



# The *Gathered View*

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

## Conference News ...

- Jim Koerber (California) and Daphne Mosely (Oklahoma), both of whom have children with PWS, are newly elected to the PWSA board of directors; incumbents Don Goranson and Barbara Whitman also were elected for three-year terms. Retiring board members Louise Greenswag and Penny Townsend were honored for their past service.
- Board Member Paul Alterman received this year's special recognition award for his accomplishments as chair of PWSA's fund-raising committee. In recent years, Paul's work has significantly boosted PWSA's income from the Angel Fund drive and conference merchandise sales.
- The conference torch was passed by Roda Guenther (Florida chapter president) to Pat Shiley, who, with Jim Boyle, will co-chair the 1998 PWSA conference in Ohio.

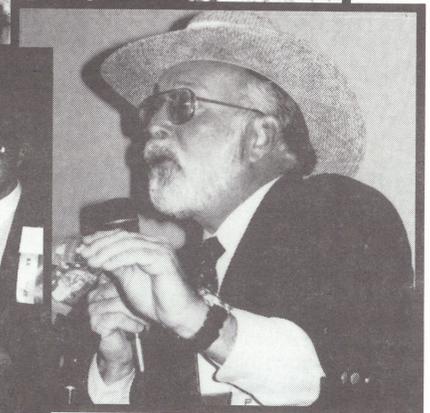
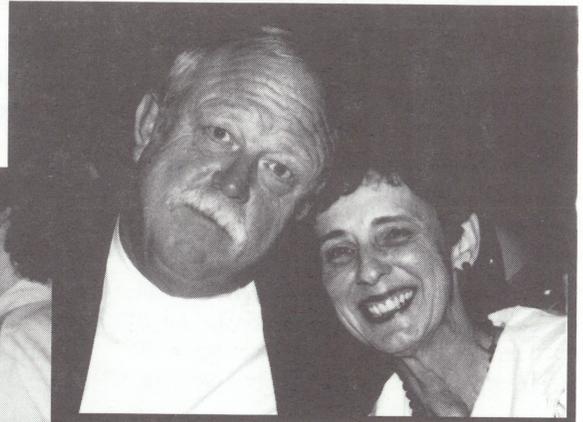
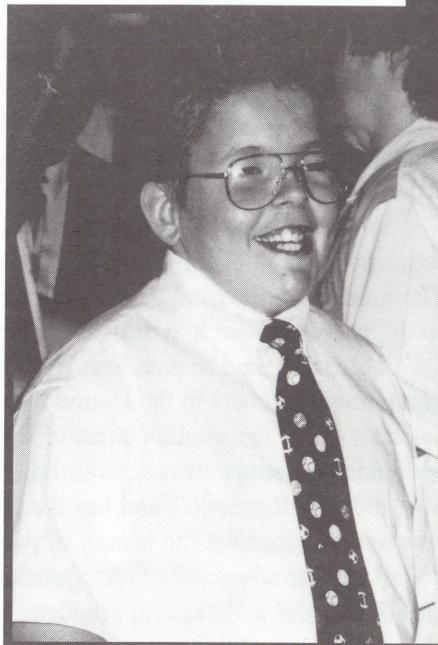
### Mark Your Calendars Now—

#### 20th Annual PWSA (USA) Conference

July 23–25, 1998

Adam's Mark Hotel  
Columbus, Ohio

*Thanks, Florida,  
for a Fantastic  
Conference!*



Photos (clockwise from top left): Josh Stallings, son of Youth Program Chairs Debbie and John Stallings, personifies the conference mood; this year's honoree, Paul Alterman, poses with fellow board member Barb Whitman; PWSA Treasurer Jim Gardner auctions donated Disney World tickets; Pat Shiley, left, accepts the conference torch from this year's co-chair, Roda Guenther; Roda, left, and Al Heinemann, right, flank KIDS by the Sea staff Connie Miles, Janalee Tomaseski-Heinemann, and Jim Miles.

*Thanks* to all our members who took the time to complete the PWSA membership survey. Look for results in coming issues of *The Gathered View*. Although the survey deadline has passed, we welcome feedback from our members at any time.

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## Out of the Office

by Russ Myler, Executive Director



**T**wenty years ago a small number of people gathered together to start this organization. At that time little was known about the syndrome, and PWS research was in its infancy. Prader-Willi syndrome was unknown to doctors and the world. If there was a diagnosis of the syndrome, it was a fluke, and no one knew what to do about it.

Clearly our founders were facing a monumental task. But probably the most challenging situation facing them was the need for research.

In 1983, the Association established the Research Fund—a rather bold act for a small (and underfunded) association. There were no great “fund drives” or consistent appeals to members. No one offered prizes or tokens for a donation. The Research Fund just “puttered” along. It soon became “the little fund that could.”

