

Families of Spinal Muscular Atrophy

Transcript of Respiratory Issues in SMA Chat, Monday, November 13, 2006, with Dr. Mary Schroth, Pediatric Pulmonologist at the University of Wisconsin in Madison.

While this transcript has been reviewed by Dr Schroth, many typos and errors may still exist - it is, after all, just a 'chat.' Identifying information and chit chat have been removed, and answers have been matched to questions.

administrator : Welcome Dr Schroth and thank you SO much for being willing to do this chat!

TL : YES - thank you!!

Dr Schroth : Hi everyone. I am Mary Schroth, a pediatric pulmonologist at the University of Wisconsin in Madison. I have been active with FSMA for the past 8 years. Today we will discuss respiratory issues related to SMA. I will do my best to answer your questions.

administrator : A question from a user who could not join us because of time difference: "My daughter (3 years old) received the flu shot. She hasn't had any side effects (for example high temperature etc.). Does this mean that her immune system is not prepared for the virus attack?"

Dr Schroth : The flu shot is a killed virus so it does not cause infection. The flu shot stimulates the immune system to make its own antibodies to fight infection when the body "sees it" during flu season. The most common reaction is soreness at the sight of the shot. So, it is normal and good that she did not have any side effects and it should help protect her from the flu this winter.

TL : When a kiddo is sick - breathing fast - increased heart rate, etc what immediate tests should we request at the hospital to determine if it's pneumonia, collapsed lungs, or just a bug?

Dr Schroth : The most important thing is for someone to listen to her lungs. Generally we can hear where the problem is. Oxygen saturation check is also important and a chest xray to see where there may be areas of mucus plugging.

TL : With the x-ray we've been told they can't really tell the difference between pneumonia or collapsed portions - is that true?

Dr Schroth : Yes, it is difficult sometimes to tell whether it is pneumonia or mucus plugging on an initial chest xray. But we also make a determination about that based on what the lungs sound like and how the child is doing.

C : Dr Schroth, is assisted coughing (by my mother to help me) the best way to help keep the lungs clear of congestion? Any long term problems with pushing too hard etc?

Dr Schroth : Yes, assisted coughing or using a Cough Assist machine is the best way to keep the lungs clear of congestion. Generally there have not been long term problems with the manual cough assist as long as it feels ok to you.

SC : Dr. Schroth --- We have a 16-year old daughter with Type 2 SMA who has some loss of oxygen (sleep apnea) and asthma. She needs oral surgery for removal of her wisdom teeth. The doctors prefer general anesthesia (without intubation). Can she go this route?

Dr Schroth : Your child can have her wisdom teeth removed with general anesthesia as long as there is another doctor watching and taking care of your daughter along with the oral surgeon or dentist. So there should be an anesthesiologist as well as the surgeon.

DM : Is there a significant difference between using a nebulizer with saline and Xopenex and if so what would indicate the uses of both during CPT prior to a cough assist?

Dr Schroth : Xopenex is a medication that contains a form of albuterol. It is not clear to me whether Xopenex or albuterol make a difference for children with SMA before CPT or coughing. My rule of thumb has been to try it especially if there is a history of asthma in the family or the child has had asthma type symptoms. However, it does not seem to help every child. I am convinced that coughing and chest PT helps every child. You are so busy taking care of your child that I recommend that we determine what works and do that. And if it does not make a difference then it may be a waste of your valuable time.

MR : Should we get an antibiotic every time he has a cold or just when he has a fever? Sometimes the colds last 3 weeks and the antibiotic is just for five days - can they become immune to the antibiotic if we give it every time he has a cold?

Dr Schroth : The purpose of the antibiotic is to prevent a bacterial infection, which can contribute to pneumonia. Colds are caused by viruses and in general there is no cure, we have to wait them out. But viruses cause a lot of secretions in the lungs and nose and colds make every one weaker. I generally treat with an antibiotic only at the beginning. I generally do not treat with an antibiotic the entire illness.

TL : My son had pneumonia back in May - they set bipap at 18-8 - he's 3 years old and doing better now - should we change the setting now?

Dr Schroth : Your son's BiPAP settings could possibly be lowered to 18/6 or even 4. That is the EPAP setting or expiratory positive airway pressure. He may have needed the EPAP of 8 while he was sick but may not need it anymore.

