PARENTS AND RESEARCHERS INTERESTED IN SMITH-MAGENIS SYNDROME

SPECTRUM

PRISMS

VOLUME 11, ISSUE 4

FALL 2007

NIH SMS Research Team to Launch Sleep Treatment Study

Researchers at the National Institutes of Health (NIH) are launching a controlled treatment study to determine if bright light alone or in combination with a melatonin preparation is effective in treating children with Smith-Magenis syndrome who have a significant sleep disorder.

Persons with Smith-Magenis Syndrome (SMS) have a disrupted sleep cycle involving early waking, frequent daytime napping, and frequent nighttime awakenings. Melatonin is a hormone normally produced at night in healthy people. People with SMS produce high levels of melatonin during the daytime and very low levels at night (Potocki et al., 2000; Deleersnyer et al., 2001). Under the comprehensive SMS natural history study at the NIH (protocol 01-HG-0109), critical sleep data collected over the last 4 years by the SMS Home Assessment of Sleep (SMS-HAS) has led to the design of the first controlled treatment trial to study two different treatment approaches alone and in combination: bright light therapy and use of a delayed time-release melatonin tablet preparation. The melatonin tablet was developed by the Pharmacy Development Services at NIH and was approved this summer by the FDA for use (IND 77,802) in this study.

The study is sponsored by the National Human Genome Research Institute (NHGRI) and will be conducted by the SMS interdisciplinary team of researchers located at the NIH & led by co-principal investigators Ann C.M. Smith, MA, D.Sc. (Hon) NHGRI/NIH and Wallace Duncan, PhD, NIMH/NIH and Wendy Introne, MD, Medical Responsible physician, NHGRI/NIH. Travel funds for the child and one parent are available.

Eligibility criteria: Children with a confirmed SMS diagnosis (del 17p11.2) between 5-16 years of age who have not reached puberty and are seizure free are eligible. Documentation from the physician/geneticist who confirmed your child's diagnosis and/or copies of laboratory reports, medical & developmental records will be requested for review by the research team to determine eligibility. Since certain medications may exclude individuals from participation, a telephone screen to review your child's medical history and all current medications (both prescribed and over-the-counter) will also be conducted as part of the eligibility review.

The two major parts to the study will be briefly described. Participation in all phases of the study, while optimal, is not a requirement.

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INSIDE THIS ISSUE:

What Is Smith-Magenis Syndrome?

Smith-Magenis syndrome (SMS) is a chromosomal disorder characterized by a specific pattern of physical, behavioral and developmental features. It is caused by a missing piece of genetic material from chromosome 17, referred to as deletion 17p11.2. The first group of children with SMS was described in the 1980's by Ann CM Smith, MA, a genetic counselor, and Ellen Magenis, MD, a physician and cytogeneticist. Although the exact incidence is not known, it is estimated that SMS occurs in 1 out of 25,000 births. SMS is underdiagnosed, but as awareness of it increases, the number of people identified grows every year.

Questions?

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A Message from PRISMS President…

Randy Beall

It can't believe that summer is over. Fall is officially here and it's back to school.

Hey - a BIG Texas "shout out" to all aspiring PRISMS board secretaries! Don't be shy. We need you!! Our current secretary, Rochelle Wright, will be stepping down at the end of this year. Please consider joining us and making a difference in the lives of our families. You will find a board application in this newsletter.

Some of our families are planning socials this fall such as picnics. These can be a great way to meet others, share information and support each other. I know of events in Houston, TX and York, PA. Check the PRISMS website under "about Us"-conference and events, for details or consider planning one in your area. Maybe just call one other SMS family and have them over. Let us know if we can help.

We have officially booked the Reston Virginia Hyatt again for our 2009 Conference. Yeah! The conference will be Sept 17-20, 2009. Start planning and saving now!

As you will read later in this newsletter, we have folks that have been beating the streets to raise money for our families. A big "thanks" to the McGrevy's and the Foster's and everyone that participated in the 5K race for PRISMS. I understand that it was a tremendous event!