In the 14 years of its existence, the PWSA (USA) Research Fund has supported, in full or part, research into almost every aspect of the syndrome. The fund led to the establishment of clear diagnostic criteria, a classification study, cancer research, a drug study, growth hormone research, and more.

This little fund to date has “puttered” out nearly \$100,000 in direct grants to researchers in the United States and Canada. In addition, the fund helped support the establishment of IPWSO (the international organization) and widen the scope of understanding to the world. Now totaling some \$60,000, our Research Fund has the potential to launch or support many new undertakings for the benefit of people with PWS.

Also, the advocacy of our association through Leonard Hacker, a grandfather (of a “beautiful grandson” with PWS) who lives in Washington, D.C., has resulted in increased federal allocations to the National Institutes of Health specifically for Prader-Willi syndrome research in the current budget and in past years. Not too shabby for a little association ... And we are just getting started!

**Note to Researchers:** The deadlines for grant requests to PWSA-USA's Research Fund are May 1 and November 15. Contact the PWSA national office for application information (1-800-926-4797).

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Opinions expressed in *The Gathered View* are those of the authors or editors and do not necessarily reflect the views of the officers and board of directors of PWSA (USA). *The Gathered View* welcomes articles, letters, personal stories and photographs, and news of interest to those concerned with Prader-Willi syndrome.

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## What Are They Thinking?

We who are safe in our "normal" world wake up each day, wrapped in the security of knowing that our intelligence and experiences will guide us as we approach whatever challenges or trials lie ahead.

But what would the experience of waking every morning be like if you had Prader-Willi syndrome? What if your first thoughts and interactions with others determined how you would function throughout that day? What if the reasoning skills to have consistent self-determination were not part of your thought patterns so it would be necessary to have a schedule on the bulletin board or Mom to take you by the hand to help organize your day? What if, at the dawning of your consciousness, the first glimmer of thought was about food and that it interrupted all else and robbed you of your creative initiative?

In the early years of a child's life, parents and caregivers naturally have more control over the day's schedule and the child's mood and behavior. But, as the person with PWS matures and becomes more independent, those waking thoughts or an early morning battle over the day's plans can be the springboard to a very difficult day.

Regardless of the age or stage of development of our person with PWS, it is the right time for us to become active and to learn all we can about the service systems that will be working with our son or daughter during those difficult days. It is a time-consuming challenge to achieve a working knowledge of city, state, and federal services, school districts, work and residential facilities that are trying to meet the needs of this disability. But it is important to take up that challenge and become partners with the system. The school-age children, the group home residents, and the working-age people with PWS depend on our continued involvement.

When the greater community begins to take on more responsibility for our family members with Prader-Willi syndrome, it becomes our job as their family to convey clearly how the day begins for a person with PWS. Without our educational efforts and advocacy, facilities serving our children rarely reach beyond simply providing services to achieve a real understanding of "what they are thinking" when they behave in certain ways. Without this understanding, services will always fall short of what our son or daughter really needs.

## Money Matters

- Jim Gardner, PWSA (USA) treasurer, reported at the annual membership meeting that the organization ended up with a surplus in 1996, after several years of net deficits. This, he said, was the result of extreme diligence in the office as well as good sales of the new edition of *Management of Prader-Willi Syndrome* and the great generosity of our members. The need for contributions continues, he stresses, since the surplus funds have already been committed to producing new publications and videos.
- At its annual meeting the PWSA (USA) board of directors approved the purchase of \$8,500 in computer equipment and upgrades for the national office.
- The board also approved expenses for a PWSA exhibit at the American Society of Human Genetics conference in October (\$1,000) and for a Scientific Advisory Board meeting in Chicago to develop "standards of care" for people with PWS (\$750).
- A research grant (\$2,460) was approved for Anastasia Dimitropoulos (Vanderbilt University) to continue her study of behavior in young children with PWS, ages 2-6. (See page 5, second column, for preliminary research findings.)

Below are condensed financial statements from the past year. (Full reports are available from the national office.)

### PWSA (USA) CONDENSED BALANCE SHEET AS OF 12/31/96

<u>ASSETS</u>	
Cash	\$186,759
Investments	103,781
Inventory	24,344
Other	2,723
<b>TOTAL ASSETS</b>	<b>317,607</b>
<u>LIABILITIES</u>	
Payroll Taxes	\$ 1,187
<b>TOTAL LIABILITIES</b>	<b>1,187</b>
<u>FUND BALANCES</u>	
General Fund	\$254,157
Research Fund	62,263
<b>TOTAL FUND BALANCE</b>	<b>\$316,420</b>

### PWSA (USA) CONDENSED INCOME STATEMENT FOR THE YEAR ENDED 12/31/96

<u>INCOME</u>	
Earned Income	\$ 98,585
Public Support	147,715
<b>TOTAL INCOME</b>	<b>246,300</b>
<u>EXPENSE</u>	
Program Services	\$187,438
Management & General	17,406
Fundraising	6,235
Research	2,900
<b>TOTAL EXPENSE</b>	<b>213,979</b>
<b>SURPLUS (DEFICIT)</b>	<b>\$ 32,321</b>

