The Gathered View
National Newsletter of the Prader-Willi Syndrome Association (USA)
July - August 1993
Volume XVIII Number 4

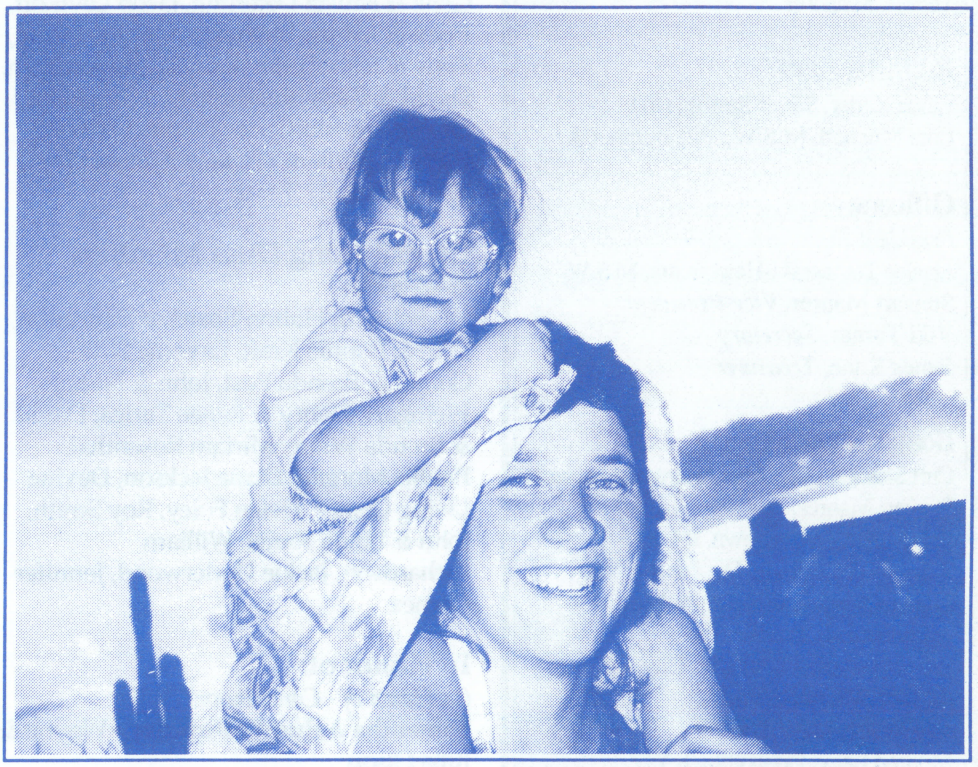
The View From Arizona

"Each conference leaves me more inspired and confident in living and dealing with the obstacles of the syndrome."
– A parent at the 1993 national PWSA (USA) conference

For both first-timers and many-timers who attended the PWSA national conference in Scottsdale last month, it was a memorable experience – seeing a hundred children with Prader-Willi syndrome in one place, talking to parents from across the nation and beyond, meeting and greeting old friends, making new ones, and hearing speakers address many different aspects of PWS. Each conference is different from the ones preceding it, yet the same in the sense of the community of understanding and support it offers parents and professionals for an intense several days each year. We return home feeling charged up, renewed, or at least less alone.

Nearly 300 parents and professionals made their way to the beautiful resort setting of the Arizona conference, including 37 group home providers who attended their preconference workshop and 27 researchers who participated in the Scientific Day. A show of hands at the opening conference session indicated that almost half of the attendees were first-timers.

In addition to 19 preschoolers and infants in child care, The Youth/Adult Activity Program served 101 children and adults with PWS, ranging in age from 6 to 35, and a number of their siblings. Highlights of the YAAP week were a visit to a wild animal preserve called Out of Africa, films at the IMAX theater, a sunrise breakfast, and, of course, the Friday night cookout and square dance. Friendships were renewed, and a few romances bloomed.



Renee Aalund gives daughter Samantha, 3, a lift on a warm Arizona evening.