In general, the BiPAP settings should be IPAP 12-18, EPAP 0-6, respiratory rate 20-30. That probably seems like a big range but it depends on how well your child is tolerating BiPAP, does the BiPAP work well with their own breathing, does their chest rise with each BiPAP breath. Some centers titrate BiPAP using a sleep study and many do not.

JS : I'm an adult with SMA type 2. Could you please discuss the effectiveness of LVR (lung volume recruitment) exercises for the group? I've doing it for two weeks and have seen impact already. It's basically stacked breathing with an ambubag, it stretches lungs, helps cough and improves ventilation. (More info at:

<http://www.ottawahospital.on.ca/rehabcentre/servicesclinics/respiratory-e.asp#2>)

Dr Schroth : The lung volume recruitment exercises are great for older children and adults who can cooperate. In younger children, I use the Cough Assist on the inhale mode (turn the exhale time to 0).

MR : What should the settings of the BiPap be when he is not sick? We just started him, he is at 8-4 and does well with it. He is 18 months. How long will it take to build up the chest wall?

Dr Schroth : I suggest increasing the IPAP pressure gradually up to 16 in order to improve his chest wall expansion. And he may tolerate an IPAP of 18. Does he have a respiratory rate on his BiPAP?

MR : We don't know what it is, what should it be? This is new to us.

Dr Schroth : It is possible that you are using a BiPAP machine that does not have a respiratory rate. This is called spontaneous mode. I recommend a machine that provides a backup respiratory

rate, because sometimes your child's breathing may be shallow and the machine cannot tell when he is taking a breath. With a backup rate the machine will provide a minimum number of breaths per minute usually 20-30 depending on the child's age.

TL : Our son's machine has this feature of guaranteed breaths per minute and it works very well - without it his breaths weren't strong enough to pick up always and he would get out of sync with the machine. While he's healthy we use 20 automatic breaths and when he's sick and breathing faster we raise it to 30.

MR : Thank you.

CC : Hi Dr. Schroth! My daughter is 3 years old and type III. When do you feel is the appropriate time for us to get her set up with a Pulmonologist? Should we do it now although she hasn't had any problems yet or wait a little longer?

Dr Schroth : For a child with SMA III, now is good time to meet with a pulmonologist to get acquainted. They may only need to see your daughter once per year, but it helps to establish that relationship before there is any respiratory difficulty.

CC : thank you

K : Hi Dr. Schroth ~ what is your opinion of RSV shots for children over 2 (my son is 3 and a Type I)?

Dr Schroth : Synagis is a humanized monoclonal mouse antibody given by a shot once per month to prevent RSV infection (Respiratory Syncytial Virus). So it is different than the flu shot where you give a flu protein that stimulates the immune system to make antibodies. Synagis is the actual antibody and it breaks down over a month and has to be redosed. It is given for 6 months out of the year during RSV season.

TL : Do you recommend Synagis for a 3 month old not big enough to receive flu shot? Does the full 6 doses need to be done to be effective? Is there any risk with Synagis shots?

Dr Schroth : Synagis is given during RSV season. In Wisconsin, we give Synagis from Nov to April, every 4 weeks. Children must be 6 months old to get the flu shot. For children less than 6 months old, their family members should all get the flu shot.

For children over 2 years old with SMA I, my advice is to ask your insurance company whether they will pay for Synagis. The current guidelines and research suggest that children over 2 may not benefit from it. The other thing to realize is that Synagis does not provide perfect protection and children can still get RSV even with Synagis. The best prevention is to avoid others who are sick and good handwashing.

TL : My 3 month old son has SMA (we assume Type 2 like big brother.) When do we start using cough machine, bi-pap, suctioning on him? No problems as of yet.

Dr Schroth : For your 3 month old, SMA II, when you start to notice symptoms of weakness, I would start Cough Assist machine. The time to start BiPAP is variable and will depend on his muscle strength.

TL : thank you

K : Do you think it's worth getting The vest or an IVP machine for a 3- year old child with Type I or is basic CPT, cough, bipap enough?

Dr Schroth : If you have been successful keeping your child healthy using CPT, cough and BiPAP, I would continue to do that. I prescribe the IPV (intrapulmonary percussive ventilator),

when manual CPT has failed to keep the child out of the hospital and also they tolerate IPV when they are in the hospital. Not every child does well with IPV. No one has shown that one form or airway secretion mobilization is any better than any other form so I recommend using what works.

K : Thanks. We've been very lucky and he has never been hospitalized except for elective surgery.

TL : How does IPV work?

Dr Schroth : IPV uses an air source like a air pump to deliver air into the lungs by mask or mouth piece at a set frequency and pressure. It sounds like a choo-choo train. It essentially does CPT from the inside using air to shake up the secretions. Some people do not like it, even less than the cough machine.

MR : We had difficulty with the cough machine - he is 18 months, does he need to be a little older to breathe with it? Is CPT and bipap enough to help him when he is sick until he gets used to the cough assist?

Dr Schroth : I encourage you to work with your child to get them used to the Cough machine. What I have parents do is to coach your child by saying "take a big breath in" while the machine is doing the inhale. And when the machine is on the exhale mode, say to your child "now cough." If you set the time on the modes the same your child will learn the rhythm of the machine especially if you coach them through it with directions.

MR : Thank you again.

Dr Schroth : What I meant by setting the time modes the same is that inhale is always 1 or 2 seconds, exhale mode is always 1 or 2 seconds and pause is always 1 or 2 seconds.

MR : Thank you.

CC : Dr. Schroth, is it safe to give our kids products like Airborne Jr. to add extra protection against catching colds? Thank you!

TL : or Mucinex?

Dr Schroth : Yes I think Airborne Jr is safe to use. I have looked at the ingredients and I am not concerned. I think Mucinex has less to offer and contains an ingredient that is very common in most cold over the counter medications, guaifenesin.

CC : Thank you

TL : Do you think Airborne ingredients would actually help prevent anything?

Dr Schroth : I do not know whether the ingredients in Airborne will help to prevent anything. I can't recommend one way or the other.

CG : My 3-year old son has SMA type 2. We live in Chile, South America. This year he has been 4 times in the hospital due to pneumonia, all of them he required mechanical ventilation, once intubated and the others with NIV (BiPAP). My question is: Is it a good practice to use antibiotics as soon as he presents signs of a cold? What type of antibiotic will be the more appropriate to start with?

Dr Schroth : The most important thing to help keep your child out of the hospital is the equipment we have been discussing. Do you have a Cough Assist machine and BiPAP for your child? The antibiotics only help if you are helping your child to clear all the secretions from their lungs that they are too weak to cough out by themselves.

CG : I do have Cough Assist and Bipap, and we use then both. The problem is that my son seems to have asthma and is hyper reactive after and RSV. What can we do to help him??

Dr Schroth : If your child seems to have asthma, then he may benefit from albuterol and/or an inhaled steroid, medications we use in asthma. These may help keep his respiratory status "more even."

CG : So far we have use the BIPAP only when he is sick since his sleep study turned out well (97% O2 all night), so my question is: . What signs should we look for to decide if our son needs to use NIV (BiPAP) during nights?

Dr Schroth : If his sleep study is ok and you are using BiPAP only with illness this is ok. If you start to see his chest sink in at the sternum, he may benefit from BiPAP every night.

CG : Dr. Schroth, considering that our son seems to have asthma, is it possible that the use of a cough-assist machine (In-ExSufflator) causes bronchial obstruction? What are the suggested parameters (pressure and time) for a SMA type 2 kid with and without a cold?

Dr Schroth : If your child has a cold the only way to clear secretions is to cough them out. In my opinion, the Cough Assist does not contribute to airway obstruction. The settings I recommend are inhale pressure 25 to 40 cm of H2O for 1 second, exhale pressure -25 to -40 for 1 second and pause time 1 to 2 seconds. Four to 5 breaths are taken continually followed by suctioning the mouth to remove secretions coughed up and a rest period of 1 to 2 minutes. We do 4 sets of 5 breaths. The goal is to gradually increase the pressure to 40 on inhale and exhale.

NB : Our son is 13 months old, has type 1 and has been through several pneumonias already. They are not able to get an IV on him and he had PICC line put in, but it got infected so we are looking at putting in a port-a-cath. Would you recommend this? Also if we have it done should we look for someone that will do it without intubation? Any suggestions, or any questions that I should ask the surgeon? Thanks.

Dr Schroth : The decision about a port-a-cath is difficult. If every time he comes into the hospital it is very traumatic for all of you to get an IV started, the port-a-cath may be very helpful. I use general anesthesia at my hospital and we are aggressive about taking the breathing tube immediately after surgery and using BiPAP.

NB : Thank you, we have been speaking with the surgeon and will again, but I am wanting to know before we go if they will do just that. Put him on bi pap quickly, they also mentioned a bag of some sort they may use instead of a tube?

Dr Schroth : Sometimes we use a laryngeal mask for anesthesia. I suggest meeting with the anesthesiologist as well as the surgeon about your desires.

SC : Our 16-year old daughter has restrictive airway and asthma with some loss of oxygen as determined by her sleep study. She will not wear her C-Pap mask nightly (she says it interrupts her sleep). She currently wears it about twice a week or sometimes every 3 nights, but she removes it sometime during the course of the night. What protocol do you recommend?

Dr Schroth : There are multiple issues to consider when your child dislikes their mask. There are lots of different styles of nose masks and I suggest you contact your home care company to try some others. There are some that are nasal pillows or canulas that do not block vision while they are on. Another issue to consider is whether the BiPAP settings are optimal for her. Possibly the settings should be evaluated by her doctor. If she is only on CPAP this may be the problem.

SC : Thank you! Our teenager likes the C-flex machine, but I will investigate your suggestions!

Dr Schroth : I suggest switching to BiPAP from the C-Flex. The CPAP will not provide her with enough support for her breathing during sleep.

SC : If I were to get a copy of our teenager's sleep study, would you be interested in looking at it and discuss an ongoing protocol with our pulmonologist here? If so, how do I go about doing that?

SC : Also, a group of us in Atlanta are interested in helping our pediatric hospital adopt some of your proven protocols to avoid invasive procedures when intubating in surgeries, etc. How can we go about doing this? We have 2 doctors who are interested in working with us. ;-)

Dr Schroth : You can contact me outside the chat to discuss further.

DM : Dr. Schroth - our 2 1/2 yr old daughter has had 2 bouts of "sudden apnea", are there any signs that would tell us whether or not they were caused by plugs vs. vasal vagal response?

Dr Schroth : The 2 sudden episodes of apnea are sometimes difficult to figure out. My suggestion is to cough her and see whether anything comes up or clears from her airway. There is a concern among those of very interested in SMA as to whether there is autonomic dysfunction and we are all trying to figure out the best way to sort this out.

TL : My son's bi-pap is like the cough machine mask - is it ok if it covers his mouth and nose?

Dr Schroth : It sounds like your son is using a full face mask with the BiPAP. I prefer a nose mask, because you can hear him talk around the nose mask. If he is doing well with the full face mask, this is ok too.

MR : When my son has a cold and his nose is stuffed up, is it safe to use the mask (we use the mini me) for the bipap? Does he just know to breathe through his mouth?

Dr Schroth : When your son has a stuffy nose it is ok to use the mask. I suggest cleaning his nose out of secretions as much as you can before using the BiPAP.

J : Dr Schroth, could you tell us a little bit about the possible benefits related to VEPTR device in children?

TL : is VEPTR better than standard growth rods? Does your hospital do VEPTR?

Dr Schroth : The VEPTR works well for children with congenital scoliosis. Some centers are using it in children with SMA. I do not have experience using it in children with SMA and I would like to see more info before considering using it exclusively. One worry I have is: are the bones in children with SMA strong enough to hold it in place? My hospital does the VEPTR procedure, but we have not done it on children with SMA.

NB : Thank you Dr. Schroth and thank you to FSMA for these chats and to my friend for making us aware of it.

CG : Dr Schroth, thanks a lot for your time and help !!

TL : YES - THANK YOU SO MUCH!

MR : Thank you, you helped me so much!

CC : Yes, thank you so much Dr. Schroth for taking the time tonight to answer our questions! You're the best!

DM : Thanks so very much for your help and dedication toward those with SMA!!!!!!

J : Brazilian parents would also like to thank you!

SC : Dr. Schroth, you are a champion for us parents and we are indebted to you! Thank you for your time tonight and thank you to FSMA for providing this opportunity. You are an angel!

CC : Thank you for putting this session together...hopefully there will be another one in the near future!

Dr Schroth : Thank you all!!

Families of SMA

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