# 1997 Scientific Day Summary

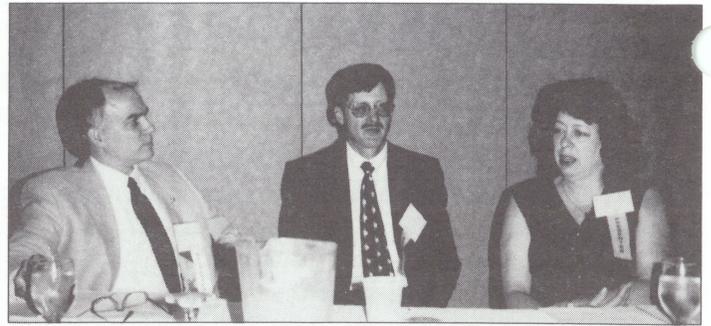
PWSA's 12th Annual Scientific Day was organized by Dan Driscoll, M.D., Ph.D., associate professor of pediatrics, molecular genetics, and microbiology at the University of Florida, Gainesville. Dr. Driscoll and Dr. Suzanne Cassidy summarized the day's reports for general conference participants. Dr. Cassidy reported that this year's scientific meeting was "one of the very best," with a good balance of presentations on genetics and behavior—current "hot topics" in PWS research—as well as a number of interesting medical reports.

## Genetics

- Camilynn Brannan (University of Florida) has succeeded in creating a mouse with Prader-Willi syndrome (PWS) by knocking out the PWS imprinting center in the corresponding chromosome of baby mice. As would be expected, the PWS mice have failure-to-thrive and die within the first seven days. Once researchers find a way to keep the tiny mice alive, they'll provide "an ideal experimental model" for study of PWS, Dr. Cassidy notes.
- Robert Nicholls (Case Western Reserve) and colleagues examined cases of PWS attributable to imprinting mutations and were able to narrow the identified "imprinting center" region of chromosome 15 from 3 million "base pairs" to 3,000. (Base pairs are the building blocks of DNA that make up genes.) Dr. Driscoll explained that there are about 50-100 genes in the Prader-Willi region, only several of which are subjected to imprinting. The imprinting center is the portion of the chromosome that controls whether imprinted genes throughout the PWS region are expressed. If the imprinting center is knocked out because of a mutation, genes that require imprinting will not function appropriately.
- University of Florida researchers also are trying to dissect the region of PWS on chromosome 15, Dr. Driscoll reported. On the premise that a person must have two or more paternally expressed genes lost in order to have classic PWS, the researchers are examining 41 patients who have many, but not all, of the clinical characteristics of PWS and

who do not test positive thru DNA methylation. They hypothesize that some of the "PWS-like" patients may have a flaw in just one of the suspected PWS genes. One of these genes, called SNRPN, is known to not be expressed in PWS patients, but was found to be expressed in all the PWS-like subjects. Examination of other PWS candidate genes is ongoing.

- David Ledbetter (University of Chicago) and colleagues are currently updating the genetic map of the Prader-Willi region, ordering the genes and looking for those which might have a role in the syndrome's characteristics.
- Drs. Ledbetter and Nicholls have adapted the DNA methylation test (first described in 1991 by Drs. Driscoll and Nicholls) to the PCR (polymerase chain reaction) technique, making possible faster and less expensive diagnosis of PWS and with smaller blood samples. A cautionary note, however, is that PCR is



Drs. Dan Driscoll, Merlin Butler, and Suzanne Cassidy described research findings at the PWSA conference.

"very susceptible to contamination," and care must be taken in sample handling.

- Dr. Cassidy looked at a group of patients who had been diagnosed with genetic testing (rather than by clinical observation alone) and found that there is "a significant amount of variability" in each of the classic PWS features, much more than previously defined. Given that the spectrum is "pretty wide," Dr. Cassidy recommends that "Clinicians should have broad expectations, a low threshold of suspicion, and a liberal use of genetic testing whenever they see people who have even a few of the features of PWS."

## Any Breakthroughs This Year?

*(Notes on the nature of research)*

Dr. Suzanne Cassidy, chair of PWSA's Scientific Advisory Board, told conference attendees that the question above, which she often is asked, reflects a common misconception about the nature of research. She stressed that "research happens in very tiny little steps, one on top of the other," and compared it to building a wall. The scientific "breakthroughs" that we hear about in the news are not sudden events but the culmination of many long hours of work—the announcement that the "wall" has been completed. While there were no "breakthroughs" in PWS research this year and none last year, she reported, a number of small steps have been achieved by researchers, promising potential breakthroughs in the near future.

