



Parents and Researchers  
Interested in  
**Smith-Magenis Syndrome**

# Spectrum

Volume 6, Issue 3

Spectrum - The Newsletter of PRISMS

Summer/Fall • 2002

*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.*

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**Questions?**

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## PRISMS Recognizes a Special Kind of Giving!



We recognize and appreciate the long hours and effort of our PRISMS Board Members. With a two-year term coming to an end let us pause and recognize our Board Members who are all volunteers and responsible for the operation of PRISMS.

Mary Scully served as Vice President. Mary, we will miss your high energy, leadership skills and creative thinking. Marsha Bach served as Secretary. Marsha got our database up to speed, personalized the information packets that she sent out and chaired the 2002 Conference Committee, among other tasks. We will miss her tireless energy and dedication. Laurie Bellet served as At-Large Member. Although Laurie will not be continuing in her board position she will continue in her role as Resource Coordinator. We thank her for her contributions to the board and look forward to her endless ideas, resources and assistance to our SMS families.

Connie Bessette will continue on as President. Maggie Miller will continue as Past President and consultant to the Board. Randy Beall will leave his position of Treasurer and move into the position of Vice President. Randy and Maggie have been invaluable to the organization. Beyond their technical skills and knowledge of non-profit charitable groups they have been available for our families, our professionals, and all others in need of assistance.

On behalf of the membership a sincere Thank You. It has been an honor to be a member of such a great team!

- Connie Bessette, President

Note: Installation of our new Board Members will take place in November; look for information about each of them in our next issue of Spectrum and on our website.



## Third SMS Golf Tournament

Rain again shortened the third annual SMS Golf Tournament at Tory Pines Resort in Francetown, NH. However, this didn't affect the turnout nor the net proceeds to PRISMS. For the first time there were over 100 participants and over \$6,000 in merchandise was donated for the Silent Auction, raffle and tournament awards. After the payment to the course for golf and meals, the tournament committee passed along \$14K to PRISMS, representing a nice increase over the previous two years despite a faltering economy and a very busy summer (PRISMS conference).

Despite a once in a lifetime storm of 2+ inches of rainfall, fierce lightning, strong winds and hail, all participants made it safely back to the clubhouse. Most players were able to complete at least 16 holes and the tournament was scored through a blind draw on the holes remaining for each group. Although nearly everyone and everything were totally drenched and the Clubhouse was without electricity and telephone, the banquet, awards and auction proceeded pretty much as planned.

*(Continued on Page 2)*

## Who's Who in



### Officers

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Houston, TX

Beth Solomon, M.S.

NIH, W.G. Magnuson Clinical Ctr.  
Bethesda, MD

## Australia's Camp Breakaway

February 7-10, 2003



Would your little SMS'er like to go to camp?

If so, why not take them to Camp Breakaway. Two of our SMS families, David and Georgina Bates and Craig and Katrina Simpson are organizing this event.

Camp Breakaway is set in 20 peaceful acres of beautiful surroundings at the northern end of Budgewoi Lake at San Remo, on the Central Coast of New South Wales, Australia, about an hour and a half drive from Australia's Olympic City, Sydney.

This camping event is not sponsored by PRISMS; however, it is PRISMS' goal to provide you with information related to education and events that may be of interest to families with Smith-Magenis syndrome.

If you would like more information regarding Camp Breakaway, please contact David and Gerogina Bates at -02 43 992 057 or Craig and Katrina Simpson at -03 52 614 820. You can also email the Simpsons at: [simpson.katrina@edumail.vic.gov.au](mailto:simpson.katrina@edumail.vic.gov.au)



## HELP HAS ARRIVED!

PRISMS is pleased to introduce to you our Administrative Assistant, Sue Makowiecki.

Sue comes to us highly skilled in all aspects of office management. She will be working out of our Francestown, NH office Monday through Wednesday each week.

Please feel free to call on her should you have trouble getting something you need. Sue can be reached by telephone at 603-547-8384 or by email at [prisms@prexar.com](mailto:prisms@prexar.com) or [info@smithmagenis.org](mailto:info@smithmagenis.org)

*(Golf Tournament, Cont. From page 2)*

The silent auction forms were destroyed by the storm and the simplest solution was to go the live auction route. The net result was a lively, entertaining event and an even greater return than the previous year. The highlight of the auction was a "tie" between two bidders for a lithograph by SMS parent Frank Gonda. When the bids reached the retail value, Frank interrupted the auction and offered to send each of the bidders a new copy of the lithograph if they both honored their bids!

Once again, our volunteers did a tremendous job, special kudos to Maggie Miller and Catherine Kelly for their lively carrying out of the special fundraising games out on the course. Likewise, Michael Herrity and Paul Duzan ably conducted the putting contest and the result was a terrific splitting of cash in a 50/50 format. Committee volunteers Patty and Dan Dubreuil assisted by Ann and Ron Smith, Maggie Miller, Catherine Kelly and Connie Bessette assembled the goodie bags and box lunches donated by Alexander's Shop 'n

Save Markets and Deli Dynamics. Kelly Kennerson and Kristel Armstrong once again handled the registration table flawlessly. Patrick McDonald, Scott Daigle, MaryAnn Duzan handled the selling of raffle.

Ann Smith's presentation on the complexity of sleep deprivation and current research, and the presence of SMS individuals helping out with the auction gives our tournament a unique insight for the participants. Several players have mentioned that they come away enriched having contributed (to assisting in the SMS greater cause).

A heartfelt THANK YOU to all who participated in the Third Annual SMS Golf Tournament and, also to our major sponsors: Armstrong Golf Architects, New Seabury Resort, Lee and Mason Financial Services and Bayer Corporation.

*- Marvin Armstrong-stepfather of Jonathan Kelly-age 32 and tournament chairperson.*



## SMS Bulletin Board

- The SMS Foundation UK is holding its AGM and Family Day at The Crowne Plaza Hotel, Heathrow, London UK on 16th November 2002. Speakers include: Dr. Carolyn Webber (Abilities and Attainments in SMS), Professor M. Patton, Dr. Orlee Udwin and Olga Ramos-Esteban. Workshops on Occupational Therapy and a question and answer session with various professionals and Contact a Family. A creche will be provided. Further details from Pat van Nederveen, Treasurer ([patricia@van-nederveen.fsnet.co.uk](mailto:patricia@van-nederveen.fsnet.co.uk)).
- United Way contributions are rolling in. Many of your United Way contribution forms will not list PRISMS, so please remember to write PRISMS in as your choice for contributions. You may be asked to provide information regarding PRISMS. The following information is usually the standard required:

PRISMS, Inc.  
76 S. New Boston Rd.  
Francestown, NH 03043  
Tax ID Number (TIN): 54-1652029  
PRISMS is a 501(c)(3) non-profit organization, per the IRS regulations.

- Final arrangements are being made to provide conference tapes from the 3rd International Smith-Magenis conference held in Westminster, CO this past summer. Additional information will be forthcoming.
- Holiday mailing to come out soon - friends and family ideas on giving, "Holiday **SMileS**" contributions to PRISMS.



## Support A Great Cause Get Great Savings

PRISMS is raising funds by selling 2003 Entertainment books!  
The books are valid right now, thru November 1, 2003.

The Entertainment book is available for 150 cities throughout North America. The coupon books are packed with Buy-One-Get-One-Free and 50% off discounts on restaurants, theatres, video rentals, dry cleaners, sporting events, and more. The savings also continue nationwide with great offers like 50% off hotels and discounts on airlines and car rental.

The price of the book varies by city with a price range between \$20.-\$45. PRISMS will earn 20% profit of each book purchased. When purchasing two or more books, shipping and handling is free. The easiest way to preview and purchase your own local book is to visit [www.entertainment.com](http://www.entertainment.com) and enter the PRISMS Group Account Number (**742332**) during the checkout process.

If every PRISMS member could sell or purchase four books, we could earn profits of over \$8,000! Remember you will get great savings with the coupon book, as well as donating 20% of your purchase to the future of our SMS children.

For more information regarding this fundraiser, please see PRISMS website at: <http://www.smithmagenis.org> or email PRISMS at [info@smithmagenis.org](mailto:info@smithmagenis.org)

Thanks to Michele Zdanowski, SMS Parent and PRISMS Fundraising Volunteer for organizing this great fundraiser. Michele can be reached at: [jzdanowski@comcast.net](mailto:jzdanowski@comcast.net)

## 3<sup>rd</sup> International Smith – Magenis Syndrome Conference Conquering the Peaks and Valleys Together



Over 300 parents, professionals, children, grandparents and significant others interested in SMS gathered in the foothills of the Rockies for three and a half days of learning about the "SMS puzzle" and connecting with a rich network of understanding hearts.

We asked participants for their thoughts.

Shirley Dechaine, mother of Paul, age 34, from Oregon wrote:

"This was an encouraging, positive four days for both parents and children—including siblings. Our keynote speaker was Judith Allanson, an outstanding geneticist who has been responsible for diagnosing many of our SMS kids.

A wide variety of topics were discussed—all of which affect our SMS families daily. One of the most predominant topics revolved around sleep disorders. We were given a research update on the 'science of sleep'.

*Abnormal circadian rhythm in SMS (Smith, DeLeersnyder, Potocki, & Duncan) was explained in detail. PRISMS website will have power point presentations of this session available soon. Also, the scientists from France, NIH in D.C., Baylor College of Medicine in Texas and Oregon Heath Sciences University have informed PRISMS that treatment trials and new protocols to study this unusual sleep disturbance are underway or expected to begin soon at each of their centers.*

"We were introduced (by Drs. Lupski and Walz) to the fascinating SMS mice which reside and assist researchers at Baylor College of Medicine.

One of the most enjoyable aspects of the conference for us was the ability to talk informally with Ann Smith and Ellen Magenis, without whose dedication and commitment to SMS research for these last 20 years we would all still be floundering in the maze. We also thoroughly enjoyed having plenty of time to talk with other parents....It is very encouraging to know that we share common problems. In one of the whole group moderated sessions, "Climbing Out of the Valleys", we helped each other solve actual problems that families deal with,...through the benefit of overhead, we shared the ways in which we in the small group had dealt with those problems."

Jennifer Comford, mother of Caroline, age 3, from Tennessee wrote:

"I feel one of the most beneficial parts of the conference was the parents sharing. We heard how many (parents) are going through the exact same things as we are ...parents who have come through the hard times say that they are seeing the light at the end of the tunnel...encouraging us to stick with it. I found the class on Behavior Management extremely beneficial; hearing that there is no cookie cutter behavior management plan guaranteed to work for every child. Mary (Beall) was very good at encouraging us to do what worked for us ... these kids are special and unique; they will not conform to any mold willingly (unless it is their idea of course). I found validation in this presentation...I shouldn't feel horrible when people who don't live in our SMS world are inconsiderate. They just don't get it."

(Continued on page 5)

Robert Goswell, stepfather of Ryan, age 10, from Australia wrote:

"The Kids Corral was a room set up with toys, books and videos, etc, where the parents could leave their SMS children while they attended the conference.

The 2 ½ hours that I was in that room were very difficult....I personally think that being in that room was the best counseling I could ever get. When the parents dropped their children off and began to leave, the children were getting themselves worked up, distressed, and injuring themselves. When the parents left the room, the kids settled down and had fun. I saw first hand what has confused me for so long, and it still does. These kids can be lovely children, happy and have fun, but not with mum and dad. When their parents came back to pick them up, they acted out again."

As Robert and his wife Kim attended many of the seminars, Robert took notes. These are just a few:

"When talking to your SMS child use simple words, short sentences, one command at a time, visual clues and signs are important.

Do not say, go feed the dog. Say, the dog is hungry, what are we going to do?

Do not fight them when they are locked in bad behavior--walk away, give no attention; make them fix it or pick up the next day.

Assistance for families was stressed.

Parents make the best advocates."

Laurie Bellet, PRISMS Resource Coordinator, wrote:

"The Make and Take sessions, particularly the first one, became areas of family pleasure. The Resource Marketplace became an area for immediate dilemma solving...everything from a recommended toy to strategies for finding caregivers. Comments about the catalog selection ranged from, 'Wow, I didn't know this existed,' to 'Wow, I have been meaning to get this for years.'

Another high point of the conference was the family barbeque—authentic Colorado!! Besides the mouth-watering beef dripping with bbq sauce, were the kid favorites: clowns, a DJ who played 'CELEBRATION' and games with prizes for the many, many winners. Adults were not left out—a Silent Auction (thanks to Janey Peterson) provided the perfect venue for healthy competition and great fun with a profit of \$2,000+ for PRISMS."

A Very special thanks to our chairperson, Marsha Bach, and our on-site chairperson, Eric Hoffman—assisted by his wife Kim. We also recognize the time and effort of many others who are best described as kind, generous, and very ambitious...this group of conference contributors defines our PARENTS, our PROFESSIONALS, all of our SPEAKERS and all of our significant others.

Because of YOU, our conference was a huge success!! Comments and evaluations overwhelmingly rated workshops as "excellent." The conference (overall) was described by many as "exceeded expectations."

We appreciate the contribution of our major sponsors: PRISMS, NIH-SMS Research Unit, Pfizer and all others who gave generously.

*Please check our website [www.smithmagenis.org](http://www.smithmagenis.org) for workshop summaries.*

## Go Ahead, Give in to Surrendering: A Parent's SMS 'Survival Kit'

by Tony Daranyi, father to Ali, age 14



Ali Daranyi (right) and friend, Ariel Belcher, enjoy fun and friendship at the SMS Barbeque held July 5, 2002 as part of the activities during the 3rd International SMS Conference.

When my wife and I were asked to be parent presenters at the recent SMS International Conference in Denver, we scratched our heads and shuddered. Although the title for the conference was “Conquering the Peaks and Valleys Together,” we had trouble with the word “conquering.” We felt that we never truly conquer the trials and tribulations that this mysterious syndrome has presented us. Conquering implies control, something we don’t feel we’ve gained. By admitting this, we didn’t want to frighten any parents who have newly diagnosed children or adult children with the syndrome by presenting too dire a picture. So we struggled to come up with a better, more appropriate word that described our particular journey. After discussing the subject amongst ourselves and with our friends and family, we settled on the word “surrender” as a good substitute that aptly describes our journey. Ironically, surrender is the antithesis of conquer. Through surrender, we give of ourselves to the syndrome and to our daughter, Ali; we gain acceptance. We then made a list of all the feelings, emotions, activities we surrender to, including: new and strict schedules, a seriously curtailed social life, loneliness, renewed compassion, much confusion, helplessness, learning about unconditional love, living in the moment, staying calm, and the SMS diagnosis itself. In the process of this preparation for the conference, we then listed our Survival Kit. These were the items, not in any particular order, that have worked for us as we surrender to the syndrome and to what we hope is a healthy upbringing of our child. We don’t want to preach, nor pontificate; hopefully sharing these items becomes useful for others. Some survival techniques have been added and/or modified since our presentation at the conference. I think we can all agree that raising a child with SMS is by far the most challenging life’s activity we have ever been involved with.

- We have had to completely discard any and all expectations — whether for the next moment, the next hour, the next day, the next week or the next month. Our day-to-day schedule is more determined by the quirky and unexpected needs of our daughter. At the same time, she sometimes blows us away by what she is capable of doing and performing, as opposed to what we expect of her.
- Ali has demonstrated extreme self-abusive behaviors since she was about two years old. (As an aside, and as a word of encouragement to other parents with a self-abusive child, as she’s aged, Ali’s episodes have dramatically decreased in frequency but not necessarily in intensity.) Along our journey we were turned on to the Skinner Behavior Model and we have found this to be the most effective way of coping. We completely, 100 percent disengage and ignore the harmful self-abusive behavior once it has escalated and redirecting (the initial strategy) has failed. One hundred percent disengagement means just that: no eye contact, no changes in body language or facial expressions, no verbal communication, etc. At the same time, we also have a strategy in place — a reward, if you will — to get Ali to do what we want of her. We continually reward the positive behaviors.
- We are the primary advocates for our child, whether dealing with the medical establishment, our families, social services or the educational system. We have inherited the role of a case worker.
- We have accepted that we cannot parent Ali alone. It takes a strong support network, including help from friends, the State, a live-in helper, Big Brother/Big Sister programs, a full-time aide at school, family, counseling.
- We strive to remain positive by looking at the glass as half-full, not half-empty. Overall, Ali has been a tremendous

*(Continued on page 7)*

blessing in our lives, a gift. She has been our teacher, teaching us about aspects of life we never imagined. There is a reason — tangible or not — for why we have been gifted with these children. It is up to us to discover just how this gift has manifested itself.

- We have accepted the situation that confronts us. This does not mean we don't repeatedly go through the grief cycle; we do. But we don't dwell in any one place, we don't dwell in denial, nor in paralysis, nor in grief, nor in devastation. Take charge of the situation and be productive. No matter how dire matters may seem at any one time, there is/will be light at the end of the tunnel.
- Always have a contingency plan in place (a "backdoor," if you will) in the event the worst-case scenario (which for us meant a full-blown, self-abusive episode — we call them "fits") is experienced.
- Along the same lines, always create a safe place, or have one available (e.g. at home, while traveling, at school, shopping, etc.). We have "Ali-proofed" her room (turned locks inside out, taken out all lights, boarded up windows, put in wainscot along the walls, anchored bookshelves and dressers to the wall). When she was little, we created a 4'X4'X4' padded box to place Ali in as a safe place for her fits. We also use the automobile as a sacrificial safe haven when traveling. For safety's sake, we have to be able to separate ourselves, and others, from Ali when she is tantruming.
- We strive to take care of ourselves. If we're not healthy, we cannot expect to have a healthy household. We schedule a night out one day a week, we try to take one day off a week, usually on the weekends, we exercise or pursue an avocation to try to achieve balance in our lives and diffuse the stresses that raising a child with this syndrome presents.
- As ironic as it sounds, keep a sense of humor and laugh with your child. These children have a unique, if not amazing, sense of humor. They want nothing more than for us to laugh, out loud, with them.
- Be compassionate — dig deeper than you ever thought possible, to walk in your child's shoes, to be your child, to be one with your child. At those moments when compassion is required of you, let go of everything else in your life and really connect with your child.
- Initiate a program of tough love. Instead of paying attention and thereby escalating negative behaviors — by hugging, consoling, holding, etc. — tough love in this case means ignoring.
- Stick to a routine and a schedule. Our daughter functions best when we adhere to a strict schedule on a day-to-day basis.
- Since Ali is not wholly proficient at expressing her needs, we have had to anticipate her needs of hunger and sleep for her, as a means of heading off at the pass her self-abusive behaviors.
- Finally, we try to celebrate our child, although we definitely don't do this often enough. The successful small steps in her life — learning how to dress herself, bathe, etc. — are actually large stepping stones to what we hope will be a life of self-actualization and completeness. For this, we must be celebratory.
- Most importantly, we have tried to tap into the incredible love that Ali pours out. We call her our Love Bug. She personifies unconditional love in the purest sense. That is her blessing. That can become our blessing, too, if we're ready to surrender to it.

Good luck!

*Tony and Barclay Daranyi have a 14-year old daughter, Ali, who was diagnosed at birth with Smith-Magenis Syndrome. They also have a 12-year old daughter, Tasha, who is abnormally normal. Ali, now a ninth-grader, had been attending a nearby public school, but has recently begun attending a residential program in Pennsylvania. The family lives in the San Juan Mountains of Southwestern Colorado, near Norwood.*

# THANKS!

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*Beverly Frey*

*Brenda Finucane, M.S.*

*Beth Kurtz*

*Tony Daranyi*

*Ann C.M. Smith, M.A., D.Sc.(hon)*

## 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 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. Here's how, contact:

Mary Beall  
414 Birch Lane  
Richardson, Texas  
Phone: 972-690-1016  
[mary.beall@attbi.com](mailto:mary.beall@attbi.com)



### Important note:

PRISMS parent to parent program will only give out information on parents who have agreed to be contacted.

# PRISMS

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