stamina or appetite and had to nap each day.) The picture below in the wheelchair shows her wearing a soft C collar because she couldn't keep her head up.

BEFORE :



A F T E R (24 months later, post NIV & amino acid diet):



I am aware that the NIV protocol, including daily use of Bi-Pap, In/E x as well as feeding tube are not normally recommended for type 2 children. I am convinced that all of these procedures have not only made her stronger but have also enhanced her quality of life. Prior to this, Erinne was just an “okay” type 2 child. Now she is a vibrant type 2 child who is flourishing. My only regret with the Mic-Key button is not having it placed sooner. I would not even consider taking it out, even though she is eating.

I think the pictures speak for themselves. Also attached to this letter are pictures of other type 2 children (some of whom you have seen in clinic.) You can see "before" pictures (prior to following the NIV protocol and amino acid diet) and "after" pictures which are of course post implementation of the aforementioned treatments. By now I am sure you are aware that the SMA family network is a very close-knit group of people. We have been comparing notes and tracked progress of our type 2 kids. We would like to draw your attention to our case histories and possibly consider recommending the NIV protocol and amino acid diet *as an option* to the type 2 families that come to you for advice and treatment.

I would be very interested in discussing Erinne's history and a more detailed daily protocol if you are interested. I would like to ask you to please give newly diagnosed type 2 families my name and phone number as a potential resource.

Thank you for all that you have done for the SMA children both those you have seen, and those who you have assisted over the phone. You are a tremendous blessing to our children. Many of them, mine included, would not be alive today if it was not for your wisdom, guidance and caring.

With great appreciation,

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Two type 2 girls, both 6.5 yrs, similar abilities, similar height & weight, prior to starting on the NIV & AA diet. One on it, one not:



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**Ryan Brown** is now 7 years old. DOB 3-19-96. He had the Gtube surgery done in July of 2002 at age 6. Some of the reasons we had the Gtube surgery done for Ryan were....he had a very hard time chewing his food (still does) but would tire out very easily and sometimes would have to spit out his food because he just could not chew it, he was always on the verge of becoming dehydrated and was underweight and malnourished. Every time he got sick, he would become dehydrated immediately. This would make the times he did get sick with pneumonia that much worse.

Prior to the surgery (7-02), Ryan weighed 32 pounds and was approximately 41 inches long. Now, as of November 2003, he is 52 pounds and is 46 3/4 inches long. (That is a gain of 20 lbs, 5.75 in in 16 months.) His health has improved. He no longer gets quite as sick as before and he no longer requires hospitalization for each and every minor cold that would turn into pneumonia. He seems a bit stronger and his chest is not sunken in as much as before.

Ryan uses the In-Exsufflator, BiPAP and nebulizer daily. He uses Pulmicort in the nebulizer every night before bed and uses Albuterol as needed. He also uses the VEST as needed. He is tube fed the Vivonex formula by feeding pump at night. I add Vit.C, Vit. Methyl B-12, Calcium, L-Glutamine and Acidophilus to the formula. It turns out to be about 600-700 cc's. During the day he just eats by mouth. He usually eats several small meals.

We are very happy we had the Gtube surgery. E ven Ryan is happy he had it done!  
Mom Lora Berhar can be reached at 541-317-5059 or [loraberhar@yahoo.com](mailto:loraberhar@yahoo.com)

BEFORE :



AFTER:



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**Brett Jacob Wilson:** DOB 12/24/1999 Currently 4 yrs old, type 2 diagnosed at 13 mos. via DNA testing. Hx of 5 pneumonias- 2 requiring short hospital stays, and 1 RSV requiring a long hospital stay- none of the hospitalizations required intubation. Brett weighed

approximately 25 lbs. at diagnosis and would lose approximately 5-6 pounds per illness, become extremely lethargic, dehydrated, and weak. Once recovered, it would take him approximately 6 months to gain weight back but never fully recovered his strength before another illness set in. Surgeries: Omphalocele Repair in December 1999, Tendon Release in August 2002 and G-Tube placement in August 2003- changed to a Mic-key button in November 2003.

**NIV Protocol:** Began using nocturnal bi-pap (18/2) in April 2001 after initial visit with Dr. Bach. Noticed an immediate difference in Brett's abilities during the day, he did not tire as easily throughout the day. Began using Cough Assist in January 2002 with pressures of +40/-40.