PWSA's annual Scientific Day was begun 12 years ago to provide an opportunity for scientists studying PWS to meet in a less formal way to discuss their recent findings and to bounce ideas off each other, Dr. Cassidy noted. "As important as this meeting is," she explained, "it is still not the major forum for discussing research on PWS," the major forums being the meetings of leading professional associations in the specific fields of study—genetics, behavior science, etc.

Nevertheless, PWSA is proud of its 12-year history of providing this annual opportunity for PWS researchers to learn from and encourage each other and to meet with some of the many families who stand to benefit from their work.

## Medical

• Dr. Jeanne Hanchett (The Rehabilitation Institute of Pittsburgh) reviewed the records of 116 consecutive patients who had come thru TRI in 1991-96 and found that 36 had a problem with rectal picking and bleeding. There was an equal number of males and females, and 40 percent were under age 20. Signs of the behavior were: spending a long time in the bathroom; unexplained frequent, loose stools; mucus or blood in the stool; and blood on clothing, sheets, or finger. Denial of the behavior was typical. Three of the 36 had medical complications from the behavior that required treatment. Dr. Hanchett recommended the following interventions for rectal picking: 1) limit time in the bathroom; 2) accompany to the bathroom, or leave the door open; and 3) if behavior is severe, provide an object to occupy the hands, especially while in bed, if that's when it occurs.

• Dr. Merlin Butler (Vanderbilt) searched for a possible role for the "obese" (OB) gene in regulating appetite in PWS, even though it is located on chromosome 7. He found no correlation of body mass index (BMI) in the PWS group to a genetic variance found in the obese control group; consequently, the OB gene can be ruled out as being significantly involved in the PWS appetite problem.

• Linda Myers (The Rehabilitation Institute) discussed the impairment in daily activities and respiratory function caused by a lack of strength and use of the upper body in persons with PWS. TRI has developed a plan of specific exercises to increase strength and range of motion in the shoulder area, improve motor planning, and boost overall endurance, using wrist weights and other aids. (This exercise program was presented as part of the parent conference as well.)

• Helen McCune (University of Florida) and Dr. Driscoll reviewed bone density scans of 13 group home residents with PWS, ages 17-35, and found three with severe osteoporosis and five with a significant decrease in bone density. Contributing factors appeared to be sedentary lifestyle, hypogonadism, low calcium intake, and low growth hormone. Patients

with severe osteoporosis were started on Fosamax. To reduce the risk of osteoporosis and fractures, the investigators recommend 30 minutes of daily exercise and a calcium-rich diet—1,500-2,000 mg. of calcium daily (with up to 1,000 mg. of that total from supplements).

## Behavior and Cognition

• Kate Sullivan (Eunice Kennedy Shriver Center) tested 12 children in their homes on tasks related to social cognitive abilities. Compared with individuals with Williams syndrome and others with nonspecific mental retardation, the subjects with PWS performed relatively well on some tests but had trouble distinguishing pictures of people who were sad from those who were angry. They also gave poor explanations of pictures showing actions (interpreting people's be-

group. Common behavioral characteristics found among the subjects with PWS were: obsessive/compulsive, disruptive, and socially offensive behaviors; hoarding; skinpicking; and hurtfulness to self but not others. GABA, a neurotransmitter involved in impulse control and anxiety, was found in higher levels in the blood of African American subjects with PWS, but a larger sample size is needed to confirm these results.



Right: Dr. Moris Angulo and PWSA board member Gail Overton-Thune. Above: Dr. Cassidy with retiring board member Dr. Louise Greenswag.

havior), and they had trouble understanding intention. This is an ongoing study that will be important in understanding why people with PWS have social problems and how they can be helped in this area.

• Elizabeth Roof (Vanderbilt) reported preliminary data from the ongoing Vanderbilt University studies, showing lower verbal IQ in PWS subjects with deletion, impaired stereoscopic vision, especially in the group with maternal disomy; and poorer motor skills, compared with obese subjects without PWS in the control

• Anastasia Dimitropoulos (Vanderbilt) studied young children, ages 2-6, comparing parental reports of temper tantrums and behavior in 86 children with PWS and 50 typical children in a control group. The surveys revealed more compulsive characteristics in the PWS group and an increase in compulsive behavior with age, while the control group showed a decrease. Age of onset of the first tantrum correlated with age of onset of increased appetite. Children with PWS under age 2 had fewer tantrums than their peers in the control group. In general, the subjects with PWS were found to be less adaptable, more rigid, and to have more problems with change in routine.

• Elisabeth Dykens (UCLA) compared maladaptive behavior among people in three different disability groups—PWS, Down syndrome, and nonspecific mental retardation—and found certain characteristics to be distinctively associated with PWS. These include skinpicking, obsessions, underactivity, increased eating, stealing, and talking too much.

