prisms

spectrum

PARENTS AND RESEARCHERS INTERESTED IN SMITH-MAGENIS SYNDROME

INSIDE THIS ISSUE

PRESIDENT'S MESSAGE

SOME REFLECTIONS ABOUT THE CONFERENCE AND A LOOK FORWARD

If you were with us in St. Louis, I hope you agree that the 9th International Conference on Smith-Magenis Syndrome was a great success. While the accounting is still to be completed, in my view the learning, sharing, support and friendship that was part of our gathering is what we are about. I came home enriched and feeling even more deeply connected to our community. Hopefully, you did too.

THE CONFERENCE BRINGS US TOGETHER IN MANY WAYS

At the most basic level, our conference is about creating a place for sharing information and learning. Whether you are a newly diagnosed family or a "veteran" with an SMS adult, you will learn and be enabled with information you can use.

Sometimes what you hear is a "refresher"—a topic or presentation you may have heard before. I find these sessions are valuable for deepening my understanding and my ability to explain the SMS "experience" to others.

I enjoy seeing friends, meeting new families and interacting with the presenters who are part of our community. Every conference, we welcome newly diagnosed families. Connecting with them and supporting them, whether they have infants or teenagers, is what our conference is about. Answering questions, sharing experiences, and perhaps helping by showing how to find humor and joy in the midst of challenging moments.

I was humbled by my casual accounting of how many people are involved in PRISMS, and also by the number of people who helped with the conference. Volunteers, speakers, presenters, exhibitors, child-care helpers, staff, consultants, donors and supporters … their efforts are aligned to make the conference successful.

And our board members, regional representatives, committee volunteers, members of the professional advisory board, international affiliates, our staff, and our members all play a role throughout the year, making PRISMS a vital and effective operation. Taking stock of everyone, watching people in action, and hearing what people had to share, I am grateful to everyone for their contribution and commitment to PRISMS.

LOOKING FORWARD

The two-year break between conferences provides a chance to strengthen the organization, focus on and complete new initiatives, and do the work needed to plan and deliver the next conference.

Strengthening the organization means clarifying goals, recognizing and stabilizing areas of need, engaging volunteers, attracting and retaining members, and fundraising.

Also, we are effective with our planning, and new initiatives are generally in the queue. We have recently added a genetic counselor who will be available for families (see more on page 13). We are working on a patient registry. And we are improving our network of regional representatives, just to name a few.

Lastly, planning for 2018 begins with a thorough review of the 2016 conference. A new committee will lead the process, and soon we'll be sharing the location.

You hear me often say, I am extremely proud to be part of PRISMS. I deeply appreciate the commitment and efforts of the members of our community who help us, and I am glad to build an organization that serves our families.

Sincerely yours,

John Mayer
President, Board of Directors
jmayer@prisms.org

PRISMS 9TH INTERNATIONAL BUILDING BRIDGES OF HOPE CONFERENCE A SUCCESS!

By Emily Fields

On July 28-30, PRISMS hosted and sponsored its 9th International Building Bridges of Hope Conference—the largest conference specifically focused on Smith-Magenis Syndrome. More than 300 people, representing 35 states and eight countries, attended. International attendees came from Canada, Mexico, France, the UK, Norway, Australia and Iceland.

With more than 40 sessions on topics ranging from Sibling Support and Behavior Strategies to Research Updates from leading researchers in SMS, Medical Management, Genetics 101 and more, there was no shortage of information for attendees to access.

It was an insightful, encouraging and meaningful conference for me. I heard our attendees found the same comfort in meeting with SMS families, professionals and a community network of support. Among the comments:

> *I received a grant and I can't thank you enough for this opportunity. As a teacher, we always want to do the best for our students, and you have just given me a much better understanding and a chance to come up with some strategies to start and ideas to pull a team together to make a plan for our student. Thanks so much.*
>
> *Great conference. Exceeded expectations*
>
> *Keep up the magnificent work you guys do with this. We travelled from Australia to attend and will more than likely be back for the next one as it was such a well-run, inviting, friendly, inspirational, emotional, supportive conference. Well done to all involved.*

As Pat Furlong, our keynote speaker, expressed, an empowered community is a strong one … capable of creating real change.

PRISMS is committed to continuing to empower the community through education, awareness and research. As an organization, PRISMS has been working for the SMS community for 23 years. But it needs each of you to make sure we can continue to create meaningful resources, such as the Guidebook for Schools or the Guidebook on Adult Residential Living, and host powerful programs such as the International Building Bridges of Hope Conference and the SMS Research Symposium.