**Amino Acid diet:** Began using Tolerex in January 2002(1 packet per day/ double dilution), plus added a multi-vitamin, l-glutamine, folic acid, and primadophilous for children. Switched to Pediatric Vivonex in April 2003(1.5 packets per day/ double dilution), plus add a multi-vitamin, l-glutamine, l-carnitine, folic acid, calcium, vitamin c, and primadophilous for children.

**Bracing:** TLSO, SSO, AFOs, Knee Immobilizers, Resting Hand Splints, Supination Splints.

**Presently:** Brett weighs 28 lbs and is 40 in long. We now use the bi-pap nocturnally as well as at most naptimes at 20/2. We use the Cough Assist daily for chest expansion and as needed with illnesses. Although, the NIV protocol has made a huge difference in Brett, the biggest difference has been made by the g-tube. I think the strength of both of these amazing things has worked wonders for Brett. Brett became extremely ill 1 week after the tube was placed and was sick for 5 weeks. During those five weeks, I was able to keep him hydrated and healthy. He never became lethargic, constantly played and actually gained strength. He grew 2 inches and gained 2-1/2 lbs. during those 5 weeks. About 2 months after the g-tube placement, Brett began to do activities that he has not been able to do since early on in his diagnosis. He has shown significant and steady improvement and strength gains while on this diet and NIV regime.

I am also aware that the NIV protocol, including daily use of Bi-Pap, In/E x as well as feeding tube are not normally recommended for type 2 children. I am convinced that all of these procedures have not only made him stronger but have also enhanced his quality of life. My only regret with the Mic-Key button is not having it placed sooner. I am not sure if these pictures will do justice, but before the g-tube he was very thin, bony and had hardly any fat.

Now his face has filled out, he looks healthy and has "chubbies" on his legs. I would be happy to answer any questions or concerns if needed.

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Before G-T ube (E arly 2003):



After G-T ube (Late 2003):



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**Aschdon Birkle**, DOB 5-17-99: Prior to G-tube surgery 10-4-02, Aschdon weighed 21 lbs and was 36 in long. December 2003, he is now 27 lbs and is 42 in.



long. He was on Vivonex which with the Mic-key we were able to give it to him every day and he gained about 2 pounds. We switched to Tolerex in June of this year and he gained 4 pounds the first month!! He is on one packet and one tablespoon of Tolerex with vitamin C, multi-vitamin, calcium added to it. It is double-diluted as well. He has no eating restrictions besides no milk- just milk not all dairy as he gets a lot of secretions in his throat if he drinks it besides that he is free to eat what he wants which is very little most of the time. Oh yes and he does get the Tolerex every day totaling about 600-800 cc's depending on his food intake for the day most of the time 600 as he eats better when he is hooked up right before he goes to sleep. We are still working on the bi-pap and cough assist issue but we do use the cough assist at least once a day for lung expansion (helps his chest not be so caved in the middle) the bi-pap was using nightly for about 4-6 hours until I broke it some how and we don't have a spare. Please feel free to ask what ever else. Dr Swoboda has seen first hand what Aschdon was like before and what he is like now so she is a good witness!

Contact Information: Mom Kenya Birkle, 307-322-1545 or

[smamom2three@yahoo.com](mailto:smamom2three@yahoo.com).



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Mia Jasmin Haq: Dr. Bach...my name is Stacey Haq...I spoke to you via a phone call about my daughter Mia (Maya) the day after your twins were born. She was diagnosed SMA type 1 by the doctors here in Boise, Idaho at 5 months of age and by the end of our conversation you were of the opinion she was type 2 because of her

ability to sit and maintain. We discussed the CoughAssist, Bi-Pap, and possibly the amino acids diet all of which I was able to get for her right away. I just wanted to let you know of our experience with these treatments. This is a picture of Mia age 3 months here and you can see some deformation in the chest area.



Another at about 8 months old, you can see the chest is becoming concave and sunken.



