July-August 2004
Volume 29, Number 4

The Gathered View

National Newsletter of the Prader-Willi Syndrome Association (USA)

In this issue

Have Faith and Set Your Goals High

By Robin MacGillivray

Robin MacGillivray, center, receives her diploma from the University of St. Thomas

I recently graduated with a double major in Elementary Education and Sociology from the University of St. Thomas in St. Paul, Minnesota. There were approximately 1,300 students who shared this honor with me, so why was this day so much more special for me? Because I struggle with that dreaded PWS — which, unfortunately, we all have experience with personally or through someone we care about.

I was asked to write an article in *The Gathered View* not only about this accomplishment but also my life story. I am flattered by this request and I strongly believe an article which mentions things I believe to have alleviated particular challenges attributed to PWS could be immensely beneficial.

One of these things was — and still is — support. I am blessed with an extremely strong support network. Both my nuclear and extended family love, encourage and support me. In addition, there never fails to be a plentiful supply of friends who are there for me every step of the way. Thus, everywhere I turn there is my enthusiastic cheering crowd, so I'm never left hang'n out on a limb struggling for dear life.

Another was literature. Fortunately, literature has always been a strong part of my family's life. I get pleasure from remembering the mammoth-sized collection of literature — the Mrs. Piggle Wiggle series, Dr. Seuss, Shakespeare, or *To Kill A Mockingbird* — available in our household for my sister and me to get exposed to. We always made a point for read-aloud sessions. These sessions started out with me — at a very early age — being read to. But as the years went on my mother turned the tables on me… now *I* had to read to *her*. With each read-aloud session I began to acquire a greater repertoire of speech which quickly improved my articulation and friendships.

Being goal-driven is also among the things that were — and still are — helpful. Establishing important goals for myself was beneficial in the long run because it helped me channel my determination, passion and energy into working toward something that in my heart I yearned for. The more significant the goal, the more contentment I experienced. In other words, my self esteem grew with each major goal I set.

Fourth, I always possessed faith. Faith in our Lord, in my cheering crowd and in myself. The stronger my faith, the stronger my entire self — including my spirit —

Robin continued on page 12

Prader-Willi Syndrome Association (USA)

5700 Midnight Pass Road, Suite 6 . Sarasota, Florida 34242
1-800-926-4797 9 a.m. to 7 p.m. Eastern Time
Local telephone: 941-312-0400 . Fax 941-312-0142
e-mail: national@pwsausa.org . web site: www.pwsausa.org

The Gathered View (ISSN 1077-9965) is published bimonthly by the Prader-Willi Syndrome Association (USA) as a benefit of membership. Annual U.S. membership dues are: 30 Individual; $35 Family; $40 Agencies/Professionals. Membership dues outside the United States are $40 Individual, $45 Family and $50 Agencies/Professionals (US Funds). We never deny parents membership for any reason.

Adults with PWS Advisory Board

Diane Arnoudse, Michigan
Shawn Cooper, Georgia
Brooke Fuller, Michigan
Kate Kane, Maryland
Andy Maurer, South Carolina
Margaret Miller, Pennsylvania
Mike Nacht, Ohio
Abbott Philson, Maine

Our Mission: Through the teamwork of families and professionals, PWSA (USA) will improve and enhance the lives of everyone impacted with Prader-Willi syndrome (PWS) and related conditions.

PWSA (USA) Board of Directors

Chair - Carol Hearn, Plymouth, MN
Janice Agarwal, Zionsville, IN
Daniel Armento, Huntington Valley, PA
Daniel Driscoll, M.D., Gainesville, FL
Lisa Graziano, Redondo Beach, CA
John Heybach, Chicago, IL
Stephen Leightman, Cherry Hill, NJ
Carolyn Loker, Plainwell, MI
Robert Lutz, Bryn Mawr, PA
Barbara McManus, Buffalo, NY
Mark Ryan, Newhall, CA
Mary K. Ziccardi, Cleveland, OH

Scientific Advisory Board

Chair- Merlin G. Butler, M.D., Ph.D., Children's Mercy Hospital, Kansas City, MO
Chair Emeritus - Vanja Holm, M.D., University of Washington, Seattle, WA
Suzanne B. Cassidy, M.D., U.C.I. Medical Center, Orange, CA
Joe Donnelly, Ed.D., University of Kansas, Lawrence, KS
Elisabeth M. Dykens, Ph.D., University of California, Los Angeles, CA
David Ledbetter, Ph.D., Emory University School of Medicine, Atlanta, GA
Phillip D.K. Lee, M.D., UCLA School of Medicine, Los Angeles, CA
Harriette Mogul, M.D., New York Medical Center, New York, NY
Sue Myers, M.D., St. Louis University, St., Louis, MO
Robert Nicholls, D. Phil., University of Pennsylvania, Philadelphia, PA
Ann Scheimann, M.D., Johns Hopkins University, Baltimore, MD
Barbara Y. Whitman, Ph.D., St. Louis University, St. Louis, MO

Clinical Advisory Board

Chair - Daniel J. Driscoll, Ph.D,.M.D., Univ. of Florida Health Science Center, Gainesville, FL
Moris Angulo, M.D., Winthrop University Hospital, Mineola, NY
Ivy Boyle, M.D., Bellefaire JCB, Cleveland, OH
James Boyle, M.D., St. Vincent's Charity Hospital, Cleveland, OH
Aaron Carrel, M.D., University of Wisconsin Hospital, Madison, WI
Elisabeth M. Dykens, Ph.D., University of California, Los Angeles, CA
Linda Gourash, M.D., The Children's Institute, Pittsburgh, PA
Louise Greenswag., R.N., Ph.D., Iowa Child Health Speciality Clinics, University of Iowa, IA
Bryan Hainline, M.D., Ph.D., Riley Children's Hospital, Indiana University School of Medicine, IN
Jeanne Hanchett, M.D., The Children's Institute, Pittsburgh, PA
Karen Levine, Ph.D., Spaulding Rehabilitation Hospital, Boston, MA
Jim Loker, M.D., Bronson Methodist Hospital, Kalamazoo, MI
Helen McCune, M.S., R.D., Shands Hospital Food & Nutrition Service, Gainesville, FL
Barbara Y. Whitman, Ph.D., St. Louis University, MO
Liaison Members:
Ken Smith, Chair, PWSA Board of Directors, The Children's Institute, Pittsburgh, PA
Suzanne Cassidy, M.D., Scientific Advisory Board, U.C.I. Medical Center, Orange, CA
Janalee Heinemann, M.S.W., Executive Director, PWSA (USA), Sarasota, FL

Check our PWSA (USA) web site
Members Only section for Special Opportunities
Limited to Members: www.pwsausa.org
User Name: members
Password this issue is FREEDOM
If asked for Domain Name it is CIMCO

Lota Mitchell

President's View

On to New Roles Within the PWS Family

It doesn't seem so long ago that I was writing my first President's Message for *The Gathered View* after becoming president in absentia because of the birth of my grandson, Joe. But now my 3-year term is ending, Joe will have his third birthday June 26, and I will pass the gavel and the responsibilities of the presidency to our new president, Carolyn Loker, officially at the Board meeting July 3.

Carolyn, a deeply caring, loving, sensitive person, is a wonderful choice. These past 3 years she has worked very hard as vice president of PWSA(USA) and as a member of the Board of Directors. It was she who started the Parent Mentoring Program, which has benefited so many anxious and grief-stricken parents of babies and young children struggling with a frightening diagnosis — Prader what? — with a strange name they have never heard of. Hours and hours, actually days, of her time go into that, as well as serving on the Executive Committee with many conference calls. With all of that, she had the dedication to PWS to take on being co-chair of the 2004 National Conference in Ohio and doing a wonderful job at the time I write this.

Carolyn Loker

Carolyn Loker, a deeply caring, loving, sensitive person, is a wonderful choice as the new PWSA(USA) president.... Help her to do the best job she can.

So what shall I tell Carolyn about her new job? Well, let me start with the hardest part for me. That was not being able to wave a magic wand and solve the problems of our chapters. They are experiencing many, whether it be chasms between the parents of older and younger children, difficulties finding volunteers willing to do the work of the chapter or just attracting people to meetings.

Chapters are so very important. Just a few of their possible functions include providing instate person-to-person support that distant national and even e-groups cannot, information about state services, education of medical and educational personnel in the state, and fund raising to help members attend the national conferences or other projects the chapter may choose to support. Finding ways to support and strengthen them is so vital.

And the best part of the president's job? The people! There was the opportunity to get acquainted with chapter leaders across the whole United States. If there was a weather disaster in one of our states, I would immediately think about those I knew in that state, wonder if they were affected, and hope they were all right. Listening to people's stories about themselves and their children with PWS was a real gift and privilege. So was working with the PWSA(USA) Board of Directors and the other officers. What a terrific group! I shall truly miss those experiences.

But I'm not leaving completely. I'll continue in my role as associate editor of *The Gathered View*, and Janalee has asked me to be chair of the Publications Committee. So I'm not saying goodbye. I'm definitely hard to get rid of!!

What message would I like to leave? Actually, there are three. First, appreciate what you have now. Don't let your child with PWS be the entire focus of your existence to the exclusion of your marriage, your other children, your extended family, and friends. Be open to and enjoy all the good things that are happening in your life. Don't wake up years later realizing with regret you lost all those years in a sea of sadness. I have learned this lesson the hard way, losing years after Julie was born with PWS and after my son David was killed in an auto accident.

Second, support Carolyn. Being in a leadership position opens you up to all sorts of criticism, much or most of it undeserved. Sometimes the best of intentions and actions get distorted by rumor and misunderstanding. Help her to do the best job she can.

And last — but not least — support PWSA(USA). It's a wonderful organization with wonderful people and wonderful goals for our wonderful (yes, they are) children. It's more than an organization — it's a family. Support it with your time, your talents, and your treasure. Together, we will reach those goals.

Peace.

PWS Awareness

PWSA(USA) Joins Coalition for Children's Health

By Richard Lutz

This spring PWSA (USA) joined the Coalition for Children's Health (CCH) and PWSA's former Board member Jim Kane became a co-chair of the Coalition.

Representatives from various groups formed CCH 4 years ago to aid children (and adults) with disorders such as autism, Down syndrome, muscular dystrophy, juvenile arthritis, Fragile X and similar conditions.

Its goal is to bring greater awareness of these disorders to Congress and Federal agencies. CCH encouraged Congressional interest in enacting comprehensive children's health legislation and helped to bring adoption of the Children's Health Act in 2000. The Act creates and expands federal programs that have the potential to greatly improve the quality of life for many children with chronic diseases and disabilities.

I learned about the CCH from David Busby, a law firm colleague of mine in Washington, D.C. He and his wife Mary Beth have two adult children with Fragile X syndrome, a genetic abnormality in an X chromosome that leads to delayed development and other symptoms. They were among the founders of CCH (Mary Beth was past chairperson) and are leaders of FRAXA Research Foundation, which has been very successful in raising funds for Fragile X research, both privately and through government sources. By their efforts Fragile X was specifically included in the Children's Health Act. That Act comes up for renewal in 2005.

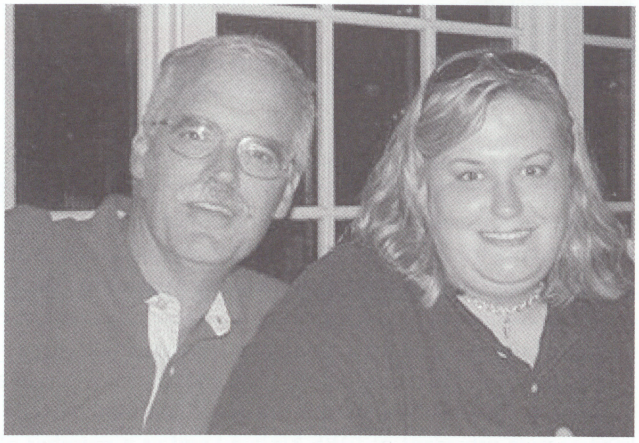
Each year CCH prepares a very substantial booklet for congressional and agency staffers. The current version, the "Children's Health FY 2005 Implementation Blueprint," seeks to implement programs under the Children's Health Act, and includes suggested appropriation language. Jim Kane, Janalee Heinemann and I drafted a four-page section concerning Prader-Willi syndrome which appears in the current Blueprint.

Jim met with agency and congressional staffers when the booklet was distributed to them. This July he will attend another CCH-sponsored meeting to again promote children's health causes to them. While Jim will speak for all CCH members, he will also try to foster greater awareness of PWS. A major objective is to obtain more government funding for research and treatment.

Substantial funding is available for all these causes. The National Institutes of Health (NIH) and all of its sub-agencies have an annual budget of about $30 billion. One agency, the National Institute of Children's Health and Human Development (NICHD) has a budget of more than $1 billion for diseases primarily affecting children.

These agencies and the Center for Disease Control (CDC) make grants primarily to researchers and research programs at universities. They are already providing several million dollars for research which directly or indirectly benefits those with PWS. Many scientists who are working

PWSA(USA)'s Jim Kane with his daughter Kate, who has PWS



with research projects for Prader-Willi, including those on the PWSA(USA) scientific and clinical advisory boards, already receive funding from these and related federal programs. These board members believe that having a higher profile with such agencies through CCH can help to increase available funds and channel the research spending in directions we believe are likely to be most productive.

At present obesity is perhaps the most prominent health issue. It was recently given recognition by the World Health Organization and undoubtedly will receive major governmental research funding. Obesity is also receiving substantial private funding because of the potential for new medications that could bring about weight loss through control of appetite. Work with hormones Grehlin and Peptide YY, which are controllers of the feeling of hunger, is now in progress. There is considerable opportunity for Prader-Willi research to be coordinated with these obesity projects. No organization representing obesity initiatives was part of CCH, and accordingly, Jim Kane drafted a section of the Blueprint discussing childhood obesity.

Jim notes that to be fully effective, we need PWSA members in their home states to contact their congressional offices and bring PWS concerns to their attention. If anyone would like to exchange ideas, or has contacts with people on congressional committees involved with appropriations or health issues, Jim would like to hear from you. Please contact him via the PWSA(USA) National Office, 1-800-926-4797 by email to jgkane@msn.com.

There is no certainty that joining CCH will prove beneficial to PWSA(USA) or to the cause of improving knowledge of Prader-Willi syndrome, but it appears to be a unique opportunity. Our various board members believe the effort is well worth undertaking, and there is reason to believe that this is another channel through which progress can be made for children with PWS.

Richard "Dick" Lutz lives in New York City. He's the father of Board member Robert Lutz, whose daughter Isabel has PWS.

CHAPTERS & AFFILIATES IN GOOD STANDING FOR 2004

We are proud to announce that the following Chapters and Affiliates have achieved Good Standing for 2004 by sending in the necessary documentation (chapter agreement, list of officers and board members, minutes, financial report and $35 annual fee) by deadline: Arizona * California * Colorado * Connecticut * Florida * Georgia * Illinois * MD/VA/DC * Michigan * Minnesota * Missouri * New York * Northwest * PWSA of Ohio * Oklahoma * Pennsylvania * Utah * Wisconsin

Thank you all!

Welcome to new Co-Presidents of the Wisconsin Chapter, Nancy and Steve Burlingame. Owners of their own diagnostic imaging business, the Burlingames are grandparents to Kyle, age 9. When he was born, their daughter Crystal was living with them, so they became very close to Kyle and involved in his care.

Camp opportunity! In October of 2003, **PWSA of Ohio** sponsored a very successful camp for children and adults with Prader-Willi syndrome. This year the organization is having two weekend camps, again at Recreation Unlimited, a totally handicap-adapted facility in central Ohio. The food and activities will be tailored specifically for those with PWS ages 8 through adult, including non-Ohio residents. The dates are June 25 to 27 and October 22 to 24. For information on and questions about the camps, check the PWSA of Ohio newsletter on their website, www.pwsaohio.org or contact Sandy Giusti at 614-876-1732 or by email at juicete@aol.com.

PWSA of Maryland, Virginia and D.C. did something different on May 22 — a walking fund raiser which was done in relay teams with up to 12 members. The day included exciting activities for the whole family, such as carnival games, miniature golf and more.

Thanks for our wonderful donations

An anonymous angel responded to our need for a laptop computer. When Janalee is not talking on the phone, she is now using this brand new, top-of-the-line laptop. Thank you, Angel!

Ray Mathews donated a Sony digital camera to the PWSA(USA) National Office. He dedicated his contribution to Ashley Braun, the daughter of Mary Braun. Mary works in Ray's Century 21 office in Freeport, Illinois and is educating the office staff about PWS.

PWSA(USA) Included in 2004 Combined Federal Campaign

Combined Federal Campaign

PWSA(USA) has been notified of eligibility on the donor list for the 2004 Combined Federal Campaign (CFC). PWSA(USA) will appear in the listing of "National/International Organizations" published in each local CFC brochure.

Those who work for the Federal Government and its agencies must use CFC Identification number 9858 to designate that PWSA(USA) is to receive donations.

If you have questions about CFC donations, please contact the PWSA(USA) national office at 800-926-4797.

Golf Tournament raises more than $12,000 for PWSA(USA)

Josilyn, 21 months, who has PWS, with her grandpa, Ira Levine

Thanks to the Josilyn Faith Foundation golf tournament fund-raiser, $12,518 was donated to PWSA (USA) for research.

The event was hosted by Josilyn's grandparents, Ira and Ronnie Levine. Family and friends came to Palm Beach Gardens, Florida from as far away as California to play golf and show their support. The star of the show was obviously 21-month-old Josilyn, who charmed all with her smile and thank you.

Reserve Friday, April 8, 2005 for the Josilyn Faith Foundation's second annual PGA national golf tournament! Anyone interested in being on their mailing list for notification, please contact the PWSA (USA) office at 800-926-4797.

Would you like to raise funds for PWSA(USA)?
Call Steve Dudrow at 1-800-926-4797
and ask for a free fund-raising packet

BBB Wise Giving Alliance Standards give.org

Genetic Testing for Siblings?

I am a sibling of a brother or sister with Prader-Willi syndrome. Should I be tested to make sure I am not a carrier?"

In the past, our typical response to the question posed above was that unless siblings know their brother or sister has an imprinting mutation, there is no more of a risk than in the normal population. If the PWS was from an imprinting mutation, there is a risk, thus siblings should be tested.

We now know more genetic information, so I asked the chairs of both our Scientific and Clinical Advisory Boards (both are renowned geneticists in the field of PWS) to respond to this question.

— Janalee Heinemann

Daniel J. Driscoll, Ph.D., M.D., University of Florida Health Science Center, Gainesville, Florida

Actually this answer is incomplete and not true in all cases.

For all cases of deletions the father should be offered to be tested by chromosomes and FISH to make sure he is not a balanced translocation carrier or has an inversion.

For all with UPD the child should also have had chromosomes done to make sure it was not a result of a Robertsonian translocation. If there is a translocation then the parents may need to be tested.

Imprinting defect cases need to be worked up to determine if they are an IC deletion or an epimutation, which have very different recurrent risks.

All families need genetic counseling since many families have their own unique caveats that need to be addressed. The siblings only need to be tested if something is positive with the parents, or they are really concerned and the parents are not available.

Dr. Driscoll chairs the PWSA(USA) Clinical Advisory Board.

Merlin G. Butler, M.D., Ph.D., Children's Mercy Hospital, Kansas City, Missouri

I agree with Dr. Dricoll's comments. The type of FISH analysis (with the correct collection of DNA probes) is also critical (need to use a centromeric probe along with the q11-q13 probe to make sure that the chromosome of interest contains the chromosome 15 centromere without an acrocentric chromosome translocation involvement).

I am aware of a family with the mother having a balanced translocation between chromosomes 14 and 15 involving the proximal long arm regions whereby a male child with Angelman syndrome was born. He inherited the chromosome translocation but was in an unbalanced form. He inherited three chromosome 14 regions involving the long arm (termed partial trisomy of proximal 14q) from the mother and had only one copy (from the father) of the long arm of chromosome 15 (termed partial monosomy of proximal 15q). Therefore, he had a deletion of the proximal long arm of chromosome 15 from the mother and Angelman syndrome occurred.

The translocation in the mother (or child) would not have been identified with routine chromosome studies. It required the use of FISH analysis, but without the use of the correct collection of FISH probes used, the chromosome abnormality would not have been identified and interpreted correctly.

The bottom line is to stress the importance of genetic counseling and to identify a genetics center in the patient's region. The genetic testing results on the patient should be reviewed with the family and additional genetic testing ordered, if needed. Complete genetic testing is required before accurate genetic counseling can be undertaken. The recurrence risk information for some families is not as simple as we sometimes are led to believe without appropriate genetic testing.

Dr. Butler chairs the PWSA(USA) Scientific Advisory Board.

From China, Praise for PWSA(USA)...

Many thanks for your and board of PWS-USA supporting to establishment PWS-China. I would like to say I am very lucky to have made so many friends like you, although I haven't seen some of you, but I can feel your warm hearts.

Although some patients have been properly diagosed of PWS by my colleagues and I in China using cytogenetic test, they are not properly managed because of lack of specific organizations that can provides services for PWS patients and their families, as well as doctors who are interested in the study of the disorder. As you know, I was invited to attend the 25th of PWS-USA meeting last year, I learned so many things at the meeting and shared my experiences with my colleagues in China. Since then, we've been making every effort to establish a PWS Association in China and have the first National PWS conference in Beijing in 2005. I think our dream will come true soon. I appreciate that you call us heroes, but we are only doing what we should do. It was nice to meet Giorgio [Fornasier] in Orlando last year. He gave me three educational packages, including General Awareness, Crisis and Medical Awareness. I am translating them from English to Chinese now. I also received Prader-Willi Syndrome-An Overview for Health Professionals from [Diane] Spencer, who is a very nice lady. All of the materials will be useful for Chinese doctors, educators and parents of PWS. Best regards to all of you. **— Jinghua**

Editor's Note: This doctor has been working with others to establish a PWS organization in China.

Highlights - International PWS Scientific Conference
Christchurch, New Zealand - April 10, 2004

Editor's Note: Part 1 of two parts, excerpted from abstracts presented at the Scientific Day. A complete Scientific Conference Abstract booklet can be ordered through PWSA(USA) at a cost of $20.

Clinical and Laboratory Correlates in Patients With PWS Experiencing Cardio-Respiratory Complications From Obesity: Keys to Early Recognition and Treatment

Linda M. Gourash, M.D., James E. Hanchett, M.D.; The Children's Institute, Pittsburgh, PA

Discussion: The insidious nature of cardiopulmonary deterioration in persons with PWS whose weight has reached the obese range is evident in the fact that cor pulmonale and hypoxia are frequently undiagnosed at the time of referral. Numerous case histories demonstrate that cardio respiratory decomposition often occurs seemingly without warning in patients who have appeared to be tolerating their obesity without imminent risk.

PWS: Phenotype, Cognitive and Behavioral Patterns

Suzanne B. Cassidy, M.D.; U.C.I. Medical Center, Orange, CA

Discussion: Reviewed typical physical findings and natural history of PWS; discussion of implications of treatment with growth hormone, review of diagnostic and testing criteria for PWS. The implications of changes in facial features and body structure (more normalization) were discussed.

The Genetics of PWS: Deletion, Uniparental Disomy and Imprinting Defect

Daniel J. Driscoll, Ph.D., M.D.; University of Florida Health Science Center, Gainesville, FL

Discussion: PWS is a contiguous gene syndrome which results at the loss of several imprinted genes that are paternally inherited. Three main molecular classes include: 1) 3-5 Mb deletion of the 15q11.2-q13 region; 2) maternal uniparental disomy (UPD); and 3) imprinting defect (ID). Of 131 patients followed at our institution, 70% are deletions, 27% UPDs and 3% IDs, frequencies that are in good agreement with those obtained by other groups. Various genetic tests are available to the clinician, including high resolution chromosomes, fluorescent *in situ* hybridization (FISH), DNA polymorphism analysis and DNA methylation. In any suspect case of PWS, all children less than age 2 need a SNRPN DNA methylation test for diagnosis.

Mouse Models of PWS — How Similar to PWS Are They and What Will We Learn From Them?

Daniel J. Driscoll, Ph.D., M.D., Anthony P. Goldstone, Karen A. Johnstone and James L. Resnick; University of Florida Health Science Center, Gainesville, FL

Discussion: The three main molecular classes of PWS in humans have been recapitulated in mouse by various investigators to learn the function of the various imprinted genes in the PWS region. Although mouse models are similar to their human counterparts in some respects, to date, no one has created a mouse model that closely resembles all the major clinical features of PWS, particularly the obesity and hyperphagia. Thus it would seem that while the PWS mouse model represents an excellent genetic (and imprinting) representation of the human PWS region, there are significant phenotypic differences with humans with the identical genetic defect.

Ghrelin and Peptide YY Genetic Studies in PWS

Merlin G. Butler, Zohreh Talebizadeh, Natasha Kiriryeva, Douglas C. Bittel; Children's Mercy Hospitals and Clinics and University of Missouri-Kansas City School of Medicine, Kansas City, MO

Discussion: A hallmark of PWS is insatiable appetite leading to early childhood obesity. Study objective is to better understand abnormalities in appetite-regulating pathways in PWS, specifically at the gene level.

Results and Conclusions: Gene expression studies showed no correlation between plasma ghrelin or PYY level and lymphoblastoid gene expression data. The expression level for receptors was measured in five brain regions in a PWS-deletion subject female and a control female, which yielded differing levels.

Clinical-Etiologic Correlation in Children with PWS: An Interdisciplinary Study

M. Terrado, L. Chertikoff, E. Baialardo, V. Araoz, K. Abraldes, G. Krochik, B. Ozuna, S. Caino, V. Fano, C. Maza; Cerrvicio de Genetica, Hospital de Pediatria "Prof. Dr. J.P. Garrahan, Buenos Aires, Argentina

Discussion: A clinical-etiologic correlation study of 91diagnosed PWS pediatric patients was divided into two groups (deleted and non-deleted) and followed by an interdisciplinary team.

Conclusions: Advanced maternal age facilitates UPD group. A greater compromise of variables (i.e., hypopigmentation, feeding problems, sleep disturbance, speech articulation defects) was observed in the deleted group. Most significant differences found are related to CNS functions. These differences must be related to the underlying differences in gene expression of the genetic subtypes.

Ghrelin and Other Gastrointestinal Hormones in PWS

Anthony P. Goldstone, MRCP, Ph.D.; University of Florida Health Science Center, Gainesville, FL

Discussion: Hormones from both the gastrointestinal tract and adipose tissue regulate appetite, primarily through interaction with the hypothalamus. Investigation of possible causes of hyperghrelinaemia in PWS and its consequences on food intake. Further studies are needed to identify whether somatostatin plays a significant role in appetite regulation.

Sex Hormones in PWS — What Aspects of the Phenotype Do They Determine?

Kate Steinbeck, Ph.D FRACP; Royal Prince Alfred Hospital, Camperdown NHSW, Australia

Discussion: Oestradiol, testosterone and adrenal androgens are all hormones which increase in the peri-pubertal period and are responsible for the major and gender diverse changes seen in this period. Changes occur in both oestradiol and testosterone pre- and post-natally in the presence of intact hypothalamic function, which may have ramifications for future phenotype. In earlier literature it has been difficult to garner effects of both growth hormone and sex hormones in those with PWS, but increased clinical use of GH has helped to clarify this. Sex hormone replacement therapy has had major

Highlights continued on page 9

Advisory for Care Providers

Exploring the Dangers of Positional Asphyxia

By Tina DiDino, REM Ohio, Health Services Coordinator
Mary K. Ziccardi, REM Ohio, Administrator

Safely using physical restraint

Recently, one of our families lost a child as the result of a prone restraint. The purpose of this article is to address the use of physical restraint, and when needed, how to practice it safely.

Often school and vocational organizations and residential agencies are trained to use face-down restraint procedures. This type of restraint greatly increases the risk of positional asphyxia, which, simply stated, means that the position of one's own body interferes with the restrained person's ability to breathe and the person cannot get enough oxygen.[1]

Further, any interference with respirations, either by the positioning of one's own body or by an external force, may result in asphyxia. The danger of death is so great that many police departments are now conducting officer training of alternative restraints and have banned the use of prone restraint. According to EMT Charly Miller,[2] one case study showed the average time between beginning a prone restraint and the onset of death was only 5.6 minutes. Use of this restraint, even for short periods of time, presents serious risks. Multiple factors place a person at risk for death or serious injury from positional asphyxia.

Risk Factors and PWS

Primary risk factors for a person with Prader-Willi syndrome include:

- one's body position during restraint, particularly prone
- obesity[3]
- prolonged struggle or physical exertion[4]
- respiratory conditions, including asthma and bronchitis[4]
- pre-existing heart disease, including an enlarged heart and other cardiovascular disorders [3,4,5]

General Risk Factors

When a prone restraint is used, each of the following factors may put a person at risk:

The individual may become upset, and his/her heart rate, blood pressure, and rate of breathing will increase.

As the physical struggle occurs, the person becomes out of breath. More oxygen is needed to fuel the struggling muscles. As the person's body is trying to get more oxygen to fuel the muscles, the person is placed in a face-down position on the floor, causing compression of the chest and limiting the person's ability to expand the chest cavity and breathe[5,6]

In addition, the abdominal organs may be pushed up, restricting movement of the diaphragm, further limiting the available space for the lungs to expand.[6,7]

Also, excessive body weight makes it more difficult to move the chest wall and expand the lungs, especially when in the prone position.[4]

All muscles, including the heart, require oxygen to function. When the heart doesn't get enough oxygen, it may begin to beat faster, or out of rhythm, potentially leading to death.

According to a case study,[8] forensic pathologist Dr. Werner Spitz indicates that there is a greater chance of positional asphyxia with increased body mass and an enlarged heart. The amount of fat located under the navel is indicative of the thickness of the fat layer under the skin, and this is associated with excess fat inside the abdominal cavity. In the prone position, the excess fat, together with the abdominal organs, push against the diaphragm, causing it to be immobilized, and ultimately interferes with breathing. Dr. Spitz concludes that the more agitated a person is, the less time it will take for suffocation to occur.

As the individual continues to struggle, more weight is added and the grip is tightened to further gain control. As the person continues to struggle to get away, exhaustion is a factor and breathing becomes more difficult. Life is threatened.

PWS Adds to Risks

In children and adults with Prader-Willi syndrome, poor muscle tone and excess weight add to the risk of prone restraint. When an authority figure assumes a "control model" mentality, restraint may become the accepted way of handling a crisis. Restraint is then used as intimidation, punishment and convenience, or for control, as opposed to a final option if imminent physical danger exists.

As a parent exploring residential, educational, and vocational opportunities for your child, it is important to ask questions regarding the support staff's philosophy in restraint use.

Are restraints used to prevent immediate harm or as threats, punishment, or control? If restraint is deemed appropriate by the team, is it part of an overall Behavior Support Plan, and have all applicable authorizations been completed? How have staff who will be implementing the restraint been trained? In extreme cases, restraint must be used to protect someone from imminent danger. Ultimately, it seems advisable to teach support staff safe and appropriate physical restraint techniques. This may prevent even well-intentioned support staff from harming someone out of anger, fear, or frustration.

Asphyxia continued on next page

Asphyxia - *continued from page 8*

Prevention is Key

Consistency, structure, trust and positive feedback are critical to building supportive relationships with people with Prader-Willi syndrome. Some additional positive strategies may include:

- Always reward the positive! High-fives, enthusiastic praise and small tangible reinforcers go a long way.
- Warn in advance when there is going to be break in the routine or a schedule change.
- Do not nag! This may only increase agitation and frustration.
- Acknowledge when you are wrong and apologize. It is helpful to know that we all make mistakes.
- Build a relationship that accepts and values each person.
- Acknowledge and empathize when a situation is unfair or upsetting, and follow up with appropriate responses and potential solutions.
- Anticipate questions and try to predict solutions during transition times.
- Don't hold a grudge! Learn to treat each minute as a new opportunity. Sometimes, it's the only way to survive.
- Never promise something that could potenntially change.
- Handle sneaking food or other crises in a matter-of-fact way. It's their nature to try and it's our job to to prevent!
- Don't minimize the difficulty the person is having. While it may seem trivial to you, it can be of great importance to an individual with Prader-Willi syndrome.
- Anticipate the outcome you are trying to generate. For example, is there value in getting the person to admit to telling a tale? Avoid unnecessary power struggles simply to prove a point.

People providing services and supports to those with Prader-Willi syndrome are strongly encouraged to explore alternatives to prone restraint. Restraint use should be considered as a safety response, not as a therapeutic technique or substitute for a comprehensive treatment plan. It is critical to remember that when all is said and done, this is a human life — someone's child, sibling, grandchild and friend — and this person, too, has value.

References

1 Schaars,Christopher; (Hampton Roads, Virginia) Daily Press; April 13, 2003.

2 Charly Miller is an internationally known EMS instructor and a restraint asphyxia expert witness. The information above is referenced from an article on her web site "Restraint Asphyxia-Silent Killer," updated August 2002.

3 Paterson, B., Leadbetter, D., and McCornish, A.; "Restraint and Sudden Death from Asphyxia," Nursing Times (94 44), pgs. 62-64, Nov. 4, 1998.

4 O'Halloran, R.L. and Frank, J.G.; "Asphyxial death during prone restraint revisited: A report of 21 cases," The American Journal of Forensic Medicine and Pathology, 21(1) pgs. 39-52, March 2000.

5 Paterson, et al.; O'Halloran, et al.; and Stratton, S.J., Rogers, C., Brickett, K. and Gruzinski, G.; "Factors associated with sudden death of individuals requiring restraint for excited delirium," The American Journal of Emergency Medicine, 19,(3), pgs. 187-191, May 2001.

6 Parkes, J; "Sudden death during restraint: a study to measure the effect of restraint positions on the rate of recovery from exercise," Medicine, Science, and the Law, 40(1), pages 39-44, January 2000.

7 Reay, D.T., Howard, J.D., Fligner, C.L. and Ward, R.J.; "Effects of positional restraint on oxygen saturation and heart rate following exercise," The American Journal of Forensic Medicine and Pathology, 9(1), pages 16-18, 1988.

8 Case study in Protection and Advocacy, Inc. Investigations Unit report by Dr. Werner U. Spitz, "The Lethal Hazard of Prone Restraint: Positional Asphyxiation," Publication No. 7018.01, p. 26, Oakland, CA, April 2002.

NOTE FOR CARE PROVIDERS and HEALTH PROFESSIONALS:

For a complete article on dangers relating to positional asphyxia, contact the PWSA(USA) National Office, 1-800-926-4797 or go online to www.pwsausa.org

Highlights - *continued from page 7*

effects on bone and the cardiovascular system which are not externally apparent but which have ramifications for health and well-being. A number of unusual clinical situations in PWS subjects provide insight on how functional disturbance of the hypothalamic-pituitary-gonadal axis may itself be altered in PWS and point to potential future therapies.

Growth Hormone — Overview of Production, Rationale and Benefits of Use in PWS

E. Martin Ritzen; Karolin's Institute, Stockholm, Sweden

Discussion: Production of biosynthetic human growth hormone (GH) in large enough quantities permitted controlled studies relating to PWS beginning in the mid-1990s. Generally, in most children with PWS, the first year of treatment induces rapid growth, increased muscle mass and physical capacity, and marked reduction of body fat if a calorie-restricted diet is maintained. However, some children respond better than others. GH treatment does not influence intelligence, but parents reported that their children took more initiative and played more with others than before. There is only one published study on the effects of GH in adults with PWS; effects on body composition are similar to that in children, but less pronounced.

A Family's Key Principles to Help Your Child with PWS

Editor's Note: It is natural and normal for parents to worry about the future of their newborn child. As we talk with the parents of children who have been newly diagnosed with PWS, a reoccurring question is, What does the future hold for my child? None of our children comes with guarantees, so no parent really knows what is in the future. We believe that with research and new management techniques, our children with Prader-Willi syndrome can achieve beyond our expectations. The following letter from a parent who prefers to remain anonymous instructs new parents how to spend their "worry energy" on some truly beneficial activities from the very first day that new little baby comes home with you.

Dear Parents of Young Children with PWS,

This letter is written to honor the memory of the late Dr. Robert Wharton, who taught us four key principles for helping a young child with PWS: Stimulate * Human Growth Hormone * Food Rules * Socialization

It is our relationship with Dr. Wharton and his advice that has transformed our lives. With Dr. Wharton's help, we went from fearing that our baby would have mild to moderate cognitive deficits, obesity and severe behavior problems, to contending with some learning disabilities, watching what our daughter eats and limiting some of her behaviors.

Dr. Wharton was a developmental pediatrician who devoted much of his work to helping children with Prader Willi syndrome. He had a knack for focusing on what could be done to help the child's quality of life. He cared for the children and families like no other and excelled at simultaneously comforting and motivating the parents.

Through his work with and devotion to PWS, Dr. Wharton developed strong beliefs about the syndrome. He believed that early diagnosis, human growth hormone treatment and early intervention brought a "Second Generation" of children with PWS. He thought that children who were born with PWS were lacking in survival skills: poor suck, weak cry, reduced movement. Dr. Wharton identified some key areas where intervention by doctors and parents would make an enormous impact on the child and his/her capabilities and happiness.

Stimulate

Dr. Wharton told us he did not believe that mental retardation necessarily was a part of PWS. He believed that it was a result of the insufficient caloric intake due to the poor sucking instinct and a lack of stimulation due to the sleepiness and low arousal of the babies with PWS. He believed that if the babies received the proper amount of calories and were awakened by the parents and caretakers and stimulated, the level of cognitive deficits could be reduced or eliminated.

He noted that these babies do not give back the same way other babies do by smiling, giggling and asking for more. This lack of response in the babies makes many parents feel that their parenting is inadequate and that they are not bonding with their baby. He counseled that the low response is part of the syndrome, but does not mean that the child is not bonding or loving. The parents need to be sensitive to the more subtle feedback of their child with Prader Willi syndrome. As the parents are *persistent* in *stimulating, playing games* and *connecting*, the child will develop the capability to love strongly and to interact socially.

Human Growth Hormone*

Dr. Wharton was an early advocate of Human Growth Hormone (HGH). He advised us to give it to our daughter and we started her at 6 months of age. That was 6 years ago. There is the obvious benefit of increasing the average height of the children. Dr. Wharton believed even more strongly in the benefit that HGH improves the fat to muscle imbalance that the children have. By improving the fat to muscle ratio, it helps in controlling weight, gives the child more power and endurance, and helps close the developmental gaps between children with PWS and their peers.

Our Food Rules

Dr. Wharton said, "Your child lacks the ability to feel full. It is your job to give that child another very consistent signal that she is full." He thought that the children should be kept slim, as once fat cells develop, they might be playing a role in signaling to the brain that the child is still hungry. The formula he gave us was 9 to 11 calories per cm of height. We developed food rules that we stick by (shown in the box on the next page).

Our goal: to establish disciplined eating habits. If and when our daughter is "full," we will have other means of signaling that eating is finished — one plate and no other food. So it is not that one cookie will lead to obesity, but it is the establishment of habit and strict discipline that is critical.

Principles continued on next page

Our Food Rules

- All snacks and meals at table when at home
- Snacks away from home in "snack bag" and of a fixed quantity
- No extra snacks in car or place to keep child quiet
- Fill plate and/or bowl at start of meal
- No seconds
- If our child wants more, create distraction or excitement over next activity
- No bites or tasting from other plates
- No tasting or snacking when child is present, unless one is sitting with her and eating from a plate
- For birthdays, special events or cooking exercise at school (making gingerbread as a class), child may have one cupcake or cookie — preferably a small one — and no seconds
- Make fewer exceptions, and the child will ask for fewer exceptions

Our three other children also follow these food rules (although they do not follow the 9-11 calories per cm). The food rules have benefited us all. There is no begging in front of any candy display, no hassles over meals. We place the food out and if there are any comments, our standard response is, "It is your choice: you can eat it or not eat it."

Socialization

Dr. Wharton and his wife, Dr. Karen Levine, believed that ultimately many of the children with PWS were not happy because they naturally were social children who did not socialize well. These children needed extra help to learn to engage socially. Dr. Wharton and Dr. Levine told us about "floor time" — a way to play with your children and engage them and teach them while you're playing. You can read about it in a book, "*How to Raise Children with Special Needs" by* Stanley Greenspan. Getting your child with other children, having more children if it's a possibility, and prompting your child to get involved are all things that can help lay the groundwork for a more fulfilling healthy socialization later in life.

PWSA (USA) gratefully acknowledges the printing and mailing of this newsletter is made possible by a grant from CIBC World Markets Corp./ Miracle Day USA.

Conclusion

So, the bad news is, our children have Prader Willi syndrome and will have it for every day of their lives. The good news is, there are steps you can take that will help your child. For us, helping our daughter and doing everything that we could do to help her develop and be more capable and happy made us feel better. Somehow, doing for your child cuts through the denial, creates acceptance of the diagnosis and fate, but brings great pleasure and hope to the present and future.

This letter is really a tribute to Dr. Wharton. We were blessed to have known him for almost 6 years and we want to spread his words and advice that we believe have been critical to improving our daughter's life, and we hope that they can improve the lives of other children and bring hope to the families!

All our love and luck for a bright future,
Another family with a child with PWS

*** NOTE: For important updated information on risk factors, please carefully read the PWSA (USA) Clinical Advisory Board consensus Statement titled *"Evaluation of Breathing Abnormalities with Sleep Apnea in PWS"* by PWSA (USA). Call the national office for a copy at 1-800-926-4797 or go to our web site: www.pwsausa.org.**

The Chuckle Corner

Another Family's Sibling Cooperation

Here is a conversation at breakfast between my just-turned 5-year-old Vincent and his brother Anthony, age 8, who has PWS.

Vincent can read the word FAT on food labels. He was looking at the cereal box contents and said, "Anthony, this has fat in it. I will eat all the fat for you because I do not have PWS."

Anthony: "OK, Vincent, that will work good."

Then Anthony turned to me and said, "Mom, I better get that part of my 15 chromosome back before Vincent gets too fat!"

Johanna Costello, Uniontown, Ohio

Please send your joke or funny story to the PWSA (USA) office. Be sure to include your name, phone number and address in case we have any questions.

Sibling View

The Day That Skylar Ran A Marathon for Jessica

By Yvonne Travis

My son Skylar Travis, age 17, recently ran the Vancouver Marathon to raise money for PWSA(USA) in honor of his sister Jessica, age 13, who has PWS.

Skylar's excitement in doing the marathon was infectious. His 26-year-old sister ended up joining him. She finished in 4 hours and 15 minutes. And their dad, the 53-year "old" man, who did not train at all, and whom I told specifically, "Do not run this marathon," dropped the kids off in the morning for the race, parked the car, signed up and also ran. He finished in 5 hours and 45 minutes.

Since Dad-the-designated-photographer was so busy running the race, we have no pictures of the event. All we have is a lot of good stories, and a good sense of achievement in working hard to help others.

Skylar was a little embarrassed to tell this part of the story: As it turned out, his feet began to really hurt after about mile 11, and he really wanted to stop. All he could think of, though, was how so many people had put their faith in him, and how much he really wanted to finish the race so he could send the money in on behalf of his sister.

This kept him going, and he somehow managed to finish the rest of the 15+ miles of the marathon. His finish time was 5 hours and 13 minutes, and he managed to come in 11th in his age group.

By this time, however, Skylar's feet were hugely swollen, and as more time passed, the pain became excruciating. He was unable to walk, and had to take a wheelchair to get through the airports to get home. There he went straight to a podiatrist, who found that he had stress fractures in both feet. Turns out he was prone to this because two of the bones in his feet are congenitally fused and therefore do not flex properly. The stress of all that running caused them to fracture.

He was unable to put any weight on them, hobbling around on crutches, and missed quite a bit of school, which bummed him out. And the real downer was missing his year-end tennis team regionals.

But the achievement of finishing the marathon and the excitement of sending in the nearly $1,600 in donations did help temper his disappointment.

Now here's the rest of Dad's story: Upon arriving home, Dad spent the next several days explaining how he never had any intentions of running, that he just started off to give the kids some support and to take some nice pictures, and then somehow it just ended up being easier to finish the race than to turn back.

Since accidents do happen — although quite honestly, 26.2 miles has never "accidentally" happened to me or anyone else I know — and because he did not fall over dead of a heart attack like I was so afraid of, I had to forgive him. Unfortunately, however, since Dad-the-designated-photographer was so busy running the race, we have no pictures of the event. All we have is a lot of good stories, and a good sense of achievement in working hard to help others.

Thanks to PWSA for all that you do. Jessica was diagnosed with PWS only about 2½ years ago, and it has been a real life and sanity saver to have your resources available to us.

Yvonne Travis and her family live in Salt Lake City, Utah.

Robin - *continued from page 1*

became. The stronger I was, the easier it was to conquer the daily food wars.

In addition, during several opportunities — positives and negatives — to assist people with special needs, I had the realization that it could be worse. That is exactly right. There are worse battles that I could fight. Luckily, though I was dealt a hand that I didn't prefer, still it is not a terrible one. Arriving at this conclusion helped me appreciate the things I have going for me.

So without further ado, I would like to encourage you to sit down with those you care about to cooperatively create lifelines (a.k.a. guidelines) to assist in your experiences with PWS. It will make your life and the PWS individuals' lives brighter.

Also, I am disturbed that almost everyone I meet has a negative outlook on PWS. There needs to be more optimistic outlook on PWS. I hope this article will help shed some more optimistic light on the issue of PWS. If there are any other pressing matters regarding my accomplishment and how I manage, feel free to contact me through the PWSA(USA) National Office or by e-mail to rwmacgillivr@stthomas.edu. I would be glad to answer questions. Hey, maybe you'll even find me speaking at a PWS conference some day.

Janalee Heinemann

Executive Director's View

When Life Changes Forever

~ Adam Romagnoli ~
10/28/85 – 5/19/04

Just 1½ weeks prior to his death, Adam walks his mother Tracy down the aisle at his stepsister's wedding

At 10:15 AM on May 19, I received the call that is every mother's greatest fear. It was my 40-year-old daughter Tracy calling from the accident site, sobbing and saying, "Mom, Adam's dead, Adam's dead!" An instant before, her 18-year-old who was tall, handsome, outgoing, and an honors student, had taken two short breaths and died. In that instant, my daughter's only child, who was also her best friend, was taken from her forever without her ever getting to say good-by. In that instant, our lives were forever changed. Instead of going to his graduation that week, we went to his memorial service. We were so looking forward to his moving to our area in Florida to attend an international business college, but instead, we will be spreading his ashes in the sea he loved so much.

When we got to Texas, Tracy was curled up in a ball on the couch and kept mumbling, "I don't know what to do, I loved him so much." Feeling helpless, I told her, "All you can do right now is try to survive." Tracy cried out, "But he was my reason for surviving!"

For me, the worst pain was, and will continue to be, knowing I cannot protect my daughter from her grief and pain. Through my more than 20 years of experience with bereavement groups for parents whose children have died, I have been a frequent and intimate visitor in the world into which we have now all been suddenly thrust. I would give anything to protect my daughter, her husband, and our family from what I know we must go through – but I cannot.

I've always been well aware that life is not fair and bad things happen to very good people. Knowing I could not protect my children from great tragedy, I tried to model for them how to live with it, and how to embrace people in pain rather than run away from them.

Many of you have shared with me the agony of finding out your child had Prader-Willi syndrome and having to face that the hopes and goals for the child you dreamed of having died with the diagnosis. Some of you will understand the helpless hurt of being a grandparent and seeing your adult children suffering emotional pain from which you cannot protect them. Some of you have faced the ultimate loss yourselves of your own child or grandchild.

So, how do we go on from here? What can make life worth living again for my daughter? I can only hope that the messages I tried to teach her and the strength I tried to instill in her will keep her going until life feels worth living again. I never made giving my children an easy life my goal. Having worked with child abuse, children with cancer, hospice, and worked and lived with PWS, I have always been well aware that life is not fair and bad things happen to very good people. Knowing I could not protect my children from great tragedy, I tried to model for them how to live with it, and how to embrace people in pain rather than run away from them.

Just two days after Adam's death, Tracy said she knew she would need to find a cause to survive. She was able to show compassion for all of the teens who were grieving, and to appreciate all that her church community did to support them in any way humanly possible. To me these are glimmers of hope. I just wish I could ease the questions that haunt all grieving parents: "Why?" "If only…" and "I wish I would have…"

We often hear it said that God will not give you more than you can bear. I say, Life will give you more than you can bear sometimes. God just helps you through it.

I know the huge hole in Tracy's heart will never go away, but my prayer is that each day brings her closer to finding reasons to live again. That is my prayer for all grieving parents.

We Remember...

Every person has something special to offer this world — and we, along with their families, want to share who they were and what they meant to the people who loved them.

Farewell to a Friend – Siobhan McGee

On March 7, 2004, our long-time volunteer, 44-year-old Siobhan McGee, died from complications of lupus. Prevailing over serious health problems and renal failure, for 6 years Siobhan would come to the office three times a week to volunteer. This was in spite of chemotherapy and many medical complications. In the last year, she would hook up her dialysis bags in our office and continue working.

Siobhan died peacefully with her "family" of PWSA (USA) staff by her side throughout the weekend.

Before she died, Siobhan arranged for the love of her life, her dog Ferghus, to go to my oldest daughter Tina and her husband John. Tina lives in the St Louis area and rescues dogs. Tina has 13 acres fenced in for the dogs, and they have full access to both indoors and outdoors, along with their own lake.

Siobhan McGee and her beloved Ferghus

Tina sent us videos and pictures of her place, but Siobhan, with her usual dry wit, said she would not show them to Ferghus because, "He might bump me off early!"

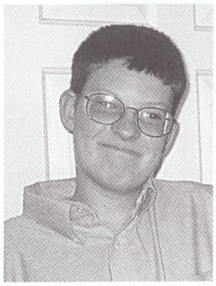
We will miss Siobhan's dedication and humor. I thank all of our staff and volunteers for giving Siobhan a loving and supportive family in her final years.

— Janalee Heinemann

View From The Home Front

Michael and Marie: Children with Prader Willi Syndrome

A Book Review by Whitman Kerric Donaldson



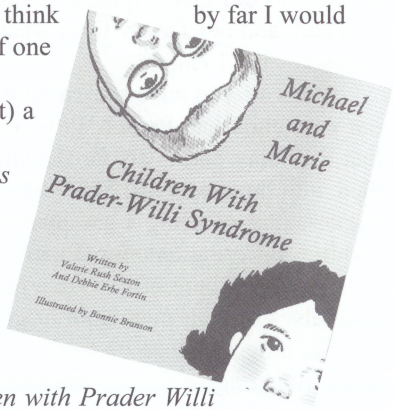
Michael and Marie: Children With Prader Willi Syndrome is perfect for explaining to your child about his or her disability; Prader Willi syndrome. It also is great for explaining to your child's classmates and getting them to understand it as well. This book talks about all the important things about the syndrome.

I especially like how it tells the readers how kids with Prader Willi Syndrome are also a lot like other kids, although there are a lot of differences. It tells about growth hormone and how people with the syndrome have weak muscles. It talks about eating only healthy foods, how parents need to be strict about food and maybe even literally locking up all the cabinets.

It says how kids with Prader Willi syndrome have many friends despite their differences. It explains how people with the syndrome have behavioral problems such as temper tantrums. It also explains something that I think is quite hard for me and maybe many others with this disability — that we have trouble with schedule changes and coping with the change as well.

Another important topic that it talks about is that people with Prader Willi syndrome might get stuck on topics, and can't move on. Also a person with the syndrome has frustrations over ideas and starts over many times before it is just right for them. It also talks about the special people that work with children that have Prader Willi — the occupational therapist, the physical therapist, the speech and language teacher, the paraprofessional and even the psychologist that the kid might see is explained.

I am a 14-year-old with Prader Willi and absolutely love this book as well as enjoying reading it over and over again. This is a great book for families and all children that have Prader Willi Syndrome. I think by far I would give this book on a scale of one through 10 (meaning one being awful and 10 perfect) a 10.



Whitman lives with his family in San Francisco, California

Editor's Note:
Michael & Marie: Children with Prader Willi Syndrome, was written by speech language pathologist Valerie Rush Sexton and special education paraprofessional Debbie Fortin. You can order the book by phone to 1-800-926-4797 or by mail to the PWSA(USA) National Office. The cost is $8.00 plus $1.50 shipping (if only ordering this one item).

Angel Fund

Golden Harp Society ($1000 plus)
Robert & Barbara Gallagher *Memory-Claire Gallagher*
Choir of Angels ($500-999)
Linda Carson *Honor-Benjamin Dawley*
Joyce Shepard
Golden Angel ($250-499)
Mercedes A. Lat
Angel ($100-$249)
John & Susan Alexander
Gary & Cindy Arstein-Kerslake
Manny Emmanuel
J. Brian & Gail R. Murphy
Kate Zawistowski Walk-a-Thon
Kate Zawistowski *Memory-Catherine Zawistowski*
Cherub (Up to $99)
Marsha Gamelin
Alan Geller
Mary Jane Gemmell
John & Christie Gibson & Family
Alan Gober
A.J. & Lillian Goldberger
Marion Goodman *Memory-Jill Goodman*
Gow Family
Lisa & TJ Graziano
Suzanne Greskovics
Hilda T. Hahn
Mike & Linda Hartnett *Honor-Reilly & Callaghan Hartnett*
Lori & Andy Hayden
Arnold & Tricia Hearn
Harry G. Hedberg
Charles & Barbara Heidel
Marilyn & Ralph Heim
Lowell & Jan Heinemann
Raymond & Kathleen Henderson
Cheryl R. Hinds
Marion J. Hogue
Margaret Hollingsworth
Doris Jean Houck
Barbara Houck-Carson
Harry E. Houseman
Carolyn Hutchens
David Ingalls *Memory-Elwin Ingalls*
Jean T. Janes
Phyllis Jen
E.Ray & Bonnie Johnson
Diane Taylor & Joyce Burke
Joan Wilson Kaiser
Mary Helen Kane *Honor-Kate Kane*
Tom Kane
Linda & John Kania
Richard Karlis
Connie Kates
Carole Kearney
Daniel & Sharon Keder
Thomas & Lechon Keenan
Jay Kelly *Honor-Steven Forthman*
Kathy Kenahan
Steven & Roberta Kenney
William Kirchhoff
Laurie & Paul Kitchin
Eleanor Kittleson
Ken & Maria Knox
Diane Koontz *Honor-Jackson Donaldson*
Sara P. Krapf
George & Pamela Kraus
James & Susan Lazarus
Pam Lewis
Kenneth & Antje Liley
Chad & Betsy Lindsey *Honor-Jack Lindsey*
F.A. Lindsey *Honor-Jackson Donaldson*
Cyr & Anne Linonis
Alan & Kathleen Luening, Sr.
Catherine Macdonald-Cooley
Ernest & Alice Marek
Terri & Bill Marks *Honor-Zoe Marks*
Richard & Vivian Marson
Donna Martin
Clara Matthews
Carol Matza
Martha McDonald
Bob & Marilyn McDonough
Calman M. Menzer
Sharon M. Milar
Kathleen Minor
Lota & Dave Mitchell
Kathryn & Glenn Moffett
John Mook
Patricia Mook & Family *Honor-Crosby Mook*
Robert & Cynthia Moser
Fran Moss
Chris & Kristen Mumford
Edward & Ruth Needel
Michael & Kay Neff *Memory-Donald Wagner*
Robert Nordin
Thomas & Shirlee Nystrom
Lisa Oppenheim
Rosemarie & Jim Ostiller
Wilma J. Palmer
Gene Pami
Catherine Partridge
Jean Peterson
Joseph & Elizabeth Preiss
Rudolph Prochazka
Frank & Gloria Rangitsch
John & Michele Raniolo
Ralph & Margaret Reimer
Jay & Linda Reinhardt
Howard & Patty Rosenthal
Michael Ruoss
Daniel P. Ryan
Bud & Nancy Savage *Honor-Jackson Donaldson*
Fred & Kathleen Schloessinger
Mary L. Schneider
Donna J. Schramm
Ron & Helga Schwarz
William & Julie Seiger
Donna Siegel *Honor-Louise Greenswag*
Sandra Singleton *Honor-Savannah Brice*
Don & Marie Sinon
Herdis Skulason
Elizabeth & Kevin Slimak
Carl & Cheryl Smith
Curtis & Vicki Smith
Norman & Joyce Smith
Sandy Smith
Mike & Jeanne Spradlin *Honor-Christina Spradlin*
Susan & James Sprowl
Bernard Starr
Tammi & Gary Steinberg
John & Pam Stocking
Eleanore Stuart
Beatrice Sturm
Stephen C. Suden
Kathryn B. Swenson
Thomas & Helen Taylor
Stanley & Tracy Thorp, Jr.
Ellen Thurber
Christina & Michael Turcola
Virginia G. Turner
Gene & Geri Ulland
John Valant
Hector Valdepena
Bonnie Ventura
Ray & Susan Wagner
Gerald & Mary Walsh
D.E. & Sara H. Ward *Honor-Mr. & Mrs. Everett Henry*
Kathy Way
Lynn & Debbie Weidenbach
Eugene & Nancy Woo
Melinda Wyatt
Peter & Kathleen Wyka
Darla Yocum
Linda A. Zick
Douglas & Nancy Ziliox *Honor-Jacob Douglas Ziliox*
JoAnne Riley Barron
John & Alvina Frisch
Joseph M. Murray
Eileen Posch
James & Karen Ward

In Honor of
Jacob Hunter-Evan Chymbor's Uncle Lindsay Nelson
Christian Coats
Alyssa & John Husby
Jessika's Birthday
Ann & Karl Everts
Ashley Fender
Wayne & Karen Wendel
Alexandria Gotoh
Robert & Marjorie Walcutt
Student Max Sznaj-Griswold School Staff
Margaret & Gregory Sznaj
Amy Hammer
Judith Lyon
Barbara Hupperich
Peter & Barbara Hupperich
Madison Riley Hurdle
Tara Firenze
Clinton & Karla Hurdle
Jack Joseph Kuna
Mike & Lori Kuna
Janis Leightman-Happy Mother's Day - Ida & Ray Leightman
Charles & Patrice McCleary's 50th Anniversary
Helen McCleary
Mr. & Mrs. Brian Moore's Wedding
Kathleen Brannan
Eloise Brannon
Helen Brooks
Barbara Esterbrook
Emily Joy
Fern Shafer
Andrea Saacks
Dennis & Sharon Saacks
Elizabeth Thorpe
Mary Allgood
Vancouver Int'l Marathan-Honor of Jessica Travis
Jon & Arlene Aoki
Ronald & Kathy Aoki
Margaret & James Avent
Leah & Scott Brockway
Gerry Brown
John & Andrea Burke
Jessica Travis *(continued)*
Regina & Joseph Drueding
Anne Yang Dwyer
Kyle & Julie Enslin
Brett & Lisa Friedman
Colin Gardner
Kyle Lee & Jill Garrett
Scott Gutting
Henry & Patrice Hemingway
Robert & Carolyn Hoffman
Verna Huiskamp
Edwin & Sharon McGough
Paula Paterson
W. Bradford & Charlene Rockwell
Brian & Janice Ruggles
Michael & Colleen Schneider
Madalyn Sue Seaman
Homer & Patricia Smith
Jean A. Stachon
June & M.W. Steinvorth, Jr.
Edwin & Joann Svikhart
Laird & Gloria Swensen
Ritsuko Travis
Yvonne & Nick Travis
John & Constance Ward
Paul & Bonnie Weiss
Virginia White
Allison & Robert Wright
David & Diana Yang
Michael Yang
Maria Vucci
Maryann Rutt
Taylor Warren - The Taylor Project
Ellisworth Bell
Gregory & Theresa Blitz
Jeffrey & Jeannie Fulbright
Roy & Vicki Hightower
Robert & Rachael Male
Norman Minton
Marlene Pearman
Neal & Angie Spradlin
Andrea & Bob Warren
Sheridan York
Beverly Kosztyo
Laverne Schultz

Contributions

Thank you for Contributions Received
March - May 2004

Board Member Fund
Jack & Kay Goldberger
Erich & Pauline Haller *Memory-Adam Romagnoli*
Michigan Annual Awareness Walk
Central Michigan Non-Profit Housing
Israels Design For Living
Showboat Automotive Supply, Inc.
Pinnacle Construction Group
Hubble's Body Shop, Inc.
National Nail Corp.
C L Trucking & Excavating, Inc.
Forest Area Federal Credit Union
Mark & Alice Alcorn
Scott & Barbara Anderson
Peter & Alice Arends
Catherine & Jon Beagle
Thomas & Pamela Bingham
Brent & Cindy Bowen
Joanne & Richard Brown
Alonzo & Barbara Burdine
Patty Burwell
Matthew & Kimberly Cairy
Rich & Suzanne Ceppos
Tyler Chambers
Edward & Sharon Clock, Jr.
Todd & Tania Cook
Sharon & Karl Cressman
Barbara Cummings
R J & Jill Dean
Beth & John Deindorfer
William & Nancy Doman
Cheri & Keith Feldman
Cindy He & Gerald Lewis
Michelle Gier
Janet Gilbert
Cami & Mark Gillette
Shirley & Dick Gilmour
Francis & Joan Hart
Richard & Jane Hawkins, II
Christine & Jon Hendrick
Nancy & Douglas Hendrick
Jeffrey & Janet Hessler
Jan Hetherington
Bobbie Hilliard
Kim Ann & Terrence Holmes, Jr.
Joann & David Holwerda
Auburn & Gloria Hubbard
Pamela & Kurt Jaeger
Mary Jankowski
Daryl & Lori Johnson
Hannah Kelbey
Harold & Linda Krueger
Kimberly Largen
Joyce Mannor
Janet McPeck
Neal & Jane Mellesmoen
Fred & Dorothy Mendricks
Kathy Mieras
Brenda Myers
Kenneth & Elizabeth Patin
Margaret C. Patterson
Brian & Nicole Powell
Walter & Kathryn Powell
Wayne & Dawn Pratt
Ronald & Nancy Prieskorn
Patrick & Charlotte Pullen
Chris & Michele Reimer
Teresa Rich
Edward & Joanne Ridgway
Russell & Susan Rosema
Signe Ruddy
Tom & Lorrie Russo
Doug & Ruth Rutherford
Roy & Teresa Sanderson
Richard & Linda Saranen
Jerilyn Scheid
Marisela & Kevin Shellenbarger
Barbara Solomonson
Patricia & Jerry Stahl
Ronald & Bonnie Stull
Helen Thomas
Sandra Ullenbruch
Barbara J. Venema
Family Sponsorships
Cinda & Curt Ball
JoAnne Riley Barron
Suzanne Brice & Family
Sandy & Frank Ditta
Raymond & Phyllis Fuja
Debbie & Nathan Jacobus
Steve & Tami Lindner
Eugene & Jean Alice Metzger
Jonah & Kimberly Storr
2004 Conference Grants
Mayim Hoya Bialik
Matching Gifts
American Express Foundation-Robert J. Baummer
Microsoft Giving Campaign - Richard L. Critchlow
JP Morgan-Katherine Grivakis
Operating Fund
United Way of Armstrong County
United Way of Brevard County
United Way of Delaware
Tolland High School
IBM Employee Services Center
IBM Retiree Charitable Campaign
United Way of Northeastern NY, Inc.

Due to space limitations, donations will continue in the next *Gathered View.*

Contributions In Memory Of

Guy C. Barron
Betsy Ford
Margaret Bailey-Schwartz
Larry Sissom
Mildred Cable
William & Judy Castle
George Castle
George & Adelaide Castle
William & Judy Castle
Goldie Effune
Janice & Alan Ehrenreich
Naava & Ken Ellis
Francine & Leon Feinerman
Lewis & Linda Herman
Joan & Ron Isaacman
Mark & Wendy Liebermann
Dr. Sanford & Elaine Silverstein
Barbara & Steve Suden
Ann & Mark Vergales
Jerri Evetts
Brian Jorgenson
John & Bonnie Kraft
Harold & Linda Krueger
Jan & Millard Wallis
Edith Falk
James & Linda Carraher
Charles Hatcher
The FIS Team
Sequel
Rikoon-Carret Investment Advisors
Cooley Godward LLP
Haselwood Services & Manufacturing, Inc.
Joseph & Barbara Curtiss
Norbert & Lynn Ensslin
Doyle & Karen Hembree, Jr.
Edward & Judith Kaplan
Adrian Pidlusky
Michael W. Self
James & Virginia Tape
Linda Trocki
Elinor S. Hearn
The Dorsey & Whitney Foundation
Eurnice Anderson
John & Renetta Kidder
Allan & Eunice Sortland
Howard Sortland
Barbara Hupperich
Edith Sparough
Margie Levine
Eleanor & Richard Weiner
Ruth Millman
Manley Bennett
Harold Olson
Sandra Singer
Allan Porter
Earl & Carole Glover
Delores Parker
James E. Price
Ann & Donald Lee
Robert & Jeannine Stayer
Anne Ryan & Richard Trimble
Lota & Dave Mitchell
Joseph Schmidt
Joseph & Billye Schmidt
Michael J. Schaffer
Invesmart of Austin, Inc.
Richard Wett
Jon & Irene Joseph
Loren Larson
Stewart & Bronnie Maurer
Lota & Dave Mitchell
Bob & Mary Ritter
Dorothy Thompson
Jon & Marcee Thompson
Adam Romagnoli
James M. Manson
George & Katharine Tizzano
PWSA of Pennsylvania
C. Lee Richards
Marion Goodman
Marcia & Judd Dunn
Donald & Barbara Dorn
Siesta Key Chamber of Commerce
Judy O'Leary
David & Vicki Knopf
John & Marilyn Bintz
Linda & Mark Ryan
Lota & Dave Mitchell
Annie & Ned Durell
PWSA of Michigan
Tina & Chuck McGovern
James & Carolyn Loker
Robert & Wauneta Lehman
Jane Phelan
Louise Greenswag
Paul DiPlacedo & William Johnson
Ed & Jacquelynn Torcicollo
Kimberley Short
Tim & Carol Hearn
Henry & Susanne Singer
Michelle & Tommy Torbert

Prader-Willi syndrome (PWS) is a birth defect first identified in 1956 by Swiss doctors A. Prader, H. Willi, and A. Labhart. There are no known reasons for the genetic accident that causes this lifelong condition, which affects appetite, growth, metabolism, cognitive functioning and behavior. The Prader-Willi Syndrome Association (USA) was organized in 1975 to provide a resource for education and information about PWS and support for families and caregivers. PWSA (USA) is supported solely by memberships and tax-deductible contributions.