*(Summary by Linda Keder, with review and editing by Dr. Dan Driscoll)*

## Chapter News

PWSA of New England held an annual family picnic in June and their second annual pool party on August 3; this was after a busy spring hosting the 5th Annual Joint Conference with the New York Alliance chapter ... PWSA Missouri donated \$500 to the national association following the O'Leary's First Annual Charity Golf Tournament, hosted by O'Leary's Restaurant and featuring actor John Goodman. (The restaurant is co-owned by Kevin O'Leary, uncle of Timmy O'Leary, who has PWS.) The chapter split proceeds of \$2,733 with a local autism organization ... Summer swim parties are popular—the Missouri folks also planned a splash on August 3 ... PWSA of Wisconsin held a "Breakfast with Your Legislators" and a PWS Training Day for providers in March and a Spring Dance in April; they've also been working to get state legislation passed to mandate services for people with PWS ... Our new Oklahoma chapter opened two group homes last fall and held a day-long conference in May ... Five chapters—Arizona, Florida, New England, Ohio, and Wisconsin—and the California Foundation now have their own Web pages on the Internet; California's includes information in Spanish ... California is also opening a 15th PWS group home and developing a video on PWS ... Georgia and Arizona held their third annual walking events for PWS Awareness Week, and Connecticut held a swim-a-thon/walk-a-thon. Utah held a meeting at the zoo to get younger families out and raised funds through a benefit musical concert ... Kentucky has held a chapter picnic, a dinner dance, and a silent auction, and is trying to open a second group home.

## Medical Alert on Antidiuretic Hormone

Antidiuretic hormone is a substance normally secreted by the pituitary gland; its function is to prevent elimination of liquid from the body. Lack of this hormone can be one cause of nighttime enuresis (bedwetting), which is a common occurrence in Prader-Willi syndrome, and medication that boosts the hormone level can help resolve this problem. An excessive amount of antidiuretic hormone, on the other hand, can stop all urination, diluting the body's electrolytes with retained fluids and setting off nervous system reactions such as seizure or coma. Several recent parent reports highlight the importance of careful dosage of medications that affect this hormone and monitoring of fluid intake and possible side-effects in conjunction with their use.

### DDAVP

DDAVP (desmopressin acetate) Nasal Spray is a synthetic version of antidiuretic hormone, sold by prescription to treat bedwetting in children age 6 and older. Although it has been successfully used in some children with PWS, Diana Baker from Virginia asked us to alert members that her teenage daughter with the syndrome ended up in a coma-like state after three consecutive nightly doses of DDAVP. (The drug is typically used nightly for four to six weeks in a controlled-dose spray, and side-effects in the general population are usually nonexistent or mild.) When the prescribing doctor checked with Dr. Jeanne Hanchett, who had recently seen the young woman at the Rehabilitation Institute of Pittsburgh, it was discovered that Dr. Hanchett had seen a similar reaction in another patient with PWS and that the drug manufacturer had a third case on record. It's likely, Dr. Hanchett says, that at least some people with PWS may have a lower tolerance for DDAVP and require either an adjusted dosage or a different solution to the enuresis problem. Diana's daughter, by the way, recovered slowly but completely once the drug was stopped.

(Note: DDAVP is manufactured in Sweden for Rhône-Poulenc Rorer Pharmaceuticals Inc., 500 Arcola Rd., Collegeville, PA 19426.)

### Haldol

In the recent issue of *Wavelength*, the international PWS parent newsletter, a father wrote about his 26-year-old son, who had suffered convulsions and a coma after several months' treatment with Haldol (haloperidol) for psychiatric symptoms. On further investigation, it was learned that the young man had been for some time drinking more water than usual, though not an amount that would normally cause problems. Professor Martin Ritzén of the Pediatric Endocrinology Unit, Karolinska Hospital, Stockholm, Sweden, commented, "It is known that in rare instances [haloperidol] can cause an oversecretion of antidiuretic hormone which prevents the kidney [from] getting rid of excess water. For a person with PWS who might decide to drink in order to reduce eating this might cause water intoxication as in [this] case." He speculates that people with PWS "may have a slightly deranged regulation of the antidiuretic hormone," noting the abnormalities in many other functions regulated by the hypothalamus—appetite control, temperature, pain threshold setting, and secretion of growth hormone and gonadotropins. "In any case, PWS patients on haloperidol should be supervised as to their water consumption," Ritzén concludes.

—Linda Keder

### MEDLINE Medical Information Searches Now Free to All

On June 26, Vice President Gore announced that the National Library of Medicine, part of the National Institutes of Health, will provide free access to MEDLINE through two products on the World Wide Web—Internet Grateful Med and PubMed. MEDLINE is the world's most extensive collection of published medical information, a database of more than 8.8 million references to articles published in 3,800 biomedical journals. Grateful Med and PubMed have different features for searching and for obtaining full text of articles. PubMed has sets of related articles pre-computed for each MEDLINE article; it also includes journal cites outside the MEDLINE database. For more information, go to the following Internet address:

<http://www.nlm.nih.gov/databases/freemedl.html>

# International PWS News

## Not Spain, but Italy, May 21–24

### *New Site and Date for 1998 International PWS Conference*

The International Prader-Willi Syndrome Organisation (IPWSO) announces that the Third International Prader-Willi Syndrome Scientific Workshop and Conference, planned for Seville, Spain, is being moved to Lido di Jesolo, Italy (35 km from Venice), and will now be hosted by the Italian PWS Association. The conference dates are just one week later than originally scheduled.

All interested persons involved with PWS—researchers, parents and caregivers, medical and other service providers—are encouraged to attend. In the June IPWSO newsletter, Board Chair Jean Phillips-Martinsson (UK) notes that many countries have succeeded in raising funds to send members to the international conferences. She suggests exploring the following as possible funding sources: “social services, local industries and authorities, handicap organizations, the media, pharmaceutical and diet food companies, airlines, tourist companies, etc.”

Conference registration is Lit. 500,000 (about \$290 US). Other costs are estimated to be \$35-50 for lodging, depending on accommodations, and \$45 for the gala dinner. Registration materials are not yet available, but additional information about the international conference may be obtained from PWSA’s parent representative and IPWSO Vice President, Mildred Lacy, Louisville, Kentucky, 502-968-2626.

## IPWSO Logo Contest

### *Win a trip to an international PWS conference!*

The International Prader-Willi Syndrome Organisation is in search of a logo that can be used on its letterheads, pamphlets, newsletters, etc., and has announced a competition to develop one by the time of the international conference in Italy.

The contest is open to all people with a “PWS connection,” and there’s no limit to the number of entries. The prize: reimbursement of conference fees for any one international PWS conference over the next six years.

The winning logo “will visually describe” IPWSO and “be easily recognized all over the world.” It will be selected by Dr. Andrea Prader and IPWSO Board Members Jean Phillips-Martinsson (UK) and Linda Thornton (NZ).

Entries must be on 8½" x 11" paper and received by the closing date of **March 31, 1998**. Mail to: *IPWSO Logo Competition, P.O. Box 143, Masterton, New Zealand.*

## Wavelength & Scientific Newsletter

### *Combined IPWSO publication now available to all*

Since its formation in 1991, IPWSO has kept in touch with its member PWS associations throughout the world via two newsletters: *Wavelength*, the newsletter for parent representatives to IPWSO (edited by Linda Thornton of New Zealand), and a scientific newsletter on PWS for the scientific representatives (edited by Dr. Ellie Smith of Australia). As of June 1997, the two publications are combined and are available to all interested persons both on the Internet and in paper form from PWSA (USA). The latest issue includes medical commentary on metabolic rates, water intoxication, and food compulsion in PWS, as well as some personal stories, a book review, and more.

A photocopy of the newsletter (28 pages, 8½" x 11") can be obtained by mailing a request for the June 1997 IPWSO newsletter, along with \$1.00 (in US cash, check, or money order) to cover copying and mailing, to: PWSA (USA), 2510 S. Brentwood Blvd., Suite 220, St. Louis, MO 63144-2326.

The Internet version of IPWSO’s *Wavelength & Scientific Newsletter* can be found on IPWSO’s new home page, hosted by Visible Ink Incorporated. IPWSO’s Web address is: <http://www.pwsyndrome.com/IPWSO/index.html>



## Life-Changing Questions

The Reverend Don Roberts, Methodist minister, CEO of Goodwill Industries-Manasota, and the dynamic keynote speaker for the 1997 PWSA conference, urged members of the audience to ask themselves the following seven questions that “can change your life.”

### **1. Are you mistaking the edges of your rut for the horizon?**

*Ask yourself this question every day, he advises.*

### **2. Do you know you have a choice?**

*Many people with disabilities and their families feel victimized, when in fact we all have choices.*

### **3. Do you realize that no good deed goes unpunished?**

*Something always gets in the way; accept that fact and continue to do good anyway.*

### **4. Are you controlled by your past—or by your future?**

*That “kernel of darkness called guilt” keeps us self-centered, locked in the past; forgiveness is needed to move forward and to give to others.*

### **5. Are you capable of being nice to yourself?**

*You do deserve a break today!*

### **6. Are you willing to stop the battle?**

*Some battles with our children are fruitless. We should accept that they have a legitimate point of view and worry about our own behavior instead—after all, we are their models for adulthood.*

### **7. Have you told somebody that you love them today?**

*This conscious daily action can change both lives.*

Your perspective on life is a determining factor in the quality of your life, and it *can* be changed, Roberts concludes.

## Ask the Parents

### How to Curb Hoarding?

My daughter is 34 and lives in a home with three boys with PWS. She has her room at the front of the home, while the boys' rooms are on the other side of the living room down the hallway, so it works quite well. I find Kim is constantly changing as she ages, so things are never really calm for long. But one thing that concerns me is that her compulsion to gather, collect and save everything seems stronger all the time—she is more protective of letting anything go. It's like she cannot save enough, whether it is pop-can tops, mugs, towels, crossword puzzles—anything and everything. We used to be able to deal with it in different ways to reduce the amount she collects, but as time has gone on, this has become more difficult. Now there is hardly space in her room—it's stacked all over and even in the middle of her floor, —in fairly neat piles, but so much of it!

She does fairly well (considering the syndrome characteristics) as long as her stuff is left alone. Any attempt to reduce it causes terrible behaviors and moods that have become difficult to deal with. How can we get her to reduce the amount of stuff without causing a serious setback in behavior? We tried major clean-outs when she moved, boxing stuff up for storage, rewards, persuasion, donating to the poor and needy, and even a major overhaul throw-out day while expecting short-term behavior tantrums. Nothing seems to work—it seems to cause long-term behavior problems.

Do any of the other parents or group home situations relate to this and, if so, how do they deal with it? I have asked the group home staff to address this issue, to make it a priority, but I would like to be able to offer them some suggestions rather than just complain.

Any information or suggestion is worth considering—HELP!

Thanks,  
Judy Ipsen  
Willard, Utah

Please send your suggestions for Judy in care of the national PWSA office so that we can share them with members in a future issue of *The Gathered View*—PWSA (USA), 2510 S. Brentwood Blvd., Suite 220, St. Louis, MO 63144-2326, Tel.: 1-800-926-4797, Fax: 314-962-7869, E-mail: pwsausa@aol.com

## Resources

### Custom-Made Clothing

If finding clothes that fit a child or adult with PWS seems an impossible dream, it may be worthwhile to consider custom-made clothing—after all, appearance and comfort can have a great impact on self-esteem!

Mary Misner of Walworth, Wisconsin, wrote to PWSA to announce that she is opening a custom clothing business, The Magic Balloon, and that she would like to “become another friend” of kids with Prader-Willi syndrome.

Having worked with disabled adults on a college campus, Mary says she is well aware of the problems of not being able to find clothing that fits, is contemporary, and is reasonably priced. She has sewn for many years and enjoys the experience of creating something special, especially for those who have disabilities. She is able and willing to make dresses, suits, play and dress outfits, sleepwear, coats and hats, slacks, and blouses. She is especially interested in producing clothing for girls, and she can make matching outfits for a girl's doll, if she has the doll's size.

For more information, contact: Mary B. Misner, The Magic Balloon, N1639 Six Corners Road, Walworth, Wisconsin 53184. Tel.: 1-414-275-2430.

### PWSA's Support Group on the Internet

Have you visited PWSA's World Wide Web site lately? We now have a “chat room,” where people can meet and exchange messages in real time with anyone else who happens to be there. (Evenings are best—try Wednesday or Friday.) In addition to the chat room, it's now easier to post messages on our Parent Connections page. Check it all out at: [http://www.athenet.net/pswa\\_usa/](http://www.athenet.net/pswa_usa/)

There's a growing gold mine of help and support available out on the Internet for parents of children with disabilities. In addition to PWSA's Web page, you might find other parents with similar concerns at the following sites:

The Prader-Willi Foundation  
<http://www.prader-will.org/>

Family Village  
<http://www.familyvillage.wisc.edu/>

Exceptional Parent  
[http://families.com/sr.asp?ch\\_no=20](http://families.com/sr.asp?ch_no=20)

Our Kids  
[http://rdz.stjohns.edu/library/support/our\\_kids](http://rdz.stjohns.edu/library/support/our_kids)

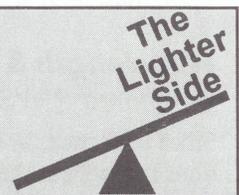
Sibling Support Project  
<http://www.chmc.org/departmt/sibsupp/>

The Arc of the United States  
<http://TheArc.org/welcome.html>

United Cerebral Palsy Associations  
<http://www.ucpa.org/html>

*Did you hear  
the one about  
the Sheraton  
desk clerk  
in Orlando who, conversing  
with a young man with PWS  
about the movie “Free Willy”  
(in anticipation of the trip to  
Sea World), then heard him  
mention “Prader-Willi” and  
figured that must be a sequel  
to the killer whale story?*

*(Thanks to Jean Shapiro from Minnesota for sharing this chuckle.)*





## 'What a Perfect Little Boy!'

*Tammy Davis, from Zwingle, Iowa, sent us this article about her son Eric, his early diagnosis with PWS, and her family's determination to "give him the best life possible."*

**O**ur son Eric is a very happy 5-year-old. ... our third child. (He has a brother, 10, and a sister, 9.) We, like many other parents, had no reason not to expect a normal, healthy infant. We knew from his first breath that there was something very wrong.

We are very lucky to have a great pediatrician. He immediately contacted the University of Iowa genetics lab to see if they could piece something together. When Eric was one day old, they drew blood for a chromosome screening. At three days old, we took him home. He had no suck reflex but could swallow. This allowed us to feed him through a preemie nipple cut with a large hole. We fed him every two hours around the clock for many months. The feedings were small and could not take longer than 20-25 minutes to be sure he wasn't using more energy than the calories he took in. He did great and gained weight slowly but steadily!

When Eric was 12 days old, we attended an outreach genetics clinic at the same hospital where he was born. His chromosome screening was normal, but he clinically fit all of the features of PWS. We went home with a lot of information and many unanswered questions—and were completely devastated. Because Eric's chromosome screening came back normal, the doctors at the U. of Iowa felt it would be to our benefit to explore other possibilities. To this day we have been through EVERY pediatric unit the U. of Iowa has to offer with no other medical findings. All indications pointed to PWS. We knew it was up to us to find out how to make his life much better.

Eric has had physical therapy since he was six months old, and today he can ride a bike, run, hit a baseball, and swing on a swing. He may not be as fast or as adept as an age-appropriate child, but he has a great time. He has had speech therapy since the age of 2. That hasn't been as

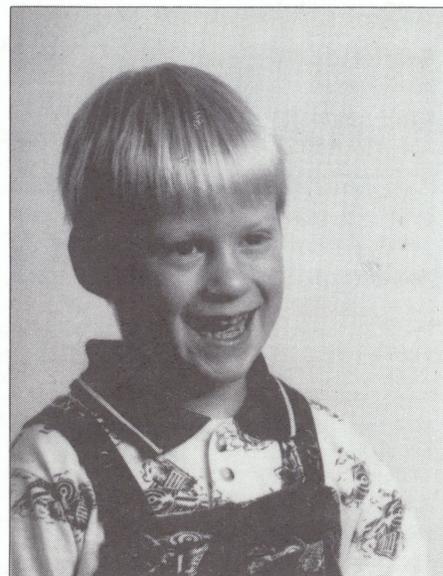
successful; he has the speech of a 2-year-old with a good pickup of sign language. Somehow he always manages to get his point across. Eric is a beautiful little boy. His smile lights up a room. He is funny, very bright and has a personality that is delightful. We don't have many bad days.

He is evaluated every six months by the U. of Iowa Hospital Schools Prader-Willi Clinic. It has been a great help to us. We see how much he has progressed and find new things that might be helpful in the future. We have made it a point not to read any information on PWS that is not current; we now know that every child with PWS is unique.

Eric has not shown any food-seeking tendencies yet. He has maintained his weight and has added about three inches in height in the last year. He doesn't have many temper problems that aren't age appropriate. He is very scheduled in his own mind, and change is sometimes difficult for him to understand. But we handle problems as they arise and try to stay one step ahead, so schedule changes don't happen too often without letting him know first.

**"I am very hopeful that early diagnosis will make the difference for Eric."**

Last December, Eric's diagnosis was finally confirmed through the methylation test for PWS. It was a relief to at last have a definite diagnosis. We have fought for the last three years for our insurance company to help pay for the testing. We won the fight, but they have now excluded his care for anything pertaining to the syndrome. They consider PWS to be simply a developmental delay. If you live with or care for a child with PWS, you know there is much more to this than developmental delay. We will continue to pursue the care issues with the insurance company because we feel strongly that this type of discrimination should not be allowed.



I don't know what it will be like in the future. But we cannot sit and wait for all of the PWS tendencies to happen. For our family, it is important that we allow Eric and our other children the opportunity to become their own person and to offer the guidance and strength needed to help them along the way. We realize that no matter what we would like them to achieve, they will do it their own way. We must set the rules and guides and watch them grow. Eric is a person first, who just happens to have PWS. I look only far enough into the future to be sure we are providing what he will need to become a productive member of our adult society. I am very hopeful that early diagnosis will make the difference for Eric.

Eric and I were paying for a purchase at a local department store, and he was being his usually charming self. A woman we did not know looked at Eric, then at me, smiled, and said, "What a perfect little boy." I smiled and said, "Thank you." He is perfect, he is Eric, and I could not imagine him any other way.

This verse was given to me by a friend, and I have it framed and hanging by my children's pictures. We must remember everyone is special and a gift.

*"I looked at my children around me,  
Each one in turn I embraced.  
Each one was a gift from their maker.  
God has such wonderful taste."*

(Editor's note: Tammy says that she would enjoy hearing from anyone who has a child close to Eric's age. Send her a letter via the PWSA office, or e-mail her at: ktcaedav@netins.net)

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