We began preventative use with the cough assist daily, began her on Bi-Pap at setting per your advice, and started the amino acids diet of Toloorex double diluted in March after she had an eye surgery that at the time we were told was necessary and later learned it was not. Mia almost died coming out of surgery and we were just barely learning proper SMA child care at the time. We put her on NIV and the diet and this little girl blossomed. She became so strong, so healthy, I decided to wait on the

stomach tube because she was doing so great and could eat almost anything given to her no problems or issues with swallowing. Mia was 13 inches around her chest prior to the CoughAssist and Bi-Pap. At last measurement she had increased 2 inches around her chest that I attributed to Bi-Pap and CoughAssist. Here she is in Hawaii in early June just prior to the conference. Look at how good she's doing and



how solid her chest is.

She was on these things until her death in June due to cardiac arrest while doing an emergency intubation. She was fine at the conference, we saw your lecture and you may remember the little girl with the piggy tails on my lap eating fruit loops giggling. A week later, she had been cutting her back teeth, which added secretions and literally developed pneumonia overnight. During transport to the hospital there was confusion about the intubation, she was supposed to stay on Bi-Pap for the ride there but for some reason it couldn't happen. She went into cardiac arrest on the way, once there they got a heartbeat but no response and she was in a coma. They would monitor her but the each day proved her body to be shutting down and not breathing on her own. They declared her brain dead and I took her off the machines. Even though Mia was a strong child on the outside, there is so much we do not know about SMA and it became obvious she was not as strong as I had thought. I truly believe using nighttime Bi-Pap for her and preventative coughing with the CoughAssist prolonged her life the way it did. If we had not had that CoughAssist Mia would have died several times, if we had not had the suction she would have died, and if we had not had the Bi-Pap

she would not have expanded her lungs and halted the deformity in her chest. These pieces of equipment to me are the most important things any type SMA child needs. Mia was type 2 by your opinion and may never have even needed these things in your opinion but in this mom's opinion, they prolonged her life, helped her gain strength, helped her rest better and were all I had to try to save my daughter when she crashed. You can see the effects on her chest and I can give you the measurements that convinced me. I think you are a wonderful doctor to our children and one of the only who will listen and value a parents opinion so I am telling you our story so you might see the real impact of these equipments for our children. They are vital to them regardless the child's type and I really feel she would not have survived after the eye surgery had I not had those equipments on hand for me to use with her. Suction, CoughAssist, BiPap, and amino acids diet..... I saw her blossom on this and I offer my opinion to any newly diagnosed parents out there searching for help. I hope you take into account our experiences and I truly have to say thank you to you Dr Bach for doing what you do and helping our children and more so for having an open mind to actually listen to us parents and take into account our experiences.

Thank you again.....

Stacey Haq

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**Emma Lockwood:** I am the father of two SMA, type 1 children. Emma, our oldest child, is three years old and living a very happy life! The first 20 months of her life, she did not require any additional assistance with breathing or feeding. The last few months of that twenty-month period however, we did see a decline in her eating capabilities and ability to control her secretions. It was at that point that we decided to go ahead with the gastronomy tube surgery. She was hospitalized for a total of 40 days, for she had difficulty being extubated after the surgery. Somewhere in those forty days we began learning about the NIV method that many families across the country are using. She now uses bi-pap, the cough assist machine, suction and is on the amino acid diet. I can't even express how thankful I am for all of these

interventions. Without these, I am certain Emma would not be with us today. There have been times where the cough assist machine has literally saved her life. We have seen Emma doing things that we never thought we would see. We were originally told that the prognosis was not good with SMA, type 1. The doctors told us that she most likely wouldn't live to see the age of two. I can tell you that Emma is extremely bright, speaks very clearly for a three year old, goes to a pre-school in the "non-flu" season. Emma also gains independence in her power wheel chair and does a great job driving! Her pulmonary function is very good considering she has SMA. Since her release from the hospital after the 40-day visit, she has only been hospitalized once for a very brief bout with pneumonia. She has steered clear of the hospital for a year and a half.

Her brother Nicholas is using the same equipment as Emma. He has been on the NIV protocol since about 6 months of age. We started as more of a preventative effort. Now that he is over one year, he is more in need of the equipment. He too is doing extremely well. He has never been hospitalized except for his G-Tube and Nissen Fundoplication surgery where he was in and out in two and a half days!

We have seen amazing things with our children, due to the NIV protocol. I am thankful that it is an option for them and that our children are as happy as they are. I believe they will continue to do well and survive SMA as we wait for a cure that gets closer and closer each day.

Kevin & Beth Lockwood

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Annah Woody:

