

SPECTRUM

July is Smith-Magenis Syndrome Awareness Month

It's time to CELEBRATE and EDUCATE!! We have dedicated the month of July as SMS Awareness Month. Celebrate all the wonderful attributes and talents of our SMS'ers while educating our community, school, churches, medical facilities, families, friends and strangers! Let's take our mission "to the streets," and teach the world about SMS. Spread the joy that is SMS and create a community of greater understanding, acceptance, and support. Let's all contribute to the building blocks of our "Bridges of Hope," and help design a future that readily understands Smith-Magenis Syndrome and regards our children as precious and important!

To kick off a first year of the SMS Awareness Month, we are having a contest! The winner of the best SMS awareness photo will receive one free night of lodging at the 2007 conference in Virginia. We would like you to send in photos, (digital is best), of all your family, friends, neighbors, etc., dressed in their grandest PRISMS wear. Any photo that shows your family or community spreading awareness of SMS is eligible for the contest. Here are some other ideas:

- ✦ Pass out PRISMS brochures in your church or school, or place of work.
- ✦ Present a lecture or power point presentation about SMS at your child's school.
- ✦ Hold a coffee for neighbors to educate them about SMS and your child.
- ✦ Get the siblings involved! Perhaps the sports team they play on would love to show their PRISMS style. (Real hockey players do wear pink wristbands!!)
- ✦ Plan a get together with other PRISMS families in your area and celebrate your children.
- ✦ Wear your best PRISMS wear, have a party and rejoice in your SMS child! Let them know how truly amazing they are!

Send us your photos and/or stories via email or snail mail the photos to the PRISMS address. Photos will be accepted until September 1st, 2006. We hope to feature the top photos/stories in our fall newsletter. So take to the streets and "say cheese" and spread the message of SMS!

PRISMS

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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

Another year is here and I feel privileged to be the President of your Board. I am looking forward to an exciting year with a strong Board, great volunteers and a great professional Advisory Board to meet the needs of all our families. Many events and activities will be going on throughout the year and I hope each of you will get involved and participate in the efforts in your area.

Is your family planning to be a part of SMS Awareness Month in July? I hope you will. We're excited that the opportunities that exist to get the word out about SMS.

Please take a minute to complete the enclosed membership form. Only a fraction of our folks have paid their dues thus far. Ugh! We need to update our contact information, and we need the support of our families to continue our work educating and supporting families with persons who have SMS. With your support we can hopefully raise awareness and educate people about SMS. Also, think of ways you or a group of parents in your area could have a fundraiser and raise awareness as well as funds for SMS. Golf tournaments have been great fundraisers in the past as well as a great time for golfers to get together for a great day of golf. Check out some of the creative fundraisers in this newsletter. The more people we can reach to raise awareness and educate about SMS the more we will be able to continue the research in to SMS.

The Board has started working on the next conference which will be held in Reston, VA on May 10-May 13, 2007 at the Hyatt Regency. It's going to be terrific!

Jay Maiher has decided to resign from the Board, and I would like to thank him for all his time and energy he devoted to our organization. We are actively seeking new board member applications from our 2006 members. Be on the look out for a board application.

SMS Awareness Bracelets Order Yours Today!

These “hi-viz” pink silicone bands are a great way to build awareness about SMS. Bands are available in adult and youth sizes with an order form on the [PRISMS website](#).

*12 bands - \$25.00
24 bands - \$45.00
36 bands - \$60.00
48 bands - \$75.00

*Please note that singles are only available at the conference.



All prices are US Dollar and do not include a \$4 shipping fee. We take Visa and MasterCard too!!

2006 Membership Drive

WE NEED YOUR HELP !!! As a new year dawns, the need for continuing education and research for families with children with SMS is greater than ever. The PRISMS 2006 Annual Membership Drive continues its efforts to help raise funds to continue its mission to provide information and support to families of persons with Smith-Magenis Syndrome (SMS) and fostering partnerships with professionals to increase awareness and understanding of SMS. Please help us by filling out the enclosed membership form and updating your information. Please return your form ASAP.

Save the Date! May 10-13, 2007
PRISMS Conference - [Hyatt Regency Hotel—Reston, VA](#)

First Annual Parents Night Out

The Baylor Pediatric Student Association held its 1st Annual Parents Night Out on November 4, 2005 at the Texas Children’s Hospital In Houston, Texas. This was held the night before the TX/LA Picnic in Houston. Four SMS families participated and it was a great experience for the students and kids.



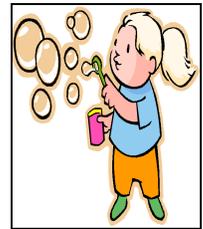
TX/LA 2nd Annual Picnic in Houston, TX

The 2nd Annual TX/LA Picnic was held November 5 at Bear Creek Park in Houston, TX. There were around 20 people in attendance. The weather was great and the kids played on the playground equipment. Everyone brought a dish and there was plenty of food to enjoy. It was a great day and lots of fellowship and sharing of experiences as SMS families. **The 3rd Annual Picnic will be held Saturday, October 28, 2006 at Bear Creek Park in Houston, TX. Please save the date!**

Understanding Sensory Integration Dysfunction

Nancy Cordrey, mother of Caroline, age 11 with SMS

We were blowing bubbles and chasing them when my two year old daughter, Caroline, fell on the driveway and skinned her knee. She let out a noise of discomfort when she hit the pavement, but was completely calm by the time she stood up. The adults who witnessed the fall and watched the blood flowing down her leg, declared her brave, but I knew at that moment that she genuinely didn't feel the pain.



Before Caroline turned three we would learn that she had Smith-Magenis Syndrome (SMS), and that one characteristic of the syndrome was desensitization to pain. It was many years later, however, that I came to understand that her response to pain was just one aspect of an overall condition known as Sensory Integration Dysfunction, or SI Dysfunction.

“Sensory Integration is the organizing and processing of sensory information from the different sensory channels and the ability to relate input from one channel to that of another in order to submit an adaptive response,” (Ayers, 1972). Sensory Integration Dysfunction, then, occurs when a compromised nervous system is ineffective in processing sensory information.

There are seven different sensory systems that can be adversely affected by SI Dysfunction: Five of these come from the five basic senses: oral, olfactory (smell), auditory, visual, and tactile (touch). In addition, we have the ability to feel our movement in space, which is our vestibular sensory system, and to feel pressure, which is our proprioceptive sensory system. Individuals can be affected in some or all sensory systems and in varying degrees.



Persons with Smith-Magenis Syndrome may have significant SI dysfunction. My daughter, Caroline has displayed SI dysfunction in nearly all sensory systems. Children with SMS are likely to be told they have motor planning issues, visual tracking problems, tactile defensiveness, over sensitivity to light or sound. These symptoms are rooted in SI dysfunction. Some of the behavior problems associated with SMS can also be traced to SI dysfunction.

Certainly there are physiological and biological differences in individuals with SMS that impact learning and behavior. Not all the challenges caused by SMS are due to SI dysfunction, but addressing SI dysfunction can improve our understanding of the syndrome and allow us to better support those struggling with it. Looking at the behavior of individuals with SI dysfunction provides clues to their sensory deficits. Insensitivity to pain has been associated with SMS and is a sign of tactile dysfunction. Children with SMS are known for self-hugging. Squeezing themselves provides proprioceptive (pressure) input. This suggests they do not have proper proprioceptive integration. Motor planning problems, and verbal dyspraxia demonstrate issues with vestibular integration.

It is important to realize that SI dysfunction can be seemingly inconsistent. A child may love it when you scratch their back but hate it when you comb their hair. Aspects of their system may be at one time overly sensitive and under sensitive at another. Occupational therapists, developmental audiologists

and developmental optometrists can help identify your child's SI issues and provide various therapies to address SI dysfunction.

Auditory issues may be common in individuals with SMS. Auditory processing problems, over sensitive hearing, and an inability to ignore background noise are some of the issues persons with SMS may face. Developmental audiologists can give hearing tests that check hearing at various frequencies to give an increased understanding of what a child is actually hearing. Specialized music therapy can be used to help improve these weaknesses. This type of therapy, usually administered by occupational therapists, can have several different names: auditory integration therapy, listening therapy, and tomatis are just some of the names of programs aimed at improving auditory dysfunction.

Visual problems may be found in those with SMS. A lazy eye, tracking difficulties, poor depth perception and light sensitivity can come with SMS. Developmental optometrists are trained to provide vision therapy that can improve these problems and have a positive impact on reading, catching, and other perceptual skills. Tactile dysfunction prevented my daughter from feeling the pain of her skinned knee. She often rubs her hands together to give herself the sensation she lacks naturally. Occupational therapists are trained in SI therapies that address the tactile sensory system. An occupational therapist may include therapeutic brushing to improve this condition.



Occupational therapists can also use SI therapies to address vestibular dysfunction. Gravitational insecurity, motor planning difficulty, poor balance and low muscle tone are symptoms of vestibular dysfunction. Swinging and jumping on a trampoline are a few of the activities commonly used to work the vestibular system.

Proprioceptive dysfunction is also addressed by occupational therapists. Carrying heavy objects, wearing a weighted vest, and other activities that deliver deep pressure are used to address proprioceptive dysfunction.

SI therapies can improve an individual's ability to process sensory information, but generally therapies require long-term treatment. A daily "sensory diet," (individually designed and prescribed by an OT,) incorporates therapy into the individual's daily routine and is often the most affective way to help patients cope with SI dysfunction.

At the recent SMS conference, I set up a display as part of the Parent Expo to make parents more aware if SI dysfunction and the therapies available to treat it. The need for such awareness was made clear when several parents of older children with SMS testified that SI therapy was the only thing that had a significant impact on their child's behavior. SI dysfunction is receiving increased attention. Training in SI therapies has become standard for occupational therapists. Vision therapy is also becoming more widely respected. These advances will make it easier for individuals with SMS who suffer from SI dysfunction to receive therapy and experience an improved ability to interact with their environment. The book, [The Out of Sync Child](#), by Carol Stock Kranowitz, is a great resource for further information on SI dysfunction.



THANKING YOU FOR YOUR GENEROSITY!

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Jeri Gawlowski has put together Memorial Packets for friends and families who have lost a loved one and wish to make a donation to PRISMS. You can contact Jeri at the following numbers or email her, and she will overnight you the packet.

Work: 734-522-0800

Home: 248-446-1094

Cell: 248-706-2434

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A special thank you to the Xerox Reproduction Center at Bank of America for their generous donation of paper and printing for this newsletter.

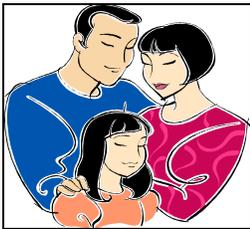
Understanding Patterns of Sensory Processing and Supporting Daily Activities in School Aged Children with Smith-Magenis Syndrome: An Occupational Therapy Perspective

Hanna Hildenbrand, MS, OTR/L and Ann C. M. Smith, MA, DSc (Hon)

Hanna Hildenbrand is an Occupational Therapist in the Rehabilitation Medicine Department of the Mark O. Hatfield Clinical Research Center, National Institutes of Health. She is a member of the Smith-Magenis Syndrome interdisciplinary team led by Ann Smith at the NIH. Hanna serves as a clinician and associate investigator with the ongoing natural history study of SMS (protocol 01-HG-0109).

This article is based on an oral presentation at the 4th International Conference on Smith-Magenis Syndrome, in Cincinnati, May, 2005.

The daily life experience of children with Smith-Magenis Syndrome (SMS) is unique to each family, and it is the parents or primary caregivers that have the best insight into their child's special challenges. Many of these parents have shared their experiences and, through their children's participation in the ongoing natural history study of SMS at NIH, have provided us with a greater awareness of the characteristics common to SMS that impact participation and performance in daily activities. Our clinical expertise and insights gained from ongoing research serve as a guide for the following discussion on clinical approaches, programs, and resources that may help children with SMS engage, adapt, enjoy, and succeed in daily life.



The Occupational Therapist (OT) has a unique role in the interdisciplinary plan of care for the child with SMS. Through consultative and ongoing services, the OT facilitates participation and performance in daily activities, social interaction, and academics. These OT services may include programming to address sensory processing difficulties and challenging behaviors, recommendations for environmental modification and activity adaptation, school readiness and skill development, splinting, and IEP/504 development and planning. In keeping with the purpose of this brief report, programming to support sensory processing and adaptive behavior will be highlighted.

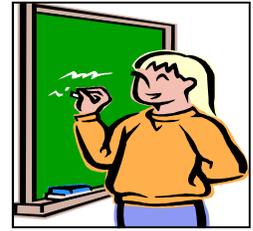
A Preliminary Understanding of Sensory Processing Issues in SMS

Sensory processing difficulties are a major issue for children with SMS as shown by significant differences found in the Sensory Profiles for 21 children (age 3 to 10 years) with SMS (Hildenbrand H, Furst G, & Smith A CM, 2004; Hildenbrand H & Smith A CM, 2005) as compared to children without disabilities (Dunn, 1999). Sensory processing is defined as the receiving and managing of sensory information which contributes to the ability to react to sensations (Miller, Reisman, McIntosh, & Simon, 2001). Children with problems processing sensory input (commonly referred to as Sensory Integration Dysfunction) may present with *hyper*, *hypo*, or *fluctuating* response patterns (Lane, Miller, & Hanft, 2000). The child that is *hyper* responsive will appear to have an increased level of sensitivity or be defensive to common sensations. For example, he/she may "overreact" to being touched by a peer, hearing an overhead announcement, or smelling the school cafeteria food or the teacher's perfume. The child that is *hypo* responsive may seem to need or want intense and ongoing stimulation. He/she may touch almost everything in his/her immediate path, constantly fidget with objects, or jump off of high surfaces. The child that exhibits *hyper* responsive behaviors sometimes and *hypo* responsive behavior at other times would be considered as having a *fluctuating* response pattern. A child with sensory processing difficulties may also appear to have difficulty with motor skills, seeming to be clumsy or uncoordinated. This child may have difficulty playing catch, cutting and writing, biking, jumping rope, or playing games that require imitation. He/she may also prefer sedentary activities such as playing video games, looking at books, or watching television.



Regardless of the pattern of response, daily activities, social interaction and academics can be impacted by difficulties in processing sensory information. The child with sensory processing difficulties may miss important sensory information

in context to the activity or interaction to which he/se is supposed to be attending. The child presenting with additional motor difficulties may expend so much effort to perform foundational or supportive motor skills, such as sitting for extended time, copying from the chalk board, or imitating a movement, he/she has less attention for important information or instructions related to the task at hand. If the child misses this essential or instructional information his/her learning and opportunity to develop related skills will be impaired.



While it is important to recognize the potential impact of sensory processing difficulties on daily activities, interactions and behavior, one must also acknowledge that there may be a blend of behavioral and sensory issues. The child with SMS often exhibits challenging behaviors such as self injurious behavior, temper tantrums, impulsivity, aggression, repetitive behaviors, and moderate to severe sleep disturbances, which may or may not have a sensory component. Together, these behaviors can significantly impact the child's adaptive capacity and ability to participate in daily activities, interact with others, and perform in academics and daily activities. The OT experienced in assessing and treating children with sensory processing dysfunction, provides a unique perspective of these difficulties and designs a care plan for guiding adaptation and developing skills necessary for improved participation and success in daily life activities and the community.

Occupational Therapy Assessment and Treatment: A sensory integration approach

To assess the child's ability to process sensory information and use it in a meaningful way, the home and/or school OT completes a comprehensive evaluation. Ideally, the home/clinical and school OTs should collaborate on the plan of care, including the parents, who provide crucial information about their child. A new model of OT intervention used by OTs specializing in Sensory Integration treatment is the STEP-SI clinical reasoning model (Miller, Wilbarger, Stackhouse, & Trunnell, 2002). The acronym STEP-SI (Sensation, Task, Environment, Predictability, Self-monitoring and Interaction) highlights the



various elements or dimensions of this new model based on long standing principles of sensory integration theory. This model provides a structure for the OT to organize information and establish priorities for occupation-based (activity-based) therapy. The focus of the STEP-SI model is to understand the child's adaptive capacity by determining his/her range of arousal and ability to organize behavior. Elements that support and challenge the child's adaptive response are identified and manipulated in order to maximize the child's adaptation and development of skills. As the child progresses, elements are adjusted according to the child's response to the therapeutic activity. In other words, the OT aims to provide the "just-right challenge" (Ayres, 1972) as the child participates in the activity and progresses from one therapy session to another. Successful occupational therapy using this approach may be observed as the child attains and maintains more appropriate levels of adaptation with improvements in organizing his/her behavior and engaging in meaningful activities.

To date, no single treatment has been proven effective for addressing the sensory processing difficulties of the child with SMS. Nor should it be assumed that any or all programs will be equally effective with all children with SMS due to many variables. In addition to a host of environmental factors (eg., school setting, available therapeutic resources, respite care, etc.), the range of developmental and cognitive disability observed in SMS may be a factor in response to care. However, clinical experience and early clinical research findings, as well as anecdotal reports from parents and clinicians outside of the NIH involved in the ongoing care/treatment of the children with SMS permit the following clinical recommendations. The various programs suggested below are commonly used by OTs working with a variety populations having similar functional difficulties. Therefore, from a clinical and functional perspective, these programs may prove to be effective in addressing the difficulties observed in the SMS child.

The Alert Program for Self-Regulation

The Alert Program (AP) for Self-Regulation, "How Does Your Engine Run?" (Williams & Shellenberger, 1996) is a model treatment program that may be particularly useful to the child as well as his/her parents and daily caregivers. This program assists students, parents, and teachers to understand levels of alertness based on sensory integration theory. It teaches participants how to identify, monitor, attain, maintain, and change their level of alertness as appropriate to a task, situation or setting. Participants learn if their engine (alert level) is too high, too low, or just right in context. As the program progresses, they also learn how sensory strategies can be used to adjust their engines as needed. The AP was originally developed for children with learning disabilities ages 8-12 years but is adaptable for other populations and ages. Most important to note is that the knowledge and strategies learned at an early age may continue to be used throughout adolescence and adulthood, as it is likely that some level of sensory processing difficulty will persist throughout the child's lifespan. The AP may be administered to an individual, but may also be administered to a group (or class) and can be an integral part of a class schedule and milieu for children with (or without) special needs. The program is designed to be administered in a series of lessons and is best implemented by an OT with experience in sensory integration techniques. Lastly, authors (Williams & Shellenberger, 1996) note that most children with regulation difficulties will also need additional occupational therapy with emphasis on sensory integration. Therefore, the STEP-SI model is an ideal framework for incorporating the AP in the comprehensive occupational therapy plan of care

The Comprehensive Behavior Support Approach

In recognizing that not all the challenging behaviors observed in SMS are purely sensory based, collaboration with a behavioral counselor and/or a Speech Language Therapist is also recommended. The Comprehensive Behavior Support (CBS) approach (O'Neill, Vaughn, & Dunlap, 1998) is well suited to the sensory integration model of practice and may be integrated with the STEP-SI model of intervention and supported by the AP. The CBS approach is an alternative to traditional behavioral management. With team collaboration and carry over, it can be used to address challenging behaviors and sensory processing difficulties, such as those seen in children with SMS. A variety of challenging behaviors that often occur in SMS include aggressive hitting and biting, self-injurious head banging and hand biting, disruptive screaming, and certain stereotypic behaviors. A main goal of the CBS approach is to understand the function(s) that individual behaviors serve and to identify variables that motivate and maintain them in order to facilitate more appropriate behaviors and skills. This approach recognizes that these challenging behaviors are a form of communication and serve as an attempt to gain social attention, obtain tangible objects, escape from a task/individual/environment, and/or obtain sensory input (Durand, 1990). Sensory needs and stimuli, sleep, task demands, changes in mood, disruption in daily routine, and personal interactions are all examples of variables that may motivate or maintain these behaviors. Based on comprehensive evaluation and team collaboration, implementation of a CBS plan may include a variety of techniques, including sensory strategies, designed to prevent, decrease, or eliminate (substitute) challenging behaviors.



Strategies to effect a change in challenging behaviors

In attempt to reduce or prevent episodes of challenging behavior and provide an environment for more adaptive responses in school, home, and the general community, sensory strategies and environmental modifications can be individually evaluated and implemented by the clinical and/or school Occupational Therapist. Trott (2002) suggests general guidelines (bulleted below) that can serve as a starting point for designing individual programs. These strategies may ease the difficulties experienced by the child and his/her caregivers as more appropriate behaviors are shaped and more adaptive responses occur.

- Adjust groupings and numbers of children/adults
- Avoid or limit time in difficult environments
- Allow the student to have a familiar adult/peer companion during challenging activities or while in challenging environments
- Modify or structure the environment per the child's response or need
- Adjust the schedule per the child's response, providing warnings for changes in routine and sandwiching or pairing preferred activities with undesirable activities
- Provide transition time
- Provide preferred activities/sensory input during waiting and difficult times
- Limit requirements and adjust difficulty level of challenging activities (Trott, 2002)

These are general guidelines which may not necessarily benefit all children with SMS. It is important to seek professional Occupational Therapy assessment and treatment for individualizing and designing the most appropriate program for the child with SMS.

In Trott's guide for caregivers (2002), information on sensory processing and challenging behavior is briefly explained, and a glossary provides clear definitions on technical terminology. Practical suggestions and tools (Behavior Change Checklist) for



addressing challenging behavior, with or without a sensory component, are also provided. As with the learning from the Alert Program and insight gained from the use of the STEP-SI and CBS approaches, acquiring a working knowledge of strategies that impact and shape the child's behavior will assist parents and caregivers in supporting the child on a daily basis. With carry over of an individually designed program and regular use of sensory and behavioral strategies, the child may develop more mature behavior that can lead to more independent engagement in daily activities.

Summary

From an Occupational Therapy perspective, enabling the child with SMS to participate in meaningful activity during the school-age years necessitates supportive services, strategies, and environments within the home, school and general community. This demands the parents' and caregivers' constant creativity, advocacy, and perseverance in the interest of the child with SMS, not only during his/her young childhood, but throughout adolescence and young adulthood. The knowledge, experience, and skills gained from effective treatment and strategies can continue to support the child with SMS as he/she moves from primary to higher education and into the community. With continued integration of this knowledge and supportive strategies into the daily life of a child with SMS, he/she may achieve higher levels of functioning and expand his/her life roles and community participation.



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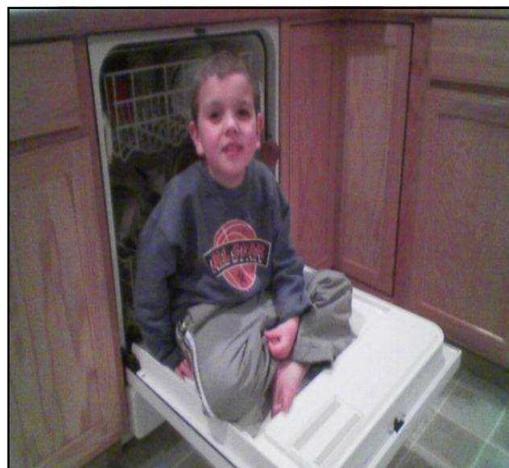
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Cheesecakes, Anyone?

By Sarah Fader

I did a little Christmas Fundraiser for PRISMS. I'm known around here for my cheesecakes and am often asked to make them. So I thought I'd sell them this year for PRISMS (I didn't want to tell anyone I was planning this in case it flopped, I've never tried to sell them before). It turned out to be a good idea and I made \$640 for PRISMS. I donated all of the baking and the ingredients and all of the proceeds are for PRISMS. I'll keep this on going as long as there's interest.

Here is a picture of my son Gavin "helping" me make cheesecakes. I think the dishwasher is a good place for him!



WAL-MART DONATES

Kellie Cooney recently contacted LaVonda Beluiso of Wal-Mart Foundation about making a grant to PRISMS. They made a donation of \$500. Thanks Wal-Mart and thank you KELLIE!!



Parents and Researchers
Interested in
Smith-Magenis Syndrome

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PARENT TO PARENT

Did you know that PRISMS sponsors a parent to parent program? If you need someone to talk to who REALLY UNDERSTANDS what your life is like, try another SMS parent. You may want to find another parent in your area, or perhaps one who has a child the same age as yours. Tell us what you need. PRISMS keeps a list of parents willing to be contacted. We'll send you addresses and phone numbers and then you can talk all that you want. Contact:



Mary Beall

Phone: 972-231-0035

mary.beall@comcast.net

Important note:

PRISMS parent to parent program will only give out information on parents who have agreed to be contacted. If you would like to be added to the list of contact parents, please email me at the above address.

THANKS!

Spectrum is the official newsletter of PRISMS. Readers are free to duplicate all or part of its contents. In accordance with accepted publication standards, we request acknowledgment in print of any article reproduced in another publication. Letters to the editor, comments on articles, and suggestions for future articles are always welcome.

Editors

Susan Voigt

Randy Beall

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Next Spectrum deadline: July 1, 2006