

## Families of Spinal Muscular Atrophy

### Transcript of SMA Questions Chat, August 24, 2004

This transcript has been somewhat edited and revised to match questions and answers, correct typos, and to remove extra remarks. Please note there may still be errors and omissions.

Our expert this month is Kristin Krosschell, MA, PT

<i>jeanine</i>	Kristin, my son is type 1, is vent dependent with virtually no mobility. What type of PT would be important for him?
<i>kkrosschel</i>	Range of motion, positioning, adaptations for play, toys and mobility are all important and are things that you should work with your local therapist on. Take into account his need for rest and support for respiration as needed.
<i>optimis</i>	I am the mother of two girls (25 and 30 years old) suffering SMA type III. Can traditional methods such as acupuncture, accupressure, or massages help to strengthen the affected muscles? They can walk but slow and sometimes fall down without any reason. Thank you.
<i>kkrosschell</i>	I am not an expert on acupressure or acupuncture - I am not aware of research supporting increased strength from these modalities for patients with SMA. I am only aware of support for some pain relief and relaxation. You may want to discuss this issue with your local practitioner and/or physician.
<i>hope</i>	How many times a week do you recommend a professional PT to work with your child?
<i>kkrosschell</i>	As far as recommended # of PT sessions per week that is a very individual answer that should be specific to your child's goals and needs. Often many exercises and activities can be carried through at home with less frequent formal sessions, other times a professional is needed on a more frequent basis to guide your sessions, especially if your child is just starting to work on a new skill or to use braces, etc. I would discuss this issue and be sure you and your therapist are on the same page in regards to goals for your child.
<i>4good</i>	What are some of the general ways Moms and Dads can incorporate PT into a child's regular daily routine, as opposed to a PT appointment?
<i>kkrosschell</i>	It is often very easy to adapt and work your child's PT needs into the daily routine you carry out at home and can sometimes be more effective and less stressful for everyone if you can try to work things into daily play, swimming activities etc. Keep in mind that you can have a professional to guide you periodically but often the best therapy and ideas come from very creative planning and solutions that you as a parent come up with on your own. There are many ways you can adapt toys - extending lever arms, using dog or cat toys that are easier to manipulate (just be sure they are safe and do not have pieces that can be broken off), swimming, stretching in the bath, playing reaching games, etc
<i>angiesma2</i>	Can a 39 year old with SMA 2 decrease her joints bent positions with stretching? how often? I am in no pain or discomfort, but would like to use less pillow support at bedtime for my legs. Also can anything in my diet make my tendons more limber? Thanks.
<i>kkrosschell</i>	Not knowing how tight you are and if your contractures are fixed makes your question very difficult to answer online. Have you discussed the question with your local PT or physician? Gentle stretching is probably OK, however, prolonged or aggressive stretching

	may lead to discomfort and other joint issues. I am not aware of any dietary supplement to address the tightness issue- you may want to discuss that question in more detail with your physician.
<i>angiesma2</i>	I look like I am still sitting down when I lay on my back. Last xrays show no "build up" in joints. I can sit "squaw like" so my knees rotate well, but don't extend beyond 60 degrees. Does this help you? I have no PT because of where I live. (Unless my husband can be considered my PT.)
<i>arietto</i>	How can we prevent hip subluxation in a SMA1 baby (3 years of age)?
<i>kkrosschell</i>	Preventing hip subluxation is a good question. Frog leg positioning and weak muscles can make babies with SMA more susceptible to hip dislocation. Discussing positioning with your therapist or orthopedist is a good first step. There are some nice simple things you can do such as pinning PJ bottoms at the bottom with a clothespin to keep the legs less abducted (rolled outward). I can respond to you re: ideas in more detail later. Also, it is important to know that in these babies hip subluxation often is not an issue if they are not in any pain or discomfort. Typically positioning and ROM are good first steps in addressing the issue. Surgical intervention should not be necessary unless pain is a primary concern, however, discussion with your local ortho doc should be helpful to determine details specific to your situation.
<i>decarlo</i>	Hi, my name is Franco, was born in Brazil, and I'm 27, diagnosed SMA II. I'd like to know if electric stimulation can prevent or slow down fibrosis.
<i>kkrosschell</i>	There is no research evidence that supports the use of electrical stimulation (NMES, FES, TES) for patients with SMA. In addition, we do know from research with other populations and how Estim works that there actually may be some tissue damage or harm that could occur with Estim in muscles that are already atrophied and denervated.
<i>administrator</i>	There is a link on the FSMA site to an article about the usefulness of TES in SMA on this page: <a href="http://www.fsma.org/links-ptot.shtml">http://www.fsma.org/links-ptot.shtml</a>
<i>hope</i>	Our PT was wondering if our daughter (18 mo. Type 2) is not showing increased strength neck extensors and has difficulty in prone positions, should she continue to focus on those postures if she won't gain anything? Do you have any ideas re: therapeutic activities?
<i>kkrosschell</i>	If your daughter is 18 months there are many other positions than prone in which you can work on neck extension. However, if the sole goal is so that she can lift her head from prone and she does not tolerate the position well then I would change your goal to work on something that is more functionally relevant and meaningful for her such as improving head control and endurance in upright postures such as sitting or in a stander if she can tolerate one.
<i>Hollandia</i>	As valproic acid and sodium butyrate seem to have a beneficial effect on the upregulation of the smn gene, I wonder if there will be nutrients or a lifestyle that will have a negative effect on the regulation.
<i>kkrosschell</i>	Your question is one that is out of my line of expertise. Great question, you may want to address that question to one of the physicians scheduled for future chats.
<i>jeanine</i>	My son (9 yrs old with type 1) wears knee immobilizers at night to prevent contractures. We are now seeing one foot turning out at a 90 degree angle when his knees are pointing straight up. Any thoughts on what's happening and what to do to correct this?
<i>kkrosschell</i>	Sometimes, and again not always, the immobilizer may in fact stretch and straighten the knee, but the weight of the immobilizer may cause the foot to look like it is turned out because the hip, not the foot, is rolled outward after the knee is straightened. Without

	seeing your child it is hard to answer this in more detail. They may also have a tendency for some tibial torsion (outward rotation of the lower leg) that may or may not be related to the immobilizer. U may want to discuss this with your local healthcare professionals so that they can actually see what is occurring and better determine if in fact it is even anything you need to be concerned about.
<i>hope</i>	Do you promote water therapy? If so, what specific activities do you find helpful for a type 2 18 month old. Any suggestions we could do at home rather than going to the adult heated pool where siblings are not welcome? Are there swimsuit options that help with head/trunk flotation?
<i>kkrosschell</i>	Swimming is great and oftentimes I think it can be some of the best therapy for children and adults with SMA. It is easier to move in the water with gravity eliminated and the water itself can be relaxing and a fun place to exercise/play.
<i>angiesma2</i>	My father had an inflatable tire tube that he placed around my neck at that age when I went swimming. Of course it was small tubes ... gotta find one that fits, if it's to your liking. I had a life preserver shaped like a ring (orange coloured) and my mother sewed a sling seat in it so I was also up right.
<i>kkrosschell</i>	Keifer products, Aquatherapy and Danmar products all sell some great adaptive aids and supportive aids for the pool. There are supports and suits that are inflatable, have floaties in them and then there is a suit called the "Wet Vest" out of Canada that actually holds the child upright so that they are in vertical and can do some walking in the pool where gravity is eliminated making it easier and sometimes quite fun. Many of these catalogues can be accessed online and/or you can call the company and request a catalogue to see the many adaptive things that are available.
<i>4good</i>	I think I've heard of extending lever arms, but could you explain them/it in a little more detail? Are they slings that position a child's arms for greater motion and less gravity?
<i>kkrosschell</i>	As for extending lever arms there are multiple ways to do this from something as simple as a direct adaptation to the toy (adding a tongue depressor or popsicle stick to a lever on a toy (such as on a pop up box) can make it much easier to be successful in operating the lever- less actual force or strength is required. Or there are slings/springs support devices that work well to support the child's arms. These are commercially available and/or can be made with theratubing or thick elastic bands. I can send you some more info later that is more specific if you are interested.
<i>lassmank</i>	Franco mentioned e-stim, and you replied that it's not effective in muscles that are already atrophied/denervated. I work with an individual who just recently has lost use of his swallowing muscles, so we tried a type of e-stim commonly called ActivStim, as the folks promoting it have some small number studies indicating that it may be one of the more effective modalities for dysphagia, at least as it relates to many conditions, although MD/SMA hasn't been explicitly included. The individual with SMA III had 11 sessions of ActiveStim, but no real swallowing gains as evaluated through FEES studies. Is this probably barking up the wrong tree, or have you run across any anecdotal information about ActivStim with folks with this diagnosis who have recently lost the ability to functionally; i.e. treatment outcomes, number of sessions, usefulness in maintaining the ability to handle their own saliva, etc.???
<i>kkrosschell</i>	Good question. I am not aware of any of the studies with Activastim done with SMA patients and as I am not an expert on dysphagia this may be a question to address to a speech and/or swallowing expert. There may be some studies underway that I am not

	aware of. Dr. Kissel, our adult expert, may be able to address this in the next chat.
<i>optimis</i>	My daughter's feet are turning inward after using crutches 3 years ago. Can they be corrected?
<i>kkrossshell</i>	This is difficult to answer without knowing more about why your daughter was on crutches- was she injured? If so what was her injury?
<i>optimis</i>	No, it is not injured. She is using crutches to balance her body while walking. She will swing her body sometime instead walking properly. She is using Canadian canes.
<i>kkrossshell</i>	Has she always walked? Is she Type 2 or 3? You may want to follow up with a local therapist to discuss your question in a more specific manner. Unfortunately, not knowing or seeing your child makes this a very difficult question to address in this forum.
<i>arietto</i>	Can “fixed” positions and postures in SMA1 babies cause cranial and facial deformities? Is there any way / technical support (pillows or other things) to prevent or avoid them? Our daughter is a SMA1 3 years old (NIV): have you got any suggestion to prevent jaw retraction (she lies, with an angle of 35-40°, on her side or on her back with the head turned left or right)? Which kind of positions/postures can we adopt to prevent and possibly avoid scoliosis? How can we mobilize the thorax, spine and the pelvis? What are your suggestions to prevent / avoid joint contractures?
<i>kkrossshell</i>	Any baby that is positioned in or that can only be positioned in limited positions may in fact develop cranial and or positional plagiocephaly (molding) issues with their skulls. There are many devices, soft pillows, car seat head supports and supports for positional deformities that can be utilized very easily with these children to try to prevent some of these issues. Target actually sells a great soft plagiocephaly pillow (“Boppy” is the brand) that can often be very effective with some of these issues. There are lots of positioning adaptations that you can try to minimize contractures. Can I get back to you on this question after the chat as I can probably be more specific and refer you to the article I recently did on Type 1 babies that addresses some of these issues.
<i>pierna</i>	My nephew just turned 13 yrs. with the mild form of SMA. How important is his physical therapy so that his condition does not worsen? He is doing wonderfully with only hip and knee bending problems, but we see that increasing slightly in the last couple years.
<i>kkrossshell</i>	You did not mention if he receives any PT at this time. If he is getting increased hip and knee tightness you do want to be sure that you are stretching and or positioning those muscles in an elongated fashion for some part of the day so that you can try to slow the contractures down. It is often hard to reverse the contractures if you do not address them right from the start. You can usually learn some great stretches and tips from a therapist in 1-2 sessions and then carry out the suggestions at home. The tightness will start to make walking more difficult if they continue to increase.
<i>hope</i>	I have read that SMA people are often low on carnitine levels which can make them weaker. Are there any SMA people getting a carnitine supplement and if so, has that made a difference in their strength?
<i>kkrossshell</i>	Hope I am unaware of anyone that is getting Carnitine that is administered other than if they are involved in some type of research trial. This is a question that you should address to your physician and/or other panel experts that will be conducting chat sessions later. I am not a physician so that is not my area of expertise.
<i>4good</i>	Kristen, there was some previous mention of massage(along with acupuncture / accupressure) therapy. Do you think MASSAGE therapy is beneficial? If not for strength than perhaps circulation and prevention of contractures etc.?

<i>kkrosschell</i>	Massage can be very beneficial for circulation and sometimes to assist in prevention of further contractures
<i>Helmer</i>	Are there degrees of SMA where exercising, swimming and other physical activities have no (physical) beneficial results ? i.e getting stronger or can all SMAs train their bodies to become a little bit stronger?
<i>kkrosschell</i>	Your question is a good one, albeit difficult to answer without knowing more about the specific child or adult. Exercise in moderation is the key, over exercising can be harmful and not beneficial. This issue is specific to each person's degree of weakness and function.
<i>angiesma2</i>	Assuming gentle stretching is neutrally safe, where does the hands go when doing hips and knee stretches .... my husband has no idea of how to do these things. Guidance or direction would be appreciated. As a teenager I went to variety village as a member. I tried weights, no difference in strength for me, but simply spinning a hand wheel for aerobics made me feel "pumped up" ... didn't rejoin.
<i>kkrosschell</i>	It sounds like you have a moderate degree of tightness. Gentle slow stretches are the key- less reps, longer time in the stretch. It is hard to tell u where the hands should go specifically without knowing more about your specific contractures. It is hard to tell from words where your tightness actually is. Maybe I can get back to you after the chat with a more specific answer after gathering a bit more info from you. Usually you want to provide support above and below the joint that is being stretched while you are stretching.
<i>Helmer</i>	So you can overdo it?
<i>hope</i>	I know that each person is different in their strength and abilities but besides a stander is there any other pieces of equipment that you have found to be extremely useful across the board with children that have a little bit of strength? (can sit well when put there, can still roll to name a few)
<i>kkrosschell</i>	Again the equipment question is hard to answer in a general sense. I try to assist parents in finding some type of toy or play/fun device that does not look or feel like equipment that may address the postural issues, allow increased function by giving appropriate support as needed so that the child can participate in and be a part of as many peer related activities as is possible. Some type of mobility device that offers independent control, if appropriate, is very beneficial.
<i>Helmer</i>	We have had great success with a loft lift at home (whole ceiling in the living room) and my son w/ SMAII then stands in a form of lift pants and "walks" around the whole living room lifted up by the lift - he can then sit down on the floor, chair or play football - this is an addition to the "standing table" where he stands for minimum 2 hours a day and preferably 4 - 6 hours a day. I have found that changing positions and different sports i.e exercises daily has made him much stronger and perhaps the whole question is not to overdo it - so he also has time to recover from all his physical activities. And a stupid question - do the SMAs become more tired from physical activity than you and me? do they need more time to recover?
<i>kkrosschell</i>	It sounds like you have quite a busy day, but it sounds like you have tried to incorporate play and FUN and that is key. Yes, time to rest and recover is very important and children with SMA are more prone to fatigue and will require more rest than you or I. As long as you are assuring appropriate rest and recovery periods you are probably just fine. I'd love to see a picture of your system if you have any way to send one or to post it to the Equipment board on line.
<i>Helmer</i>	Certainly will send you some photos over - both training football and while dancing ! let

	me get hold of a camera and I'll send you some photos from Denmark !
<i>angiesma2</i>	It sounds like an adult version of a child's jolly-jumper, ;-)
<i>kkrosschell</i>	Thanks for offering to send the photos. Anyone else with creative or adaptive ideas that have photos please share at the website or to my email. They are great to use when providing others with ideas. Often it is the parents' creativity that sparks the solution to many others problems and questions.
<i>administrator</i>	A plug from your webmaster: If you have a photo please share it on the Equipment message board here: <a href="http://www.fsma.org/mb/view-equ.asp">http://www.fsma.org/mb/view-equ.asp</a>
<i>jooolsie</i>	My daughter SMA2 age 19, luxation in one hip, sublucation in other. We "hang" her legs alternatively over the side of the bed to stretch the hip down. Can we do anything else? She has a stander and can use it for between 30 - 45 mins before she tires.
<i>4good</i>	what is luxation and sublucation?
<i>jooolsie</i>	dislocated and partly dislocated
<i>kkrosschell</i>	It sounds like you are saying one hip is dislocated and the other is sublucated. If this is in fact the case there will be some associated tightness that you need to be cautious about during your stretching program. There are stretching positions that may actually increase and make the hip sublucation worse. Have you visited a therapist or has your physician instructed you in correct positioning during stretching?
<i>jooolsie</i>	We only visit a therapist once a year and I was told that would help when she was young, years ago.
<i>kkrosschell</i>	Those exercises may have in fact been the right stretches years ago and may still be now. However, it sounds like she may have changed, as most of us do, and it would be good to have a review of those stretches with your local therapist to be sure they are still helping and that they are in fact the best stretches for you to be doing.
<i>hope</i>	We have a standing Dani and was wondering what type of goal you would have for time in it. I am sure it is probably up to how well your child tolerates it. Is it better to have her in it for many short times throughout the day or one long period if she doesn't mind?
<i>kkrosschell</i>	It is up to tolerance, however, let me preface that. Remember standing time should be somewhat age dependent also. We do not stand as young children for long extended periods without bending, squatting, stretching etc. Normal play involves lots of movement. Oftentimes, even as adults, prolonged standing can be less comfortable so shorter periods may be better. When children are tight they sometimes tolerate shorter and more frequent periods better also.
<i>arietto</i>	What are your suggestions to prevent / avoid joint contractures? Which kind of exercises do you suggest to stretch the muscles? How many times and how long do they have to last during the day? A scarce mobilization of the trunk can be a cause for respiratory problems?
<i>4good</i>	Kristin if you could send me some more info on extending lever arms it would be appreciated.
<i>kkrosschell</i>	Your questions have all been great and thought provoking. I'd love to stay and chat however, have other commitments at this time. I will get your email addresses and respond to any questions I missed as soon as possible. And Angie I am sorry without seeing you I do not know if you are a grown kid or an adult but your assistance in answering questions and your ideas, as well as everyone else's were great. Bye for now.
<i>Lots of people</i>	Many rounds of "Thank You" to Kristin for sharing her time and expertise.