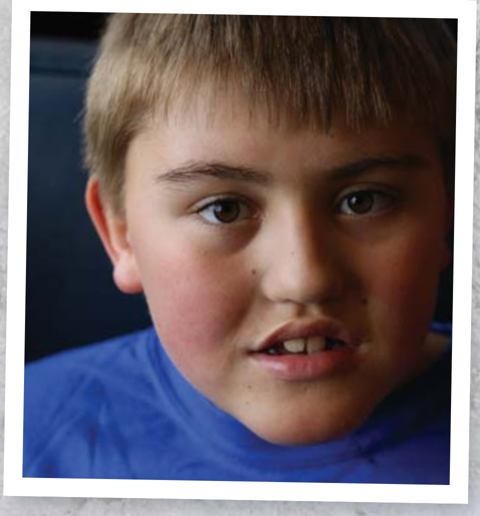
As you read through this edition of the Spectrum, I ask each of you to consider: How can I be a part of building the future for the SMS community? With opportunities to give of your time, talent and resources, I invite you to contact me at info@prisms.org and let's start empowering the community together.

Thank you to all of those who were able to join us for the 2016 conference! I look forward to seeing you again and meeting new faces at the 2018 conference.

Together we are stronger.

Emily Fields
Executive Director

CELEBRATE COMMUNITY!



DIGNITY *of* RISK

By Zach Kon

ZACH KON, 24, WAS DIAGNOSED WITH SMITH-MAGENIS SYNDROME AT AGE 15. HE LIVES NEAR BOSTON. HE DELIVERED THIS TALK DURING THE 9TH INTERNATIONAL BUILDING BRIDGES OF HOPE PRISMS CONFERENCE IN ST. LOUIS JULY 28-30.

Hi, my name is Zach Kon. I am here today to talk about my Dignity of Risk story. I will talk about how I took a chance, or a risk, and applied for a job. I will tell you how it worked out.

But first I want to define the Dignity of Risk. I looked it up on Google, and this is what I found. "The dignity of risk is the right to take risks when engaging in life experiences, and the right to fail in taking these. This term was first coined around the issue of care for people with intellectual disabilities in the 1970s."

But first a little of my history. I've been with a group called Massachusetts Advocates Standing Strong for many years. One of the many things I like to do with Mass Advocates is to go to the state house almost every month and talk to my legislators and representatives about issues that are going on with the special needs community.

Being part of Mass Advocates has helped me gain more control in my life by giving me empowerment and confidence to be part of the world. Knowing that there is a field of successful people with disabilities around me gives me encouragement. Being part of Mass Advocates taught me how to stand up better for myself.

My dignity of risk story is that I want to work. I want to make money. I want the satisfaction of knowing I am doing a good job. I want to be recognized by my boss and coworkers. I want to get out of my day program more and be productive in the community.

I find it important to be around people because I am a social person and I like to talk to other people, and I like to move a lot. So it was important to me to get a job where I could do those things.

It was hard for me to think about applying for a job because I thought I would never get an offer. I was concerned I wouldn't know how to do the job without the proper training. I had mixed feelings about it. I was both excited and nervous about getting a job.

A friend of mine said that you can't experience the world if you don't get out and do it yourself. You can't have your parents do everything for you in the long haul or you will never truly learn how to experience the world for yourself.

I am different. It is hard to get a job in the community for anyone, especially for those with disabilities.

I saw this job at a local food store, Wegmans, and I thought this would be a great opportunity because I know the people there are really nice because I have shopped there.

So now I will tell you the steps I took to apply for a job. To start, when I lived at my residential schools I worked at small jobs. I also practiced my interviewing skills and my cover letter skills, and that's where I built my first resume.

My school had a coffee shop called Somethin' Brewing, where I trained and learned how to make coffee and tea and prepare bagels. I took orders from people, made their orders, and delivered them to them.

For example, I knew how a customer liked her tea, and she would rely on me to make it for her whenever I was working. After that residential placement I started at my day program where I worked at some volunteer opportunities like Meals on Wheels and the Salvation Army.

I also participated in job camp, which helped me prepare for real life work. Job camp was a year-long class that taught me interviewing skills by practicing in front of a camera and watching the video with the whole class.

It also taught me resume building skills and how to dress for an interview. The teacher showed me how to take initiative in a job interview. In the practice interviews, I was very nervous and shaken up.

A little over a year went by and this is what took place. I applied for and got a job at Wegmans. The way I got this job is, with the help of my job coach I looked on-line at the Wegmans site. On the site we saw there were positions open in their Helping Hands Department. I applied for the job.

They did not take me the first time because the hours that worked for me did not match what they were looking for. So I tweaked the hours and then they called me in for my first interview.

I was nervous at the practice interview with my job coach. So I practiced thinking good thoughts to calm myself down and do well at the Wegmans interview. By being calm, I knew I would do a better job going forward.

The interview went really well. They called me back for a second interview that went amazingly well. My job coach was with me for both interviews. They called back within a week and told me I got the job.

I did not think that I would ever have an opportunity to work in the community. I worked hard and proved myself completely wrong. I went through all the right steps from building a résumé to applying for the job and having an interview. Taking that risk actually led me to have the world's most amazing opportunity.

I thought back to my first practice interview, and that just proved to me that I can do anything. Taking a risk shows how far I or anyone can go. From residential schools to my day program, and then to competitive employment makes me happy.

I worked at Wegmans for about one month and it was a nice place to be, with nice co-workers and bosses, but it wasn't suitable for me. Working for a big company is not the right fit for me. So I stopped working there.

Since then, my job coach has been helping me look for other jobs. He found one at a local gas station where I'll be stocking shelves and cleaning up the trash. I hope to start in December.

The Dignity of Risk is trying something new and seeing how it works out. I took a risk by working at a large company. That did not work out for me. I'm glad I took the chance because I learned what works best for me.

I want to thank the members of the PRISMS board for letting me give my speech and opening the conference. I want to thank you all for taking the time to listen. And I want to thank Ann Smith and Ellen Magenis. If it wasn't for them we would never know about the syndrome.

CELEBRATE COMMUNITY!

FROM THE COMMUNITY…

The 2016 International Building Bridges of Hope Conference was the first SMS conference for many families.

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Building Bridges of Hope
9TH INTERNATIONAL CONFERENCE • ST. LOUIS 2016

Allison and Alex Stephanouk

Where from:
Columbus, GA

SMS child: Natalie, five; diagnosed at two weeks old.

One thing we learned at the conference: We really appreciated the session on special education since our daughter is starting kindergarten this year. We learned about educational advocates and how they can help families navigate through the school system to obtain the services that are best for the child.

The best thing about the conference: Meeting other families affected by SMS.

John Roseborough

Where from:
Wichita, KS

SMS child: Elsie, four; diagnosed in January 2016.

One thing I learned at the conference: That the PRISMS SMS community is very approachable, loving, and supportive.

The best thing about the conference: Getting to know other folks and learning.

Geoff and Marika Thomsett

Where from:
Australia

SMS child: Sarah, eight; diagnosed at one week old.

One thing we learned at the conference: Our emotional state, recognizing and accepting it, is a major contributor to us as parents being able to provide the best support, guidance and encouragement to help Sarah thrive. Be the best parents we can be, change what we can, make a difference, and let go what we have no control over.

The best thing about the conference: Being so far from home, so far from friends and family, yet finding ourselves at home among (PRISMS) friends and family.

The Gateway We're All Traveling Through

By Tony Daranyi

When our daughter, Ali, was diagnosed at birth in 1988 with Smith-Magenis Syndrome, our lives took a sudden and unexpected turn. Without a doubt, we were unprepared for the journey ahead. Our faint road map was a stack of nine case studies examining the only other known cases of SMS. Except that, when Ali was diagnosed, there wasn't even a name for the syndrome. It was simply called "17 P-minus". The case studies were dire, painting a bleak picture of what lay ahead.

"The good news is she doesn't have Down syndrome," Ali's pediatrician said, when he informed us over the phone that she had a chromosomal abnormality. "The bad news is she has something much worse." The case studies reflected patients who had mostly been institutionalized their entire lives. These patients were mostly non-verbal, self-abusive, physically handicapped, and socially maladjusted.

We were devastated. And we were scared. But we committed ourselves then and there to accept our fate and to take care of Ali to the best of our abilities. We would try to remain optimistic and live a day at a time.

Fast forward to the recent International PRISMS Conference in St. Louis. My wife, Barclay, (who presented on residential options at the conference), Ali and myself all were able to attend. It was a powerful three days that far exceeded all expectations.

Before registration on Thursday, Ali and I took a field trip to the Gateway Arch, the famous St. Louis monument. It was there that I was struck by a powerful metaphor on the journey we all take as parents, researchers, physicians and teachers of children and adults with SMS.

The arch symbolizes the westward expansion of the U.S. St. Louis played a key role for settlers seeking a better life, and for some fame and fortune. The city was at a crossroad between the east and the west. Once past St. Louis, the settlers were headed into the unknown. They had few trails to follow, and those they did follow were usually obscured and not well traveled. There were no maps, few markers, and little direction. Travel was often dangerous. Life on the road was difficult, to say the least.

The parallels between what those settlers experienced and what those of us involved with SMS children and adults are interesting to say the least.

Parents are faced with daily challenges: behavior management, sleep disorders, obesity, hearing aids and eyeglasses, general health issues, residential options, school placements. We're forced to become experts on so many fronts.

Like the settlers crossing through the proverbial arch, we are all—parents, researchers, geneticists, teachers, caregivers, doctors, fundraisers, and PRISMS directors—mostly traveling into uncharted waters. The path is arduous. It's challenging. It's difficult. There is little guidance on what the "best path" might be.

How many of us have been to mental health "experts" who really don't know much about self-abusive behavior? Who out there understands circadian rhythms, and those of children whose melatonin production is turned upside down? What are the implications of having pain tolerance levels that are off the charts? Who knows? On the medication front, it's mostly experimentation, a guessing game at trying to figure out what prescription cocktail might work for any one of a number of issues we're trying to address with our children. What educator knows of the best classroom environment for our children? What disability service center can explain to us what the best residential options might be for our adult children?

Apart from PRISMS commitment to research and support services, and its fabulous resource guides ("On the Road to Success with SMS" for educators and "Exploring Adult Residential Housing Options" for parents) there aren't many maps for us to follow to keep us on course. We're essentially on our own.

We have each other, which, as the conference so powerfully demonstrated, is crucial to our "making it." We all travel in the same wagon train together, down a road never trod before. We're adventuring through this life together.

Like the Western settlers, this travel requires teamwork to be successful. Most important, it requires unconditional LOVE, some tolerance and lots of patience. Those are the qualities that will get us through the challenges and allow us safe passage crossing under the proverbial gateway. Isn't that what these SMS children and adults are ultimately teaching us? They are gifts we all need to cherish.

CELEBRATE COMMUNITY!

PRISMS 2016 CONFERENCE BY THE NUMBERS . . .

316

registered attendees (representing 35 states and eight countries)

10%

of attendees diagnosed with SMS

40 PLUS

conference sessions, including Genetics 101, Residential Settings for Adults with SMS, SMS and Sexuality, Medical Management, research updates and behavioral therapy

21

attendees in childcare services

11

attendees in PRISMS Den services

50%

of regional representatives in attendance

91

attendees for Night at the 360, PRISMS inaugural fundraising event

400 PLUS RAFFLE TICKETS SOLD

16

teachers and support staff funded by PRISMS to attend the Teacher/Support Staff Workshop

33

PRISMS members who attended courtesy of the Family Support Scholarship program

$35,000 PLUS

amount contributed by PRISMS to support conference attendees with registration, lodging and travel costs

25

professionals supported by PRISMS to present and engage in the conference

$90,000

approximate donations received by PRISMS through sponsorships, contributions, raffles and the Night at the 360

Building
Bridges
of Hope

Welcome Dianne Samad, MS, CGC to the PRISMS Team!

PRISMS is pleased to welcome Dianne Samad, MS, CGC, as the newest member of its team and support for the Smith-Magenis Syndrome community.

Ms. Samad is a genetic counselor at the Baylor College of Medicine. As part of PRISMS commitment to research and education, it has partially funded this genetic counselor position.

Ms. Samad will work with Dr. Sarah Elsea on establishing and maintaining a registry for individuals with SMS, the overall aim being to improve patient care through a better understanding of the nature of the disorder. In the lab, Ms. Samad will collaborate with Dr. Elsea to coordinate and recruit participants for ongoing research studies and clinical trials for SMS. She also will assist with maintaining the PRISMS website and will be a person of contact for questions regarding the genetics and inheritance of SMS.

Ms. Samad earned her master's degree in genetic counseling from the Oklahoma University Health Sciences Center (OUHSC) and her undergraduate degree from the American University of Beirut (AUB) in Beirut, Lebanon.

SEVERE PROBLEM BEHAVIOR AND SMITH-MAGENIS SYNDROME

By Mindy Scheithauer, PhD

Problem behavior is a common issue among individuals with SMS, with estimates of 93 percent of individuals engaging in self-injury (Arron et al., 2011). Problem behavior can be incredibly stressful for caregivers of these individuals; not only can it cause physical harm, but it also can interfere with family functioning and development of adaptive skills.

Individuals with SMS may engage in problem behavior for a variety of reasons, but frequently it can be a result of communication deficits. That is, individuals with communication deficits may use problem behavior to communicate wants or needs such as:

- To get attention (e.g., reprimands, comforting/soothing, or a surprised reaction).
- To get access to a preferred item or activity (e.g., food, iPad, toy, going outside).
- To get out of (or escape) something that he/she does not like (e.g., brushing teeth, chores, environments with loud noises, or academics).

Caregivers often provide consequences like these when their child engages in problem behavior in an attempt to get him/her to calm down or to prevent injury. However, over time this teaches the child that he/she can obtain such outcomes through engaging in problem behavior. When this is the case, this pattern of learning is best corrected by using a combination of the following strategies:

- Stop providing the preferred outcome following problem behavior.
- If the problem behavior occurs to get attention, ignore the problem behavior.
- If the problem behavior occurs to get preferred items/activities, avoid giving any items or activities after problem behavior.
- If the problem behavior occurs to escape, follow through with all demands.
- Provide the preferred outcome after appropriate behavior.
- Provide the attention/item/escape after a period of time without problem behavior (e.g., every 10 minutes without problem behavior)
- Provide the attention/item/escape after a specific appropriate behavior, such as requesting attention or complying with a demand.

When using these procedures, it is crucial to start small and go slow. If a child is currently engaging in problem behavior as soon as the iPad is removed, start by removing it for only few seconds. If a child engages in problem behavior as soon as you attempt to brush his/her hair, start by only touching his/her head with the brush. After initial success, expectations can gradually increase.

Sometimes the most challenging aspect of the process described above is identifying exactly why problem behavior occurs. Also, with these treatment strategies problem behavior often gets worse before it gets better.

Given the dangerous nature of problem behavior, it is important that caregivers of individuals with SMS identify a professional in the field of behavior analysis to assist with assessment and treatment. This individual may be a Board Certified Behavior Analyst (http://info.bacb.com/o.php?page=100155) or other professional (psychologist or special education provider) with experience treating problem behavior using behavioral strategies.

Mindy Scheithauer is an assistant professor in the Division of Autism and Related Disabilities in the Department of Pediatrics at Emory University's School of Medicine. This is a summary of a talk she presented at the PRISMS conference in St. Louis, July 28-30.

FIND US ON SOCIAL MEDIA

Host a fundraiser or awareness event in your area

Contact info@prisms.org for support and ideas to make your event a success

Join the PRISMS community by becoming a member for 2016

Learn more about membership by visiting our website at www.prisms.org

Make a donation

Support PRISMS mission to increase awareness, education, and research for the SMS community by visiting our website and making a donation today at www.prisms.org

Share your story with us

We want to hear from you. Reach out to PRISMS Executive Director at efields@prisms.org

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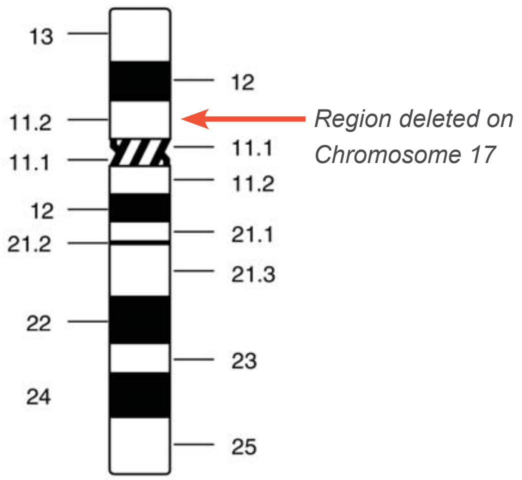
What is Smith-Magenis Syndrome?

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 1980s by Ann C.M. Smith, M.A., a genetic counselor, and Ellen Magenis, M.D., a physician and cytogeneticist. Although the exact incidence is not known, it is estimated that SMS occurs in 1 out of 15,000 births. SMS is under-diagnosed, but as awareness of it increases, the number of people identified grows every year.



VOLUNTEER OPPORTUNITIES

PRISMS is always looking for helping hands from the community to serve as members of one of our various committees, assist in a program, or coordinate an event. Interested in sharing your time and talents with the SMS community? Contact us at info@prisms.org

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