



The **Gathered View**

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

Feeling Grateful in the Midst of Apprehension

By **Betsy Ford**

Not a day goes by that I don't look at the golden little boy I have been entrusted with and give thanks for the wonder that he is. He is unique, loving, tender, excited, *alive*. His four years have seen things many of us don't see in a lifetime of medical procedures and conditions, and he takes it all in stride.

He is a gift, and I wouldn't trade this child, beset with a genetic condition many would describe as a burden, for anything in the world.

These sentiments were renewed for me especially strongly this summer. My son and I had the opportunity to attend the 2002 National Prader-Willi Syndrome Conference in Salt Lake City, Utah, from July 11 to 13. There were some hard moments, but without a doubt it was an absolutely exceptional experience. I want to share a bit about what I felt as a first-time attendee.

We arrived in Salt Lake City excited and a bit apprehensive. It was our first national PWS conference, and I was more than, how can I describe it.... Nervously curious? Anxiously interested?

Well, whatever I was feeling exactly, the time had arrived for us to face what had frightened me from the time of Jacob's diagnosis: scads of Prader-Willi kids and their parents, all in one place, all asking questions and sharing insights and learning from one another — watching one another, quietly comparing their kids with my kid, those people's problem areas with these other people's difficulties; assessing where each set of genetic manifestations fit with which part of the syndrome, which part of the ongoing research, and which part of the group, the statistics, the average Prader-Willi situation.

Gads. Maybe it wasn't too late to fly home.

No, I had been lucky to come here this year. A single mother,

I am raising my special son on one income in an expensive area of the country. Generous grants from both National and my local PWS chapter greased the way and put me over the edge financially. There would be no turning back now.

My son Jacob, my mother and I, arrived in Salt Lake and registered for the conference. I began seeing little, mostly blonde

kids, glasses slipping down their noses in many cases, little faces and hands bearing the tell-tale signs of a genetic misfortune, meekly appearing with mommies and daddies from all over the country.

My little Jakey, a lover, like many of the other kids, noticed the little faces immediately as well. He seemed taken from the start with the fact that, for once, he could run around with little people who were actually his size, his speed, who seemed to peer back at him with the same quiet curiosity, the same ancient eyes, and share a familiarity and language imperceptible to those outside their strange world.

I felt grateful for my son. I felt grateful to be here, to see him connect, recognize himself, and smile at what he saw.

The conference opened with an inspiring speech by a young man named Jason, who had the misfortune to have become a quadriplegic due to an accident in his teens. Jason really set the tone as he brought us, through loud laughs and quiet tears, into his living room and into his life. You can't be depressed, he said, when you feel grateful. You can't see only bad, he had observed, when you allow yourself to see how much good there is.



Jacob Ford, age 4, who has PWS

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The Gathered View welcomes articles, letters, personal stories and photographs and news of interest to those concerned with Prader-Willi syndrome.

Communications regarding *The Gathered View* or PWSA membership and services should be directed to the national office of PWSA (USA) in Sarasota, Florida.

New Members Added to the Scientific Advisory Board

By Merlin Butler, M.D., Ph.D., Scientific Board Chair

The PWSA (USA) Scientific Advisory Board (SAB) is composed of scientific investigators who have actively contributed to our understanding and scientific knowledge of Prader-Willi syndrome (PWS).

There are usually about 10 members, representing a wide range of expertise relating to PWS, including clinical and laboratory geneticists, psychologists, and endocrinologists. They are physicians and scientists who are knowledgeable about PWS and actively engaged in studies either through basic research (e.g., behavior, genetics, use of animal models) or applied clinical research (e.g., outcomes of growth hormone treatment). All have expressed an interest to willing volunteer their time to participate as active members of the SAB.

This group of interdisciplinary scientists meets annually during the PWS conference, usually following the Scientific Day Session, to discuss research issues pertaining to PWS, including grant applications and providing scientific expertise, collaboration and future direction to PWSA (USA). There are liaison members from the governing and clinical advisory boards attending these meetings in order to assure collaboration

PWSA (USA) now approved for the Combined Federal Campaign

PWSA(USA) is now in the Combined Federal Campaign (CFC) brochure for charitable organizations. This is the federal government's version of the United Way, and all employees of the federal government are encouraged to give through the CFC. This includes postal employees, veterans hospital employees, the FBI, the Secret Service, etc.

Our PWSA (USA) identification number for CFC is 9858.

The CFC annual campaign drive usually starts around October/November, running through December 2002.

For Federal government workers, each agency will have designated CFC coordinators to collect contributions. These coordinators will carry forms and brochures with them for co-workers to review and decide to which agencies they will make donations.

Employees will use either payroll deductions for ongoing bi-weekly payments, or give via cash or check.

Coordinators are totally objective in canvassing employees. They are not allowed to push for any particular organization; otherwise an obvious conflict would occur.

So PWSA (USA) needs to get the word out to families and friends of PWS children. Family members connected to the PWSA (USA) who work for government agencies can tell their co-workers about PWSA (USA) and how their contributions can help.

Government employees wanting to help can then contact their designated CFC coordinators to get the material necessary to donate their contributions to PWSA (USA).

Only by telling families and friends of Federal government workers that PWSA (USA) is now an eligible charitable organization and giving them the CFC identification number 9858 will this become a value to our association.

and summarize information to share with their respective boards.

During the year, the SAB members are asked to review and critique grant applications submitted to PWSA (USA) for possible funding, review and help develop consensus statements for PWSA (USA), and discuss future scientific directions and missions. They also respond via e-mail to the Executive Director concerning various questions involving pertinent research and the networking of researchers.

At the most recent meeting in July, a decision was made to invite new members with additional expertise to join the SAB. These new members include:

- **Ann Scheimann, M.D.**, a pediatric gastroenterologist with expertise in GI problems that occur in PWS. Dr. Scheimann is affiliated with Johns Hopkins University.
- **Harriette Mogul, M.D.**, an adult endocrinologist with expertise in treating adults with PWS with growth hormone and obesity in general. Dr. Mogul is affiliated with New York Medical Center.
- **Sue Myers, M.D.**, a pediatric endocrinologist with expertise in treating PWS children with growth hormone. Dr. Myers is affiliated with St. Louis University.
- **Joe Donnelly, Ed.D.**, an internationally known researcher in obesity, recently involved with obesity studies in PWS. Dr. Donnelly is affiliated with the University of Kansas.

We welcome the new members and know that they will become important to SAB and PWSA (USA). Their presence will increase our diversity and expertise as we enter a new era in the understanding and treatment of PWS.

Raise \$\$\$ for PWSA (USA) with The Coolest Gadgets in Cyberspace

How far did you walk yesterday? How many calories did you burn? What percentage of your weight is fat? How much muscle have you gained?

These questions, so essential to the lifelong health of all people, especially those with Prader-Willi syndrome, can be easily answered and raise money for PWSA(USA) at the same time.

In addition to the full line of Oregon Scientific fitness electronics: pedometers, heart rate monitors, and body fat percentage scales, CyberGadgets also offers lots of electronic gadgets, which make great gifts. **Plus, sales of all items can earn a 25 percent donation to PWSA(USA).**

Just go to www.cybergadgets.com and place your order. As you check out, find the box that asks where you heard of their site. Type in PWSA(USA) and 25 percent of your order total will be donated to PWSA(USA)!

Lori DiCola, Prader-Willi Alliance for Research



Love Overflowing Into Doing Something For PWS

By Lota Mitchell, PWSA (USA) President

A couple weeks after returning to Pittsburgh from the national conference in Salt Lake City, I drove the 450 miles to North Carolina for granddaughter Colleen's sixth birthday party and then brought her home with me for a visit. We had a delightful week or so here of fun and activities; then it was time to take her back to NC.

I learned later that as I drove away from her house, Colleen burst into tears and cried and cried. Shannon, her mom (and my wonderful daughter-in-law), held her very close and told her, "Those tears show all the love you're feeling. When there is so much love inside you, there just isn't room for all of it. And so it just brims over and comes out in tears in your eyes."

Isn't that the way we often feel about our children with PWS? We love them so much that it overflows into the tears in our eyes. Sometimes the love is mixed with grief at the loss of the normal child we had expected, or fear about what the future holds, and sometimes with determination to do something for the cause of PWS.

The "something" can take many forms — reaching out to other parents to offer support and understanding, doing fund raisers, getting involved in our state and national organizations, contributing our time and money.

The chapter presidents I've been writing about in a number of issues of *The Gathered View* have carried out their resolution to do "something," and they work hard to do it. Their normal load these days is made even harder by the fact that many of our chapters are struggling with a viewpoint difference between the parents of the young children and the parents of the older children and adults.

The leadership challenges they face are significant. Leadership is needed which can create an atmosphere where those of differing views can still work together to fulfill the purposes of the group.

We are proud of our chapter presidents and their commitment, and we wish for each and every one of them the energy, the tact, and the wisdom, to meet those challenges successfully. Here are a few more for you to meet.

NEW YORK — A new chapter president in 2002, **Daniel Angiolillo** was elected a Supreme Court Justice of the state of New York three years ago. A resident of Westchester County north of New York City, he has been on the board of the New York chapter for six years and secretary for the past four years.

Doing "something" for PWS can take many forms — reaching out to other parents to offer support and understanding, doing fund raisers, getting involved in our state and national organizations, contributing our time and money.

The Angiolillos' daughter with PWS, age 17, is the oldest of their three children. So far they have not had to lock up the kitchen cabinets.

NORTH CAROLINA — Another new chapter president in 2002, **Sally St. John** is a single mom of two children and a school teacher. Son Elliott with PWS, 17, lives at home where everything is locked. He is an avid University of North Carolina fan. Sally enjoys going to the beach, camping, the outdoors and reading.

KENTUCKY — The mother of four boys ages 18, 16, 13 and 10, **Kim Settles** is deeply involved in her kids' activities. She describes herself as a stay-at-home mom. The youngest, Patrick, has PWS. He is very social, in Special Olympics and has many activities.

PENNSYLVANIA — **Maria Silva** is mom and stepmom to six—husband Steve's two, her two and their two. The youngest is Ari, age 8, who has PWS. A woman with many talents and involvement in several organizations, Maria was Chairman of the PWSA (USA) national conference in Pittsburgh in 2000.

MD/DC/VA — Making use of her journalistic background for PWS, **Linda Keder** was the editor of *The Gathered View* for several years and just this past fall completed the highly acclaimed PWSA (USA) booklet on Growth Hormone. She has volunteered her writing and editing skills on behalf of PWSA (USA) for many years. Daughter Leslie, 13, who has PWS, is an only child.

Reuniting With Our Brazilian Angel

By Pam Eisen, PWSA (USA) IPWSO Delegate

Once again my "sister," Helena Portugal (from our twin country, Brazil) and I were united. This time I had the pleasure of introducing her to all who attended our exciting 2002 conference at Salt Lake City. Throughout this experience, I was keenly aware of the Angels Among Us.

As you may recall from the past few issues of *The Gathered View*, Helena, a nurse, educational psychologist, and the single mom of a 15-year-old with PWS, established the PWSA of Brazil in January 2002. In a nation of 150 million people, there was a paucity of information on Prader Willi syndrome, from diagnosis to management. Helena had a prodigious dream to educate professionals and parents – to make a difference in the lives of children and families dealing with the challenges of PWS. With the spirit of unity, United States and Brazil initiated the first IPWSO Twinning Project.

In the short life of this project, huge accomplishments have occurred and are a testament to the power of international sharing and cooperation. With help from IPWSO and PWSA (USA), Helena has made many contacts and has had access to necessary information and research. As twins, we've supplied educational material and constant support while Helena, on an unequivocal course, has performed miracles.

On her own, she has translated publications, visited hospitals and clinics, educated and recruited professionals to join her in her mission. Among her many accomplishments, Helena published an article in a pediatric journal, was interviewed by a prominent reporter for a national newspaper, appeared on a national educational TV program, and has convinced the government to make a declaration providing a disability pension to mothers and their children with PWS.

In October, a PWS Awareness float (with a popular children's musical group performing) will appear in a grand parade in Rio de Janeiro. Presently, Helena is working with the Brazilian government to establish a multi-disciplinary program for PWS in already existing neighborhood clinics.

Helena's quest has drawn much attention in her country, helping several families secure a diagnosis. In June the first official Brazil PWSA was held with 15 families present!

Knowing that Helena and her organization needed more information on research and management of PWS, PWSA (USA) awarded her a conference grant, and Helena joined our membership in Salt Lake City. Her radiating presence at the conference reminded us of our mission and enriched our lives.

A warm, intelligent, vibrant woman, Helena instantly charmed all, made new friends, collected every possible morsel of information, and observed each detail.

Helena's presence brought an overwhelming and generous response from our members. We set up a booth with pictures of Helena, Fefê, and other members of the Brazilian association. For sale was a beautiful array of colorful Brazilian art, painted by Helena's artist friends. As Helena left for the airport with \$1,521, she was already thinking of how to best spend the money for her newfound organization.

With all of the recent discoveries and new possibilities for our children, we must remember there are families in this world who are just as much in the dark as we were before the birth of our organization. Knowing the difference that education about early diagnosis, new medical breakthroughs and management of PWS can make, we must reach out throughout the world, providing information and support. Joining together, we have hope and can all fly with the angels!

A Thank You From Helena...

As we hugged good-bye, Helena asked me to thank her beautiful "American Angels" for providing her with this invaluable experience. In her own colorful, touching words she wrote:

"My experience in this Conference was quite fantastic! It was everything that I needed to have forces, and not to give up, of this long walk to help our PW children."

The affection and the love to the neediest countries... the certainty and the gratefulness to God and the science, mainly of those that were favored by the early diagnosis... the most apparent peacefulness, a more tangible hope contemplated in the small children's parents' attitudes...

I will never forget all those children jumping... the grief and the glance friend... the attempt of softening, through experiences, the problems...the tears and the smiles... mainly of the oldest children's parents...

All this learning... the books, the scientific material so wanted... a child's hope in each sentence, in each folder... I thank to God, to Giorgio, to my family...TO OUR AMERICAN FAMÌLIA... aH! I am very proud of being part of this MARVELOUS FAMILY!

Thank you for the opportunity to have participated, of having lived together, of having divided, learned and known such special persons...

I remember each gesture, each glance, each hug... of the unconditional love of all those that were to my side physics and spiritually, during the whole event....

THANK YOU ON BEHALF OF ALL THE CHILDREN FROM BRAZIL... AND AFFECTIONATE BLESSINGS OF ALL OF US PARENTS, RELATIVES AND FRIENDS OF OUR BRAZILIAN ANGELS.

Helena Portugal





Another Mother is Crying.... Another Child is Dying

By Janalee Heinemann

On Sunday, our crisis counselor, David Wyatt, was discharged from the hospital.

Tuesday morning, with an IV line threaded from his arm to his heart and his leg covered with a medical stocking, he came to work.

Before he got a chance to sit down, the phone rang, and within five minutes, he was talking to a crying mother who has multiple sclerosis (MS) and a 160-pound acting-out 9-year-old daughter with PWS. Her MS is getting worse, and she has begun dropping things. To add to her concern and grief, her daughter was suspended from school due to her behavior and stealing food. Mom feels she is losing her ability to control her daughter's foraging for food, and she is gaining weight rapidly.

David's next call was from a desperate couple who were driving around in their car rather than going home to face their 10-year-old daughter. They had just come back from a plane trip, where their daughter got out of her seat to go the bathroom and sat down where her parents could not see her. She then rang for the flight attendant and said that she had not gotten her snack — so the attendant brought her several snacks.

"What was I to do?" the dad asked. "If I took the snacks away and she created a disturbance on the plane, in this day and age, who knows what would have happened!"

One other of the many stories these parents told David was about their daughter breaking into a neighbor's home to get food. Although these neighbors did not file charges (as happens in many cases), they refuse to unload and lock up the loaded guns they keep in their home.

The next call was about a 12-year-old with PWS in Moldova (a former part of the Soviet Union) who is chained to a radiator in an orphanage. The American mother who was calling had lost her son who had PWS last year at age 18. This mom had befriended via e-mail the mother of the child in Moldova, and the two had been communicating for seven years. The American mom would adopt her Moldova friend's child, but due to the political climate, there is a freeze on adoptions.

Next, David opened his email. One was from a mother who wrote, "HELP!!!!!! My daughter has PWS. She is 9 years old. For the past 2 years she has been having hypoxemia, cyanosis, edema (predominantly of the left lower extremities) and she also has SEVERE scoliosis. She is having spells where she is lethargic, pupils are dilated, she gets a lazy eye, and her body is either mottled or ghostly pale. When she is having one of these spells, she can't think clearly. Any help would be greatly appreciated."

These are the calls and e-mails that fill David's day — and sometimes spill over into mine.

While David was in the hospital, I took all the crisis calls, and ironically, we had calls on three families from Tennessee, all dealing with weight gains so enormous that the medical complications are probably irreversible.

One child is 12 years old and well over 300 pounds. He was admitted to the hospital for congestive heart failure and is on a ventilator. He lives with his grandmother.

Another is 16 and over 360 pounds. This child has been in state custody since 1997 — yet still allowed to gain this much weight! He now is dying of obesity-related causes. The third is 24 years old, five-foot three inches tall and 448 pounds. The physician said he is low normal in IQ and a nice kid, but is now having right-sided heart failure, respiratory failure, severe edema, sleep apnea, etc.

In asking myself why, each of these cases, people had been unaware that PWSA (USA) exists, I realized that we have no state chapter in Tennessee. Although PWSA (USA) continues to improve national awareness and education about the syndrome, these Tennessee cases are glaring examples of why we need a team effort between our national organization and the state chapters.

The call from those parents driving around brought back memories buried in the shadows of my mind of our darkest days with our son Matt, who has PWS, and one particular event when Matt was screaming and crying in the car over some food we had denied him.

Neither Al nor I have volatile personalities, and we can usually handle situations calmly even when we are not feeling calm inside. But on this day, we were worn down and "snapped." We pulled off onto a country road, turned to Matt, and started hollering back — just to give him a sense of what it felt like. We then got out of the car, walked down the country lane, and sat in a ditch on the side of the road where we could see but not hear Matt crying at the top of his lungs in the car.

A farmer drove by slowly, and stopped to ask us if we knew there was a boy crying in the car down the road. We just stared at him blankly, and I answered, "Yes, we know." At that point, we did not care whether that farmer reported us to the authorities. A quiet jail cell sounded like a good respite. It is easy to block out experiences like this now that Matt is doing so well. Although there is a great deal of hope and optimism for the future of PWS, the reality is we cannot ignore that there are many parents still sitting in a ditch on the side of the road.

Taking all the crisis cases for a week while David was in the hospital was an important reminder for me that our fight for

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That talk, I remember most clearly, by itself would have made the trip worthwhile.

The hardest moments were seeing the grown Prader-Willi kids. At this year's conference there were two young men, both apparently in their early twenties. I hadn't fully prepared myself for this part. Both youths appeared physically more as you might picture, with their respective weights, postures, facial structures.

You know, in our situation, you make these deals with yourself. You figure out what you can live with and what you can handle, and you cross your fingers and groan inwardly and dream, on those bad nights, over the rest. One of the young men almost wrestled his mother down in the middle of a conference session for a bag of sun chips. I haven't had to see that yet, to deal with that. Jake isn't there yet.

Standing right next to this scene, I gazed in horror, and disbelief. Slowly, all those things I had read in the pamphlets and books, all the stories I had sympathetically listened to and nodded at in the talks, the glazed eyes I had seen on some other mothers, all the things that I had successfully avoided having to deal with up to now came crashing in on me.

I stood there, in my head desperately bargaining with God, one last time, to take this cup away from me, from Jacob. I stood there, not knowing how to move or what to feel.

As I watched, the mother slowly, fluidly almost, calmed her clenched-jawed son, gently retracted the crushed sun chips from his shaking grip, smoothed herself and him, and went on listening — quiet, resigned — as though nothing had happened.

I stared at the side of her head: her chin high, her gaze unwavering; her face, while faintly drawn, remained unmoved, as though wrestling a bag of chips from one's adult child in the middle of a speech was a normal event that might occur on any given day. This wasn't going away, was it? I stood there, closed my eyes, and, in that moment, somehow fully accepted what Jacob is.

The specifics of the rest of the conference remain a blur in my memory. I took pages of notes, listened intently to the speakers and other parents, asked my own questions, took

names and phone numbers and exchanged earnest promises to stay in touch, sealed with strong hugs and more than a few tears.

That I also got to learn a ton about the syndrome and current research, to meet nationally known doctors and to share stories with other families facing the same issues I am facing, save only for the uniqueness associated with every one of the little developing personalities involved, made the conference the trip of a lifetime.

Here as I write, months after returning from Salt Lake, I have to say that Jason had it right. What I feel most is, well, grateful. Crushed sun chips, scary possibilities, bad dreams and all.

When I see other 4-year-olds, I still realize how far behind Jakey is. I drag him off to therapy session after specialist appointment, knowing his gains are incremental, his "milestones" are more like "inch-pebbles," creeping forward along a twisting path bearing no resemblance to all the nice charts that come in the mommy books.

But I also see the sunshine and energy and life in Jakey's eyes, the delight in his laugh, and the trust and unrequited love he gives, effortlessly, to me and to his world.

He fearlessly charges forth every day, unhindered by his hindrances, and in the process melts away for me the hazy what ifs, and the sheer weight of this responsibility.

In his vulnerability, his fight to survive, he needed me to champion his cause. Unknowingly, he became my champion in the process. For all the hassles my son will have to put up with in his life, I know that there will be pain he will never have to feel. And I continue to learn from him every day how to love, how to give, how to live. This little angel, with his tiny hands and tiny heart, has saved me. Grateful can't come close to describing the thanks I feel.

Betsy Ford and her son Jacob, who has PWS, live in Laurel, Maryland..

Grieving

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awareness and education, and our continuing need for our support programs is far from over. In the past six months, I have finally had the opportunity to put more of my time into advocating for research and awareness programming on a broad scale.

I also am often haunted by the question, what happens when there is no more David, who is so right for the job and will dedicate his life to this very difficult cause for so little money?

Last week was a reminder to me that we must continue to balance our needs as an organization. Research, education, conference, support and awareness — each plays a crucial role in the fight for our children's lives. As our annual Angel Fund Drive approaches, I hope each of you will try to remember that although life is positive and hopeful for our new generation of children with PWS, we cannot turn our back on the needs of others. Every single day I remind myself that there, but for the grace of God, go I.

Praise for PWSA (USA)

Here are some recent comments from our members...

◆ There are times I ask myself "why me?" Then I'll pick up the latest copy of *The Gathered View* and re-read some of the articles. Somehow a sense of peace — don't give up — you're not alone comes through.

◆ As we learn... our simple lives are enriched from what we've been through and the wonder of what lies ahead. I was very glad to see the article on single parents as this situation is unique in itself.

Securing PWSA (USA)'s Financial Legacy

By Maureen A. Wheat

PWSA (USA) Director of Development

I am writing this on the plane as I return from the PWSA (USA) conference in Utah. Even though you will not read this until October, I wanted to share my immediate enthusiasm, hope and ideas with you for securing the financial legacy of the Prader-Willi Syndrome Association (USA).

Working with the Prader-Willi Syndrome Association, I have witnessed the many avenues PWSA (USA) uses to fulfill

Members came forward to ask how they could help guarantee the future of PWSA (USA) so that we can continue our vital work.

its mission to "improve and enhance the lives of everyone impacted by PWS and related conditions."

Fulfilling that mission means sharing hope, promise and information to all affected by the syndrome, such as the mother in crisis who told us that attending the PWSA conference helped her "gain the hope and courage to fight for my daughter's quality of life." And the teacher who thanked us for the booklets and handouts on PWS that will help to inform her as she prepares to teach a student with PWS. These stories of support offered to people in crisis are countless. Through the dedication of its chapters, members, staff and hard-working volunteers, PWSA (USA) is trying to meet the challenge of giving support to all who ask for help.

At many points during our dynamic conference, generous members came forward to ask how they could help guarantee the future of PWSA (USA) so we can continue our vital work.

The answers that grew out of those conversations were many, including volunteer support to the national office for special projects, and Chapters raising more money and donating a portion of the proceeds to insure the funding of research projects and support the everyday operating costs of PWSA (USA). These operating costs include printing and mailing the resources that are the very foundation of education and awareness about PWS. The operating costs help to cover the salaries of office personnel like Gilda and David, who field hundreds of crisis calls per month.

Seeking the dream and securing our financial future does come with a price, but the payoffs will be rewarding. Everyone will be invited to join hands and walk together toward a brighter horizon.

In the November/December issue of *The Gathered View*, I will extend an opportunity for each of you to leave a legacy to PWSA (USA).

In the January/February issue, we will have a conversation with Chapter presidents about what it means to work in community with the national association and how that journey can benefit everyone involved.

There is a phenomenal amount of compelling and exciting work before us. Just as PWSA (USA) uses many avenues to transform desperation and fear into hope for the future, we must discover new pathways to generosity so that PWSA (USA) can continue to foster more answers and a cure through research, and secure the hope of all who are impacted by the syndrome. Thank you for your commitment, generosity of spirit and the gift of your time.

Sam Beltran Back in the Saddle

PWSA of MO-KAN held their first annual PWS Trail Ride Benefit on May 4 at the beautiful Beltran home. Dr. Sam Beltran was the first board chairman of PWSA in 1977 and its national president in 1980. Sam, his wife Linda, and their daughter Sarah, 30, now live in Andover, Kansas.

Proceeds from the benefit went toward funding the formation of a new Missouri-Kansas chapter.

Joy Holmes of Hutchinson, Kansas, mother of 6-year-old Joey, who has PWS, organized the Trail Ride.

About 25 riders rode for 2 hours while the kids enjoyed fishing at Sam's pond, playing with new kittens, feeding the horses and jumping on the trampoline. The kids got a horse ride when the trail riders returned. A picnic afterwards featured great food and a real snow cone machine. The weather was beautiful and the day was perfect.

A reporter from a Wichita TV station interviewed Sam and featured the PWS Trail Ride on the evening news.

The MO-KAN Parent Support Group has been meeting for the last four years at Children's Mercy Hospital. The group receives a great amount of guidance from Dr. Sam Beltran, Dr. Merlin Butler and Dr. Travis Thompson and is looking forward to more awareness about PWS in Kansas City. Chapter members want to establish a dedicated PWS residential facility in Kansas because none are currently available there.



Dr. Sam Beltran with his daughter Sarah, who has PWS.

California Walkin' for Prader-Willi Syndrome

By Mary Hill

About a year ago, Executive Director of the Prader-Willi California Foundation, Fran Moss, asked our Northern California support group to help organize a fundraising walk for PWS.

Honestly, at first none of us really wanted to lead this effort. It sounded like a lot of work, and we had no experience with fundraisers. So I was quite surprised when I found my husband Paul at the computer one night typing an e-mail to Fran offering to help coordinate the event! But now I am so glad he volunteered — organizing and attending the walk turned out to be a great experience.

Thankfully, Fran and her Southern California crew took care of many of the administrative aspects, including the brochures, registration, T-shirts, accounting and much more. Paul found a site for the walk in our home town of Berkeley — then he asked for help from the other members of our support group. Those who could pitched in with enthusiasm — it was a true team effort. And working on the walk turned out to be a fun way for us all to get to know each other better, too.

The event was a wonderful success in every way. About 270 people showed up to walk with us on April 20th at the Berkeley Marina. And we raised more than \$45,000 (before expenses)! We were all absolutely touched by generous responses to our solicitation letters. The Southern California walk the following week was also quite successful.

One of the wonderful and unforeseen results of these two walks is that the Prader-Willi California Foundation will now be able to fund additional (and much needed) projects this year.

This is very exciting, and our local support group has been brainstorming ideas to present to the board of directors. A few of the many ideas being considered are funding outreach to Spanish-speaking families and creating a video for school personnel that fully illustrates the difficulties that children with PWS encounter in the school system. This should enable children to more easily get the extra help that they need to succeed in school. Either of these projects would be a wonderful and unexpected legacy of our walk.



Oscar Hill, age 1½, who has PWS, with his dad Paul

The walk itself was only about 1.5 miles, but we all enjoyed views of the East Bay hills, Mount Tamalpais and the San Francisco Bay.

Our older son Abraham, age 3½, decided he would ride his bike rather than walk. He was so excited when the walk finally started that he immediately sped off into the crowd ringing his bell. Our son Oscar, age 1½, who has PWS, spent much of the walk on Paul's shoulders, which Oscar loved! He bounced and clapped (and banged on Paul's head) most of the way.

Instead of name tags, many people wore stickers with a picture of the child for whom they were walking. We were hoping the stickers would make it easy for people to introduce themselves and mingle with fellow walkers. However, we only made stickers of the younger kids with PWS and really didn't have enough for everyone. We had so many requests for stickers that we gave out

some extras of Oscar. During the walk it seemed like everyone was "Walking for Oscar."

Paul made great signs and posted them along the route to direct people through the park. The signs also provided some information about PWS, and many people told us they learned a few new things about the syndrome. In addition, a couple of representatives from Pharmacia came with water and souvenir sport bottles and answered questions about growth hormone treatment. We served a PWS-friendly lunch and gave away kites as door prizes.

We met many wonderful families, friends and caregivers of people with PWS. Additionally, about 30 adults with PWS came from Northern California PWS group homes. One woman, Jean, instantly took to Oscar — she wore his sticker, held his hand — even carried him a few steps of the way. She was a delight to walk and talk with.

All in all it was a wonderful event — one that we will never forget. We are already thinking about ways to improve for next year: more stickers, bigger signs, better outreach.

It's almost time to start planning again, but no one will have to twist our arms to help out this time!

Mary and Paul Hill live with sons Abraham and Oscar in Berkeley, California.

Update on PWS Research

By Janalee Heinemann, Executive Director

The following is from a presentation from an outstanding scientific research panel at our national conference in Salt Lake City, Utah in July. An audiotape of the entire presentation is available for members only through our national office for \$10. To order, call 1-800-926-4797.

In response to our new mission statement of expanding our mission to supporting research in obesity, we have had contact with several researchers in the obesity field.

Ghrelin Studies

Dr. David Cummings from the University of Washington is studying a gastric peptide which may be a factor in regulating appetite in PWS. Dr. Cummings' initial research on ghrelin was recently published in the New England Journal of Medicine.

Individuals with PWS have been found to have abnormal levels of ghrelin, a hormone produced by the stomach. It is the only known appetite-stimulating hormone made in the stomach, which makes it much more feasible to find a treatment to reduce ghrelin levels.

Ghrelin not only increases appetite, it decreases metabolism. When people attempt to diet, their ghrelin levels rise, which fights against weight loss.

The ghrelin levels in both children and adults with PWS are dramatically higher than in the "normal" population, thus the interest in PWS for future research.

Although ghrelin levels drop with gastric by-pass surgery, Dr. Cummings does not advocate this type of surgery for PWS at this point. One of the reasons is the risk that someone with PWS might still be hungry, but with little stomach in which to put food, this could create severe complications.

As we have said many times, the answer to all obesity may be through Prader-Willi syndrome. Now the world of research is observing what we have known for a long time and is taking an interest. The possibilities are very exciting for our children.

Another ghrelin researcher who presented at our Scientific Conference is Dr. Andrea Haqq, from Oregon Health & Science University. She has measured the ghrelin levels in children with PWS and also found their levels to be extremely high.

The Bear Study

Dr. Ralph Nelson, Director of Research at Carle Foundation Hospital is conducting a Bear Study relating to the etiology of excessive appetite in persons with Prader-Willi syndrome. Dr. Nelson has been studying the control of appetite in bears for more than 25 years.

Twenty years ago he wrote a chapter in our first Management of PWS book on clinical nutrition, and has never forgotten PWS. He hopes that his study of bears will be helpful in finding a way to control appetite in persons with PWS.

Bears are remarkable because they are able to go many months without eating or drinking. They prepare for this hibernation, or "denning" period with several weeks of ravenous foraging and feeding. In other words, during a normal

We will keep you updated with articles in *The Gathered View* on the progress of research. You will only receive *The Gathered View* with a PWSA (USA) membership. Call 1-800-926-4797 become a member today.

year, there is a time in which the same animal eats as much as can be found, and then later consumes absolutely nothing. There are several substances that may control this process.

Every parent of a child with Prader-Willi knows that in the first two years of life, getting your child to eat is a problem, since the appetite is usually poor and sucking difficult. Of course, by age 4 to 5, most children with PWS begin a period of runaway appetite.

Dr. Nelson's research team is comparing substances that control this process in the blood of both the child/adult with PWS. (PWSA (USA) was able to get private funding for this study to expedite the research before denning season.) Our hope is that these studies of bears will be helpful in finding a way to control appetite in people with PWS.

More on Appetite

Although it was not reported until after the conclusion of our Utah conference, another new player in the world of appetite is PYY3-36, or PYY for short. It has been dubbed "the fullness hormone." PYY switches off the appetite. The problem at this point is that PYY is only effective in injectable form because it is destroyed by stomach acid.

Both the September 2002 edition of *Discover* magazine and *The New York Times* have published articles discussing how Prader-Willi syndrome is a window to understanding the complex system that regulates metabolism and appetite.

PWS will soon be part of a *60 Minutes* segment dealing with current research on appetite. Watch the PWSA (USA) web site for the air date and time.

CoQ10 Studies

Dr. Daniel Driscoll of the University of Florida and Dr. Robert Nicholls of the University of PA are pursuing research studies on CoQ9.

Note: We are quite aware of the flurry of interest in CoQ10 being used for PWS, with some parents reporting dramatic improvement in energy strength and others reporting no improvement. Until we have scientific information through well-designed controlled studies, PWSA (USA) is currently not able to make a specific recommendation for or against the use of CoQ10 in the medical treatment of individuals with PWS.

Meanwhile, we are not ignoring the issue. Since our

Research continued on page 11

A Grandparent Reports on Conference 2002

By **Barbara McManus**

While I have been involved with PWSA (USA) for the past three years, this was the first conference where I represented our New York State organization. I found the session on research exciting (read more about this in Janalee Heinemann's article on page 10), and enjoyed the smaller conference model that was introduced this year in Utah.

Research

The doctors associated with PWSA (USA) are on the leading edge of current research in conditions related to PWS. As an organization we can pool our resources through our willingness to volunteer our dollars and our support to make a real difference for our people with PWS. I know that together we will accomplish so much!

I think we'll hear more about this in the near future. To read more on the research session at the conference go to: <http://www.pwsausa.org>

The Smaller Conference

The Utah conference was our first attempt at a smaller, less problematic method of state-hosted conferences. The programs for the children were limited to the 0-8 age group. The general conference was designed the same as most previous ones, with programs targeting all age groups.

I talked to many parents who had traveled to this conference without their children, and they reported that it was great to network with the other parents without their children and the daily obligations. Some parents brought along someone to watch the older children while they attended the conference. I hope more will attend the next smaller conference in 2004 wherever it may be held.

The smaller conference makes it more appealing for state chapters to take on the obligation of hosting a conference. Maybe someday soon, we in New York State can host one of these smaller conferences. It was a great learning and networking opportunity.

Barbara McManus of Niagara Falls, New York is a grandmother to Jessika Dickinson, age 10, who has Prader-Willi syndrome. Barb has volunteered to be Secretary to PWSA (USA), co-webmaster to the National website, monitor to the age 6-12 eGroup and now a director of the New York Alliance. You can e-mail her by clicking on the link for Webmaster on the PWSA (USA) web site, www.pwsausa.org.



Jessika Dickinson, age 10, who has PWS, is the granddaughter of Barb McManus.

Research

continued from page 10

conference last summer, we have been working with researchers in an attempt to encourage and support CoQ10 research.

Dr. Driscoll and Dr. Nicholls have begun initiating the process of a study on CoQ9 treatment in newborn PWS mouse pups to determine whether this treatment helps get them through the neonatal failure-to-thrive phase (CoQ9 in mice is considered similar to CoQ10 in humans). It may be noted that there are currently mouse models for the three main molecular classes of PWS.

There are reasons for doing both a CoQ9 study in mice and a human trial. Among the advantages of a mouse study are: 1) It will cost less money and is easier to get approval for the study. 2) All tissues are available to assay biochemically, and strains of mice can be used that allow a good control population. 3) If CoQ9 or any other treatment helps keep the mice alive, this will also serve an important purpose of allowing us to do further research on adult PWS mice, since at present these mice are dying as neonates.

We will keep you updated with articles in *The Gathered View* on the progress of research. You will only receive *The Gathered View* with a PWSA (USA) membership. Please remember, no parent is denied membership. If parents cannot afford to pay, they need to notify us, but every family should become members of PWSA (USA).

The Chuckle Corner

At the 2002 national conference in Salt Lake City, all meals were provided. The children each morning came to the office area to receive their box breakfast, which contained among other items different cereals.

When 4-year-old Danica arrived to pick up her breakfast, she was asked, "Would you like MiniRaisin or Granola?"

In the spirit of Prader-Willi, she responded with a resounding "YES!"

*Grandparents John and Sue Alexander
Whittier, California*



Do you have a joke or funny story to share with readers about Prader-Willi syndrome? Send it to the PWSA (USA) office. Be sure to include your name, phone number and address in case we have any questions. We'd love to hear from you!

Thanks to PWSA (USA) Staff for Help With Home Placement

We would like to thank Janalee, David and the PWSA (USA) staff for the time and assistance you offered us to achieve our goal of getting our son who has PWS appropriately placed at the Gilbough Center in Cape Cod, Massachusetts.

Matthew had his first home visit with us from August 31 to September 3 for the first anniversary of his twin brother Jonathan's passing away. Jonathan also had PWS. This year has been a terrible struggle for all of us, especially Matthew. Our getting Matthew placed in a home with other Prader-Willi people has given us a sense of purpose for Jonathan's death. We try to see it as a Gift from Jonathan to have made this all happen for Matthew.

Matthew has lost 55 lbs. He now weighs 176 and has 20 lbs. to go to reach his goal. He has many new friends his age. He had told us that losing Jonathan was a double loss for him. He not only lost his brother, but his best and only friend.

Danny, Justin, Colin and Maxx are housemates and new friends. They have really helped Matthew deal with his loss and loneliness. Danny reminds Matthew a lot of his brother

Jonathan, and they bonded from the moment they first met.

Matthew keeps telling us how lucky he is to have such a nice home to live in, and in a resort area, too!

He is very excited because Cape Cod Community College offers a 2-year program for the disabled. He just sent in his application to take the course on Large and Small Animal Care that they offer. There is a waiting list, but he is hopeful he will get in next year.

In September Matthew is participating in a job training program at a local horse farm. He is very excited about it, as both he and his brother loved horse and were in the Equestrian Special Olympics. Matthew now has a brighter future, thanks to so many caring people that he has never even met. We have told him of all the wonderful efforts that were made for him.

We will be forever grateful to for whatever part you played in helping Matthew to have a future. We will try to keep you updated as new things happen. We will always remember you in our thoughts and prayers. Thanks again for your time and support to our son.

Irwin and Roberta Langewisch, Milford, Connecticut

Plan now for PWSA (USA)'s 2003 – 25th Annual Conference

Mark your calendar for July 2 – July 5, 2003.

Bring the family and spend the Fourth of July just minutes from Walt Disney World Resort and Universal Orlando and within walking distance to Sea World. Our goal is to have a group event for the evening of July 4th that you will never forget!

July 2nd will be a day of outstanding programs for the scientific community, and also for providers. Our general conference program will have separate sessions for parents and caregivers of the 0-5 population, school-age children and adults. As with our 2002 conference, we are planning to have "The Latest In Research" presentations for all attending the general conference, plus a summary of the scientific presentations.

Our conference will include a **Youth and Infants Program as well as a Youth and Adult activities program**. All but the infant/toddler groups will visit one of the major theme parks as part of their program.

The **Sheraton World Resort** is the headquarters hotel for the PWSA (USA) Conference. This recently renovated, year-round resort features three outdoor swimming pools, a fitness center and lighted tennis courts. PWSA (USA) has secured a block of rooms at the discounted rate of **\$98 a night** for up to four in a room.

To avoid any overcharge or problems with registration, please make your room reservations by contacting Globetrotter Travel at 800-322-7032 (press 2), or by e-mail to pwsa-usa@globetrottermgmt.com. Hotel reservations may also be made online by contacting www.globetrottermgmt.com/pwsa-usa

All reservations require a credit card guarantee or a check deposit of the first night's room and tax. Checks should be

made payable to Globetrotter Travel. This deposit is non-refundable if canceled within 72 hours of the arrival date.

For special airfare and car rental rates, please call Globetrotter Travel at 800-322-7032 (press 1) or e-mail to pwsa-usa@globetrottermgmt.com to take advantage of the special airfare and car rental discount rates. Conference attendees can save up to 15 percent on airfare. Discounted car rental rates have been negotiated for PWSA (USA).

PW
POST OFFICE
For people with PWS



My name is Wayne John Cummings I am a none smoker age 38 born with Prader Willi syndrome and hearing difficulties I am looking for a teenage boy and girl penfriend who is deal hard of hearing or hearing impaired age 13/16 from the Merseyside and North Wales areas I love sign language golfing discos computers e mailing family and friends living worldwide writing letters to pen-pals haveing a lasting friendship with people going to the cinema travelling outside the UK such as New York which I went last April listening to rap music plus pop and rock n roll music and socialising as well I also like to have loads of friendships I like kind and honest people and sencible people as well I even love to teach sign language when I learn alot more about it. My address is:

5 Rawlins Street
Prescot Road Fairfield
Liverpool L70JE
Merseyside GREAT BRITAIN

Information for School Staff:

Supporting the Student Who Has Prader-Willi Syndrome

(Compiled by Barb Dorn, Outreach Program Director, PWSA of WI, Inc.)

All students with PWS are individuals. Each has varying strengths and needs.

This chart does **not** reflect the behavioral needs of all children and young adults.

Common Behavior Characteristics In Students With PWS	Possible Management Strategies
<p><u>Rigid Thought Process</u></p> <p>It is common for people with PWS to receive and store information in a very orderly manner. There is a strong need for routine, sameness, and consistency in the learning environment.</p>	<ul style="list-style-type: none"> • Foreshadow changes and allow for discussion. Do this in a safe area where they can share feelings. (The student needs time to adapt to this change.) • If there is a change -use visuals; put things in writing – lists, schedules. • If able, communicate changes in personnel ahead of time – but not too far ahead. • Don't make promises you can't keep. • Break down procedures into concise, orderly steps. • To resolve "stubborn issues" try using "compromise". Both the student and the educator have to come up with a totally new solution. Not only is this a successful problem-solving strategy – it can also be a form of diversion. • Provide praise when being flexible.
<p><u>Perseverative or Obsessive Thinking</u></p> <p>This is the tendency to get "caught" on one issue or thought to the point where it overshadows the main theme of the learning or social event. This behavior can contribute to difficulty in transitioning from one topic/activity to another. Students often have a great need to complete tasks. It can lead to loss of emotional control.</p>	<ul style="list-style-type: none"> • Use reflection – have student restate what you said. • Put in writing; use visuals. Carry a small notebook if needed. • Less is best – give less work at one time rather than more, add work as time allows. • Avoid power struggles and ultimatums. • Ignore (if possible). • Don't give more information than is necessary especially too far in advance. • Use "strategic timing" of activity the student has difficulty ending just before snack or lunch. • Set limits. "I'll tell you 2 more times, then we move on to next topic. This is #1."
<p><u>Tenuous Emotional Control</u></p> <p>Any combination of life stressors can lead to emotional "discontrol". The result may be exhibited as challenging behaviors such as tantrums – yelling, swearing, aggression, destruction, self-injury. During these episodes, reasoning is lost. Recovery of control takes time and is often followed by sadness, remorse, and guilt. Because of a problem in sequence processing, students are not always able to turn what not to do into what to do.</p>	<ul style="list-style-type: none"> • Be aware of "hallway over stimulation" – especially before the school day begins. Have student enter the building at a less popular entrance. If possible, have arrival time 5-10 minutes after school starts. Dismiss early. • Start the day by allowing time to go over the schedule for the day and work through any changes there may be. Putting a new schedule in writing often helps decrease anxiety. • At the start of the day – set daily goals with the student. Limit to no more than 3. Communicate behaviors you wish to see. Make it a cooperative task that provides concrete behavior expectations. Put goals in writing. Avoid the word "DON'T" ... focus on the word "WILL". (Ex: "I will talk in a quiet voice... instead of "Don't yell. When I feel frustrated, I will tell Mr. Smith or another adult.") • Provide positive attention and praise when student is maintaining control, especially in difficult situations. Celebrate success! • Encourage communication, acknowledging feelings. Words are important, listen carefully! • Include student in behavior plans. Their input elicits cooperation and a sense of support. • Be a role model. "I always say "darn" when I am angry. Let's try that for you... darn, darn, darn." Practice when the student is not agitated or angry. • Depending on the student and the situation – use humor. It is often effective. • Anticipate build up of frustrations and help him/her to remove self to "safe area." Create a key word or phrase to alert the student it's time to go. Practice using these words/phrases when the student is calm. • Develop a plan and teach the student what to do if he/she feels angry or frustrated. Many students substitute a means of releasing this pent-up anger: long walks/exercise, ripping paper, tearing rags, popping packaging bubbles, etc. • Don't try reasoning during times when out of control. Limit discussion. • Have a plan in place if student becomes more violent. Safety for all is a priority. Consistency in approach is imperative • Provide positive closure. Don't hold a grudge. • If using consequences – they should be immediate and help the student learn from the outburst – saying "I'm sorry," sending a note to say he/she is sorry.

Common Behavior Characteristics In Students With PWS	Possible Management Strategies
<p><u>Food Craving and Diet Restrictions</u></p> <p>For people with Prader-Willi syndrome, the message of fullness never reaches the brain – they are always hungry. In addition to this craving for food, food is metabolized at a rate that causes extraordinary weight gain. Food must be monitored and the individual supervised.</p>	<ul style="list-style-type: none"> • Make sure lunch is placed with a bus driver and /or an assistant on the ride to school. • Educate and inform all people working with this student – including bus drivers, custodians, secretaries and volunteers. • If the student states he/she has not had breakfast – call parents or caregiver before giving more food. (Oftentimes they say this to get more food.) • Supervise in lunchroom and in all food related areas – including vending machine areas. In some cases, student may need to eat in classroom (with peer/friend). • Many require supervision in hallways or near unlocked lockers at all times. • Avoid allowing the student to have money. Lock up all sources of money – including purses. Money buys food! • Address any stealing or trading of food in private. • Follow guidelines for treats or eating of extra food. Communication with home is very important. • Follow calorie-controlled diet. If a special calorie diet is needed and served by the school, a prescription must be obtained from a health care provider and should be a part of the student's educational plan. • Don't delay snack or lunch; if this is necessary discuss ahead. • Limit availability and visibility of food. Be aware of candy dishes or sources of food. • PRAISE situations where student does not take food when you see they could have. • Avoid using food as a reward or incentive. • Be aware of smells – there is nothing like the smell of popcorn to add make a student with PWS agitated. • When going on a field trip or other outing, discuss all food-related issues <u>ahead</u> of time. Will you bring snack along or will it be purchased? If purchased, what will it be? Will the outing interfere with the time of a meal or snack? • Obtain weekly weight by school nurse if indicated.
<p><u>Poor Stamina</u></p> <p>People with PWS tire more easily and may fall asleep during the day. Morning is typically their optimal learning time, when energy level is highest.</p>	<ul style="list-style-type: none"> • Daily exercise should be a part of student's schedule. • Get the person up and moving. Send on errand. Take a walk. • Schedule high energy, mobilizing activity after lunch. • Offer items /activities which stimulate large muscles and deep breathing: balloon blowing, party- blowers. • Provide scheduled rest time or a quieter activity if needed.
<p><u>Scratching and Skin Picking</u></p> <p>These two behaviors are often seen in individuals with PWS and may be worse during times of stress. Combined with a higher pain threshold, these behaviors can result in tissue damage if not controlled.</p>	<ul style="list-style-type: none"> • Use diversion - provide activities to keep hands busy (coloring, computer time, play dough, hand-held games...). • Keep nails short. Apply lotion liberally – it keeps skin slippery. Skin that is soft and moisturized is more difficult to pick. Applying lotion can also be an effective diversion. • Provide supervision. Reward and praise for not picking. • Cover area with band-aid or similar covering. • Don't just tell him/her to stop picking – it won't work. • Apply mosquito repellant before any walks or outside activity.
<p><u>Difficulty with Peer Interaction</u></p> <p>While children want and need other children and value friendship, it may be difficult for them to be exposed to the unpredictability of others for long periods. The need for order frequently translates into fairness issues and comparing themselves to others, often resulting in anger.</p>	<ul style="list-style-type: none"> • Many do better in small groups and at times alone • Pre-plan outings. Keep time short • "Supported recess or social outings" – planned activities with a friend • Include child in planning activities that are of interest to him/her (board games, puzzles, computer games...) • Provide social skill classes that emphasize sharing, taking turns...

Students with Prader-Willi syndrome are very caring, sensitive and conscientious. They want very much to be successful, have friends and be a part of their school community. Although they face some unique challenges, with proper support and understanding they are playing, learning, working and living successfully in our communities.

We Remember

Each of our young people with PWS had 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.

— Janalee Heinemann and Norma Rupe

A Gift from Harriet and Mathilde Budner

By Diane Spencer

The box arrived stuffed with medical reports, letters, books, county reports, lists of doctors to call, daily record keeping of behavior, cancelled checks, school recommendations, school rejections, doctor's reports, poems and tender apologies from Harriet to her mother, beginning "Dear Mom, I'm sorry..."

Harriet was born July 15, 1951 to Mathilde and Lou Budner in Bronx, New York. Mother was a teacher and dad was a dentist. This is the story of two parents trying to find out what was wrong with their only child, a little baby girl. The Budners started looking for answers when Harriet was 3 months old, but it was not until she was 30 years old that the diagnosis of Prader-Willi syndrome was accepted.

Harriet's parents were determined to provide her with a rich intellectual environment. They regularly attended Broadway shows, plays, etc., but Harriet was a "Dark Shadows" soap opera fan, and even attended a convention in Los Angeles by herself. Harriet did volunteer work at the library and helped with childcare at a community center. She was accepted as she was, but Harriet always said she just wanted to feel like "a real person."

One day when she was having a tantrum at age 4, her mother said, "Harriet, I've done everything I can think of, what do you want?"

Harriet answered, "A little of sitting-on-the-lap kind of loving."

From then until the day she died, after a tantrum she would ask for "a little sitting-on-the-lap kind of loving."

Another time, Harriet stole \$20 from her mother to make her cat Angelique well again. Then she wrote to her mother, "She is the only creature to help me when I need her the most. She may just purr and sit by me... but she is a wonder. I know I do some bad stuff like stealing and lying, but she helps me from going crazy and talking to myself. I need a non-human someone... No I don't need a psychiatrist, I understand and know myself. With this note you will find \$20, the amount I stole..." The note was signed, "Peace, Harriet."

Harriet once wrote to the *Examiner* after seeing a picture of herself under the headline, "They Eat Themselves to Death." She told the newspaper she was going to sue because: "1. I have a right to say when and where I have my photo taken; 2. I asked the photographer not to take my picture, he took it anyway; 3. Your paper is known to be bird cage stuff (slandorous and trashy); 4. I think you have a hell of a nerve printing stuff which is full of lies."

Harriet wrote in one of her "I'm Sorry" notes that one of the hardest things was knowing what to be. "I want sometimes

to grow up and be away from you, other times I want to hide like a young child and act like a child."

I think she was allowed to be both, happily for Harriet.

I recently talked with Harriet's mother and she told me that as she was going through Harriet's things, she was amazed at the many objects Harriet collected. As I read through this voluminous diary it really doesn't surprise me at all. I don't think the apple fell far from the tree.

In Memory of Harriet C. Budner 7/15/51–8/23/87, we gratefully acknowledge this meticulously recorded history given to us by Harriet's mother, Mrs. Mathilde Budner. The material sent to us will be carefully copied, to be used as a case study for university medical classes and our own group of doctors.

Diane Spencer is support coordinator in the PWSA (USA) national office.



Harriet Budner

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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.