We have developed a number of materials that can help make hosting an event easy. Fundraisers raise awareness too and they're fun. I will never forget that one of our SMS moms told me that hosting a fundraising event was one of the best days of her life! Wow!! Won't you please consider hosting a fundraising event in your area? It could be a run, a walk, a bowl-a-thon or golf tournament, just to name a few event ideas. If you'd like to kick around some ideas, please contact Percy "Mr. Fundraiser" Huston (info@prisms.org).

I look at the Bridges of Hope slideshow from time to time to remind myself of our precious kids. As an organization, we're working hard to provide information and support for our families. Here's to a brighter future for our families!

Randy

Randy@prisms.org

Hyperacusis in Smith-Magenis Syndrome

Carmen Brewer, PhD[1], Jennifer Bentley[1], Chris Zalewski, MA[1] and ACM Smith,MA, DSc(hon)[2]
[1]NIDCD/NIH, [2]NHGRI/NIH

This research was conducted by audiology doctoral student, Ms. Jennifer Bentley working with audiologists Carmen Brewer, PhD and Chris Zalewski, MA as part of the IRB-approved comprehensive natural history study of Smith-Magenis syndrome (protocol 01-HG-0109) being conducted at the National Institutes of Health (NIH). This article is based on Dr. Brewer's presentation at the May 2007 PRISMS Conference in Reston, VA.

ENT abnormalities are documented in as many as 94% of individuals with Smith-Magenis syndrome (SMS). Most children with SMS experience chronic otitis media that begins in early infancy and hearing loss (conductive, sensorineural, or mixed) is common. Additionally, oversensitivity to loud sounds is an expressed concern of a number of parents.

Hyperacusis is defined as an oversensitivity to certain frequency ranges or certain sounds that are not bothersome to listeners with normal hearing. Due to the similarities between SMS and other genetic syndromes with known hyperacusis, such as William syndrome, NIH researchers sought to investigate this question further.

The purpose of this study was to determine the occurrence and severity of hyperacusis in persons with SMS and to document the types of responses, triggers and management techniques. Children with a confirmed diagnosis of SMS participating in the IRB-approved natural history study of SMS at NIH (01-HG-0109) were recruited in the United States or at Camp Breakaway in Australia. A 2-page questionnaire originally designed to evaluate the severity of hyperacusis in children with Williams syndrome (Cohen et al, 2006) was mailed to 90 families in the USA (SMS-USA) and distributed to 16 families attending Camp-Breakaway (SMS-AUS). Unaffected SMS siblings of the USA group served as a comparison (control) group.

Results: Preliminary findings are summarized below based on analysis of 83 returned questionnaires including: 47 (29M/18F) from SMS-USA, 16 (8M/8F) from SMS-AUS and 20 (13M/7F) from the sibling control group:

Sensitivity to loud sound (hypercusis) was present in 78% of the total SMS group compared to only 10% of the sibling control group (Fig 1). While there was a slight difference in the frequency between the SMS-USA group (74%) and SMS-AUS group (87%), this difference was not significant. Thus, in this study, hyperacusis was about 7 times higher in the SMS group.

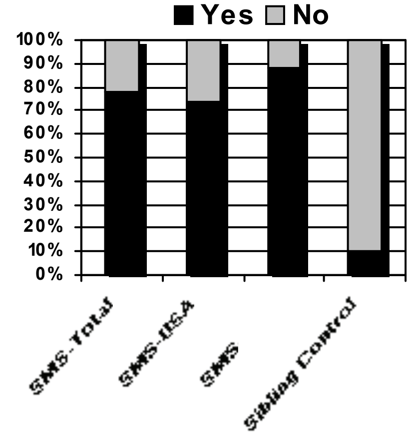
While the majority (59%) of parents indicated that their child's intolerance for loud sounds remained unchanged over time, 33% felt that it had improved and 2% felt that it had cleared up.

Triggers and Response to Loud Sounds

The major trigger for distress was tiredness (47%) followed by mood (29%). Tiredness was also the most often cited reason for a heightened reaction in 50%. Common behavioral responses to distressing sounds were covering the ears with hands (88%), becoming upset (58%), or displaying anxiety/tension (52%). Self-injurious behaviors (head banging, biting self), common in SMS, were triggered by loud sounds in 28% of the SMS group.

Parents reported the most severe reations for sudden environmental sounds such as fireworks, balloon burst, and a sudden shout. There did not appear to be a relationship between the presence/absence of hyperacusis and the child's degree or type of hearing loss.

Fig. 1: Hyperacusis in SMS



…...Continued on page 15

Mystery of the Auction

May 11th

My name is Noah Nuthin, ace reporter for the Reston Virginia Times and Country Diner. I am here at the Reston Hyatt to investigate a phenomenon known as the SMS Auction. Apparently there are some shenanigans going on in the Lake Fairfax room and I am here to break this story.

8 am, Friday

I walked into the room that was filled with amazing items. Books, toys, gift certificates, autographed collectibles, household goods and so much more. Where did these items come from I wondered as I walked around. I tried to be inconspicuous as I marveled at the array of wonderful items available for people to bid on. But to no avail. Some loud fellow walked up to me, shoved a pen in my hand and demanded that I bid. Okay, to blend in, I put down a bid on the Live with Regis and Kelly tickets. That Kelly sure is cute and I could probably write it off as a business expense but before the ink was dry, some lady named Rochelle marked my name out and put hers in there. With a sour face I moved on until I bumped into a very nice lady named Claudia. She was one of the people in charge of the auction and seemed willing enough to talk so I primed her for information. Seems this auction was part of a larger effort to raise money for Smith-Magenis Syndrome.

Claudia explained that the hundreds of items in the room were given by donors and all proceeds went to PRISMS (an organization that advances SMS through research, information and support). I moved on quickly, interrupting the nice lady Claudia because that loud, funny looking fellow was coming back around, telling me to "Bid early and bid often!" As I passed the Nintendo, Ipod, wine, books and John Deere clothing, something caught my eye. Basketball and football Jerseys. Being the reporter I tell everyone I am, I eased up for a closer look. THEY WERE AUTOGRAPHED! Did I yell that out loud? Wade and Manning. I quickly put my name down on both jersey sheets and walked away. As quick as I could say FUMBLE, that Rochelle person marked my name out on both sheets, put hers in there and walked away with a smirk. I wrote down on my pad to find out which hotel room she was in and glue the door shut later that night.

I moved around the room, noticing the books, food gadgets, deer antler wine rack, Autographed Princess Diaries DVD from that pretty girl who was the star, gift certificates from restaurants, hotels, airline vouchers, more DVDs, candles and so much more. It was time for me to leave because I was hungry and that loud fellow was heading my way again.

11am Saturday – May 12

I got a late start because someone glued my door shut. I think it was that Rochelle person but I can't be sure. As I walked in, that loud fellow yelled at me to bid. I did for a ticket to see David Beckham play soccer. I looked for that Rochelle lady but she was across the room, looking sad. Apparently there was a new bidder in the jersey war. Some fellow named Percy and he was taking Rochelle to the mat, figuratively speaking. I smiled as Rochelle walked by mumbling something about getting another case of glue.

LATER SATURDAY –

As I entered the auction room, there was a buzz going on. Diamond jewelry was being bid on, airplane tickets flew, and there was a raffle. The place was electrified as people positioned themselves for the last bid. That loud fellow counted down the minutes and people started to bid. 5,4,3,2,1 and the auction was over. As I later walked away after paying for some lovely yellow candles (for my wife), I heard the total for the auction reached close to $9000. What a great auction and such nice people… except for that loud fellow.

5K Race for PRISMS

On Saturday August 25, seven SMS families met in Springfield, Ohio for the first annual 5K Race for PRISMS. More than fifty runners and walkers helped to raise over $1000.00. Every participant received information about Smith Magenis Syndrome.

A few of "our kids" with the Ohio National Guard

A family carnival was held in the afternoon with food, games, and prizes. Between the 50/50 drawing, raffle, and auctions, another $3000.00 was raised for PRISMS. Best of all our kids were able to enjoy a "normal" day. The Police Department fingerprinted and made identification cards for all the kids. The Fire Department held fire safety classes and the kids learned how to exit a simulated smoke filled room. The Ohio National Guard performed special maneuvers in the Hummer for all to watch. Our kids entertained the crowd at the carnival games tent with their winning one-liners. They bounced around in the inflatable barn and were tattooed with flowers, horses, and rainbows in the face painting booth. Removing the usual triggers helped the kids stay calm. So many friends and relatives told me how surprised they were at how few meltdowns and outbursts they saw. It was a record day for Garrett.

Last but not least, we reached a family through the free public publicity received from the local newspaper. Although this family only lives 2.0 miles from our house, our paths had never crossed until the morning of our fundraising event. They were given the newspaper article from a family member and went to the PRISMS website for more information. They came to the carnival with their fourteen year old daughter and the "SMS family-experts" agreed that she probably has Smith Magenis Syndrome. On Monday, 2 days later, the parents requested a FISH test from their pediatrician, and received the official results that their daughter did indeed have SMS.

Thank you to all the SMS families who came and worked a shift. A Special Thanks to our co-event planners, Robyn and Harold Foster for their blood, sweat and tears. A special appreciation to Jeri Gawlowski, and Joe and Michele Zdanowski, for all the help and the hundreds of answered questions in the months leading up to the fundraiser. And finally, thanks to the families that kept the game tent running smoothly; John and Kellie Cooney, Faith Nobilucci and Billie Sparks, and Katherine Talent and her children.

Charlie and Tina McGrevy

If you have hosted a fund raiser for PRISMS and wish to share your success please contact the newsletter editor with information. If you would like ideas on how to organize a fund raiser, please contact Percy Huston (percy@prisms..org).

Taylor and Jeremy

Stories of connecting through PRISMS

After our three year old daughter was diagnosed with SMS, I obsessively researched everything I could about SMS! I think I read every word of every article and every story on PRISMS. And when I saw that there was going to be a conference 6 months from that time, I tried to sign up right away. However, I was ahead of schedule and had to wait a couple of months to register. My husband Ken and I had several reasons we wanted to attend the conference. One of those reasons was that we were hoping to meet other families from our state (Illinois) that might be parents of children close to our daughter's age.

The first day of the conference I noticed a couple holding a newborn and was curious if she had been diagnosed at such a young age. When I struck up a conversation with them, I learned their newborn did not have SMS but that their two-year old daughter did. They were the first couple I had met with another SMS toddler. And though they were from Kansas I still found a comfort in talking with them. That night we met two other families from Illinois. And found out that they knew of two other families also from Illinois but all the children were ages 10-15. So, we were somewhat disappointed. So, our focus turned to trying to connect with parents of toddlers.

The next day, as Ken and I were walking to our room we saw a little boy with SMS and his mom entering their room. I was so excited to see another toddler with SMS that I introduced myself. I learned that Susan and her husband Paul were from Los Angeles and their sweet little boy was two years old. He had been diagnosed around the same time as our daughter. And to our surprise, we learned that Susan is actually from the Chicago area!

Though we could have asked several more questions, Susan was holding a very sleepy-eyed little boy and we knew it had to be naptime. So, I hate to admit it but I got the address list and put a star next to Susan and Paul's names. I wanted to make sure that if I did not see them again at the conference, that I at least had a way to contact them.

They were the first couple I had met with another SMS toddler.

Thankfully, we were able to talk a few more times. And then Susan asked how far we lived from Chicago. When I said about two hours, she mentioned that they were coming back to the area in a month to visit relatives. Susan asked if we would be interested in coming to visit them while they were in Chicago.

I believe I emailed her the day we got home from the conference just to make sure she knew how to contact me. She replied with the dates that they were coming to Illinois and asked if we would want to celebrate their son's 2nd birthday with them and their family. (Oddly enough we were having our son's 2nd birthday party the day before).

We were happy at the opportunity to get to know their family better. And even more excited that Taylor would get to meet another toddler with SMS. Not to mention their siblings would also get to meet someone going through similar experiences. From the time that we arrived, Taylor and Jeremy kept looking at each other. Both of their siblings were there but even with all of the commotion and distractions, I noticed that their focus always seemed to come back to each other. Neither one of them had a good nap so were quite tired (imagine that), so they did not play together a lot. I think had they not been so tired they may have been more interactive. They sat by each other for a while just taking each other in. They seemed quite content just looking at each other. Even though they are very young, I think they still have a sense that they are similar.

I think for Ken and I there is just an unspoken bond between our families. And it is almost more comforting to be able to go through the experience with another family where

you will face the same obstacles around the same time. I think of Susan and Jeremy at every mealtime. Because I know as I am fighting to get through one meal at a time without tantrums, etc…. Susan is doing the same thing in California.

And though it would be even better if we lived closer together. Email and phone calls still allow us to give each other support in the challenging times and cheer for each other with each stride our children make. Our hope is that we will be able to make it an annual tradition to get together and allow our children the opportunity to have a friend that understands them.

Though there are several opportunities available at the conference, to our surprise, connecting with other parents was the best gift that we received!

Shannon LaFave

This past February, our son Jeremy, was diagnosed with SMS at 19 months of age. After a year of tests and doctor's appointments, we finally had our answer to his development delays, short stature, low muscle tone, and sleep disturbances. In the first days after his diagnosis, we learned about the SMS conference in Reston from the PRISMS website. Our lives had been turned upside down, and we hoped the conference would give us a greater understanding of the challenges ahead. We knew we'd be meeting other parents and families, but we didn't realize that making these connections would be the most positive aspect of the conference for us.

I first met Shannon and Ken in the hallway of the hotel. We saw each other's badges and introduced ourselves. I was immediately drawn to the fact that they were from Illinois. Though I now live is Los Angeles, I was born and raised in a suburb west of Chicago and still have family there. The next day at lunch, I mentioned to Shannon that we'd be visiting my family to celebrate Jeremy's second birthday the following month. Shannon said they could drive up so see us! I couldn't believe that someone I had met the day before would drive four hours (two hours each way) to spend an afternoon with us!

In the emails that followed between Shannon and I, we finalized our plans to get together. Shannon, her husband Ken, her daughter Taylor (age 3 - SMS), and her son Blake (age 2) would drive up to celebrate Jeremy's birthday at a small family barbeque with us. (It turned out that Shannon and Ken have close friends in the next town over, so even if our barbeque was a bust, they had backup plans - ha!)

We were so excited to meet another SMS child that was close to Jeremy's age.

We were so excited to meet another SMS child that was close to Jeremy's age. By the time they arrived, Taylor and Blake were weary from the trip, but played eagerly alongside Jeremy, my 5-year old daughter and my 2-year old nephew. It was a great opportunity for us to share stories, discuss issues, and get to know each other. We hope to make this an annual event and watch Jeremy and Taylor grow up together. Thank you PRISMS for helping us make these connections!

Susan Diamond

Save the Date

PRISMS Sixth International Conference

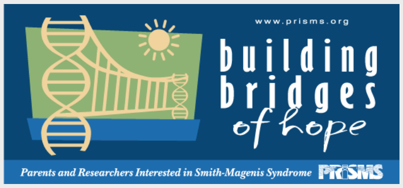
"BUILDING BRIDGES OF HOPE"

Sept. 17-20, 2009

Reston Hyatt Hotel

Reston, VA

*This is the same site as the 2007 conference. Our families loved the location and were able to easily enjoy the shops and restaurants in immediate walking distance. We hope to see you all return AND we welcome new families and faces to the 2009 conference. Start planning and saving now! Be a part of the celebration!!



Last year "Chicken Soup for the Soul" publishing was looking for stories for their "Special Needs" book. Congratulations to Tina McGrevy for submitting a story which actually made it into a page layout format, but was cut in the final editing. Although her story didn't make it into the book, Tina is receiving some glory as her story is being highlighted on the website, www.soulsupporter.com.

What's Your Fundraiser?

Who is your United Way charity?

Across the United States the **United Way Campaign** supports charity organizations. You can write PRISMS in during your United Way Drive.

Who is your online shopping source? Register with **iGive.com** and let PRISMS benefit from your purchase. Shop online through iGive at over 680 stores. Shopping and donating made easy.

Who is your internet search-engine?

Make **Goodsearch** your internet search source. With every search raise money for PRISMS.

Find more information about these fundraisers go to the PRISMS website **www.prisms.org**.

The New 2008 Entertainment Books are now available!

DINE, SHOP, TRAVEL, and SAVE

Last year PRISMS earned nearly $500.00 and sold 79 books.

This year for every book sold over the 79 of last year, PRISMS will earn a bonus profit of 50%. Help PRISMS sell over 80 books and increase their Entertainment Book fundraising this year!

Just buy one book and sell one book too!

The Entertainment Book is available for over 150 cities throughout the United States and Canada. The easiest way to preview and purchase your own local book is at www.PRISMS.org and click on the Entertainment Book link. As you go through the purchasing process you will receive the discounted group price and PRISMS will automatically receive 20% profit of each book purchased. If ordering 3 or more books, contact me directly to save on shipping fees.

Michele Zdanowski Fundraising Volunteer mzdanowski@comcast.net (248) 437-7251

United Way Donations

As you may know, now is the time of year that most companies begin their United Way campaigns. Each United Way chapter lists a variety of organizations that may be chosen for payroll deductions, most of which are locally based.

If your local United Way chapter accepts write in donations (most do), the 501(c)(3) non-profit status of PRISMS should qualify us for the write-in option.

Please consider asking your fellow employees to join us in our struggle to solve the puzzle of SMS. Many folks give generously to the United Way and this is a great way to spread awareness and increase our funding. Most co-workers are eager to donate to a cause that is personally tied to a colleague. You can easily direct them to our website (http://www.prisms.org) to learn more about SMS and the mission of PRISMS.

Here's all of the information you'll need:
PRISMS, Inc.
21800 Town Center Plaza**,** Suite #266A-633
Sterling, VA 20164
Tax ID 54-1652029

To obtain a write-in campaign in your workplace, contact your company's United Way chairperson or your local United Way agency to ask if there is a write-in or designation option. Or, you might see the option on your pledge card.

If you need to contact your local chapter, there is a search tool by postal code at the top of the United Way national page. http://national.unitedway.org/

Thanks in advance for your continued support. Together we are making a difference in the lives of our kids!

The Question Box

Question: *What strategy can I try to keep my child from picking at sores on his hands and arms and creating new sores?*

Answer: Since our children are so” visual”, any little teeny, tiny paper cut or small mosquito bite that they can ***see,*** can become a huge and ongoing sore that never heals, scars, or can get infected. They also pick because they crave the sensory input, and have a reduced sensation to pain. Skin picking can be a huge problem and hard to deter. One method that we have used at home, is to have a constant supply of “medical” non-latex gloves on” hand” at all times. (Sometimes we find them in colors). My daughter will wear them when she is in a “picking” cycle, and it helps to hide any “boo-boos” or “ouchies” she can see, but also provides the sensory input that she is craving. We have also alternated with white cotton gloves, (that can easily be washed…don’t machine dry them), and also “silk” type gloves, when we can find them. She will wear the gloves out in the world, when she needs to, or just at home to decrease the need to pick. It has been a huge help for her, and enables her to **self-manage** her picking, as **she** will go put on her gloves, when **she** feels the need. I don’t have to get involved, and it does reduce the amount of picking and the anxiety associated with the skin picking.

Margaret Miller, parent of Deirdre

The Question Box is a new feature of Spectrum. Please email your questions/suggestions/answers to notfar@earthlink.net
Fax: 843-986-9369

Question:

“What can I do to keep my child safely buckled up in the car?”

Please share your knowledge!!! Email your suggestions to notfar@earthlink.net

WANTED:
PRISMS SECRETARY BOARD POSITION

PRISMS is currently seeking to fill the Secretary Board position. Rochelle Wright, our current Secretary, will be stepping down at the end of this year, or as soon as the position is filled.

We are seeking to fill this integral position as soon as possible and we are anxious to see our membership stepping forward to become part of the PRISMS Board. This is a wonderful opportunity to volunteer your gifts, talents and time, while sustaining the mission of PRISMS and celebrating all individuals with Smith-Magenis Syndrome.

We ask that applicants be "current" PRISMS members, (dues are current for 2007). We have listed the job description and hope you will give the position great consideration. PRISMS needs your help and we welcome your interest and abilities!

PRISMS Secretary Responsibilities

The PRISMS Secretary is charged to:

Take minutes on all monthly teleconferences. We have teleconferences every first Wed. evening of the month. Everyone on the board is expected to attend.

Making sure Share 360 is up to date and cross checking Share data with board members, (Share 360 is an online repository that holds pertinent PRISMS documents and historical records and accounting), and updating by-laws and other governing documents.

Take minutes for our 2 face-to-face board meetings each year, at a designated site. (We have our board meetings usually in a town that accommodates one or more members of the board, so we save money on airfare and travel.) The meetings involve arrival on a Friday and departure late Sunday afternoon.

Preparing and distributing itineraries for the teleconferences and board meetings with input from the board.

Helping to prepare any planning/making arrangements for the board meeting.

Monitoring the PRISMS Bulletin Board; daily review of new users, and approval or denial of new users, and triaging emails as needed.

Supporting other board member positions as needed.

Term of service:

The term of service is **three** years and ends at the last day of the designated year.

PRISMS

Parents and Researchers Interested in Smith-Magenis Syndrome

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Sterling, Virginia 20164
Tel: 972.231.0035, Fax: 413.826.6539
Website: http://www.prisms.org
Email: info@prisms.org

BOARD MEMBER APPLICATION

Name ____

Date ______________________

Address ____

City, State, Zip____

____

Home Phone ______________________ Work Phone________________________

Cell Phone ________________________ Email Address______________________

Occupation __________________________

Please state why you are interested in serving on the Board of Directors:

____

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Each Board member has a specific responsibility within the organization. What do you see as your role, scope of work, job description on the Board?

____

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____

What specific talents and experience do you possess that would be an asset to our Board?

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How much time do you realistically see yourself giving? (We are committed to 2 in-person board meetings per year and monthly telephone conference calls in addition to our individual duties)

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What specific goals and ideas do you have for the future of PRISMS?

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Additional information

____
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____

____
Signature

Please return this application to the PRISMS office, 21800 Town Center Dr., Suite 266A-633, Sterling, VA 20164.

If you have any further questions, please contact PRISMS at info@prisms.org.

SMS SuperKid!

LAUREN KURTZ

My family has been involved in soap box derby racing for the past 40 years. Just about all of my cousins, my brothers, and myself have raced in our local home town race in the 70's. My Uncle is very involved in the organization and has been our local derby director for years.

After having twins my Uncle told me he would have car kits waiting for us when they were 8 years old, the age you need to be to race. As the years went by my son Michael, Lauren's twin brother started racing.

Lauren would go along to the races to watch her brother and would always ask me if she could race. I knew that Lauren would not be able to race by herself. It would not be a safe situation! I later found out about a race that they have for children with special needs. It is called the National Super Kids Classic (www.nationalsuperkids.org).

They use specially designed cars that can accommodate two drivers. The child with special needs is paired with a co-driver (an experienced soap box derby racer). Inside the car are 2 steering wheels and a brake. The co-driver does all the driving. The steering wheel for the special needs driver is not hooked up to any cables, so they think they are doing the driving!

SuperKid Lauren!

Last October Lauren raced in a town about an hour from us that had this special race. There were 17 cars and she came in 6th place. She had a great time. I ended up getting on a committee in our local county to get a race started in our home town. We tried to get it going last June, but ran into some road blocks. Our first race is going to be on September 22, 2007 in our home town.

Since our local derby already paid to send a child to the National Super Kids Classic, they approached me to see if Lauren would represent Bucks County in the National Super Kids Classic in Akron, Ohio. At this event local champions compete against other local champions from all over the United States.

This was such an honor, so of course I said yes. It's every derby racers dream to win a local race and make it to Akron to race in the All American Soap Box Derby.

Lauren had such a good time and was so proud of her self. She finished 7th out of 56 racers. She went down the hill in Akron more than I did when I was a child! The racers attended an awards banquet and everyone got a trophy.

Lauren is so proud of her trophy and has it displayed in our house right next to her brother's. Our local newspaper came out and interviewed Lauren…she felt like a celebrity. It was a great experience and we hope we will be returning next July!

Beth Kurtz

Do you have a SMS SuperKid?
Send a story with pictures to
Julia Hetherington at
notfar@earthlink.net
Fax: 843-986-9679

PRISMS would like to thank the following individuals who have made contributions in the last year (Sept 2006—August 2007). PRISMS relies on the generosity of its supporters to continue the pursuit of our mission.

Many Thanks.

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Continued from page 1….NIH treatment study

Part 1 Inpatient Trial: involves two separate inpatient NIH admissions of 2-3 nights each for the bright light and melatonin treatments. During the month before inpatient admission, participants will be asked to wear an actiwatch (wrist-watch like device). Parents will be asked to rate the child's behaviors, mood, and sleep patterns and obtain frequent body temperature measurements.

During the bright light treatment, children are asked to remain in their hospital room for the periods of controlled light treatment, but will be able to watch TV, play games, etc. An electroencephalogram (EEG) with additional electrodes to track eye movements is used to monitor the child's attention. The parent will be asked to rate the child's mood and behavior during the trial period. Between 8AM and 6PM serial blood samples are collected to measure melatonin levels and other biochemical markers. Twenty four-hour urine and/or saliva (spit) may be collected to measure melatonin and/or biochemical markers.

During the melatonin treatment children will take a single dose (tablet) of melatonin or placebo at bedtime. During the daytime, EEG electrodes are placed to track eye movements and left on to track EEG-sleep patterns. The parent will be asked to rate the child's behavior and mood as described for the bright light study. Between 7PM and 7AM serial blood samples are collected to measure melatonin levels and other biochemical markers. 24-hour urine may be collected to measure melatonin and/or biochemical markers.

Part 2 Outpatient Trial: is an outpatient trial conducted in the home setting over 11-consecutive weeks using bright light alone and in combination with melatonin (or placebo) tablet. They undergo the same procedures outlined in the pre-trial at-home phase of Part 1 (Actiwatch, behavior assessments, body temperature measurements, saliva samples) over an 11-week period. If saliva samples cannot be collected for melatonin testing, 24-hour urine samples may be collected instead.

Parents interested in having their child participate in this treatment study should contact: Ann C.M. Smith, MA, D.Sc.(hon), Principal Investigator, Tel 301-435-5475 or Rebecca S. Morse, MA, Study Coordinator, Tel 301-451-3085

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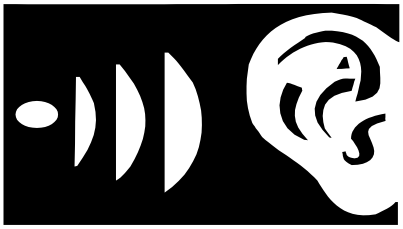
Continued from page 3…..Hyperacusis



Palliative Strategies Tried (See Table 1)

Many parents reported that their child with SMS showed **less** distress when they were prepared or warned of an impending sound. Other effective management techniques included sound reduction through a variety of techniques, avoidance of distressing sounds, controlling known triggers, and familiarization with the offending sound.

Summary: A significant percentage (78%) of children with SMS were reported to have hyperacusis in this initial study. Research is ongoing to characterize this newly appreciated finding in SMS and identify strategies to aid in management. Assessment of hypersensitivity to loud sounds should be included in the audiologic evaluation of persons with SMS. This can be achieved via traditional loudness discomfort measures or by questionnaire. When a problem is identified, parents/caretakers should be counseled regarding palliative strategies.

Table 1: Successful Palliative Strategies Tried

Strategy	Number Reporting
Preparation, explanation, warning	14
Avoidance of sounds	4
Control triggers (e.g., nap to avoid fatigue)	2
Reduce sound level	
Earplugs or headphones	5
Cover ears	1
Turn off hearing aids	1
Increase familiarity with sound	2

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New in this Issue!!

I hope everyone has enjoyed the increased frequency and the format of the SPECTRUM over the previous year. SPECTRUM has two new sections in this newsletter. They are ***SMS SuperKid*** and ***The Question Box.*** **Both require your input**. One of the most frequently heard comments is how important parent support is in PRISMS. We would like to give members the opportunity to let their SMS child shine through ***SuperKid***. It is always good to hear of SMS success. ***The Question Box*** is also about sharing and supporting others. Sharing your knowledge with "been there, done that" experience. Please do not hesitate to contact me if you have something to share. We need both your stories and your questions/answers. The success of these two new sections requires your input!! I can be contacted at notfar@earthlink.net, or by phone at 843-521-0156, or by fax at 843-986-9369.

SPECTRUM Editor in Chief, Julia Hetherington

THANKS!

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