Conference attendees had a rare opportunity to meet New Hampshire artist Stuart H. Williams, a 42-year-old man with PWS who raises goats and draws lively animal scenes using markers and oil pastels. Our thanks to *Prader-Willi Perspectives* for bringing Stuart and his colorful works to the conference and to everyone's attention.

This issue of *The Gathered View* is longer than most so that we could include information and photographs from the conference. We hope to report on more of the conference sessions in future issues, as well as bringing you stories from families and professionals we met in Arizona.

Special Conference Reports in this issue:

Also in this issue . . .

and much more.

Prader-Willi Syndrome Association (USA)
1821 University Ave W. #N356
St. Paul, MN 55104-2803
800-926-4797 612-641-1955
Fax 612-641-1952
Office Hours: 8:30 am - 4:30 pm CDLST

Special Thanks to Our Contributors

(Donations received from June 11 to July 29)

Angels ($50-$99):
Ray & Linda Sheeran

Patron Dues Members ($100+):
Rich & Barbara Carter, Peter & Lucia Smith, Norman & Cynthia Barkeley, Gene & Fausta Deterling, Jacob Levikoff, Rhett & Jeanne Eleazer, Jay & Sue Jacobi, Tony & Jean Antin, Amanda Kavanaugh, David & Carol Burleigh, Mercedes Lat, Herb & Gerald Gross, Reinard & Rose Nanzig, William & Diana Kirchhoff, McCall Family Trust

Contributing Dues Members ($40-$99):
J.Y. Belliard, Halton Board of Education, John & Bonnie Kraft, Laverne Sidlo, Gerald & Brenda Post, John & Nancy Weingart, Henry & Renee Tarica, David & Brenda White, Miriam Rosenthal, Phyllis Morgan, Diana Jackson, Devine Living Center, Helen Foley, Roy Smith, John & Linda Kania, William Richardson, Jackie Underwood, Jennifer Gomez

In Honor of:
(In Honor of - Contributor)
Samantha Aalund's Birthday - Wayne & Julie Olson

Memorials:
(In Memory of - Contributor)

Thomas Kujawski - Society Bank, Sylvania Chamber of Commerce, Pat & David Zaski, John & Cathy Goetz, Century Equipment, Albert & Linda Kennett, Duane & Bonnie Guhl, Robert & Arloa Wheeler, Charles & Nancy Johnson, Jackson & Evadine Bryan, Carolyn Nies, James & Elizabeth Boldt, Irrigation Association of Ohio

Adam Trachtenburg - Abe & Katherine Braitman, Hope Wolk, Jacklyn & James Gibson, Donald & Janice Bauer, Leonard & Rosalie Lewy, Samuel & Ann Pearlman, Anita Granoff

Dabebruzzo - PW Florida
Edward Cantor - Ida & Judy Singer
Fred Hockbery - Ida & Judy Singer
Eviline Gregers - Frances Van Zomeren
Lucille Thorsen - Frances Van Zomeren
George Moerke - Frances Van Zomeren
Olive Croal - Lota Mitchell
Robert Jones - Ed & Ruth St. John

Renee Davis - Gerald & Janet Wade, Mr. & Mrs. Carey Highsmith, Patricia McBryar, Jean Housley, Evelyn & Sandra Rogers, Ooltewah Middle School, Countryside Cafe

Harry Kricheff - Jackie Lovenworth, Richard Ruf, Rotelle Inc. Food Services Division, Rita Agrons, Martin & Elaine McNamee, Alrod & Sarah Barison, Robert & Rose Graham, Roselyn Cacciatore, International Multi Foods, Muriel Kazansky, Evelyn Pinerman, Hass Co. CPAs

Diane Chausow - Suzanne & Carter Meldman, Scott & Betsy Forester, John & Kathleen Grede, Henry & Patricia Moughamian, Jerry & Barb Siegan, Polly & Eugene Wachtenheim

Research:
Stu & Marnie Boyd, Women's Fellowship First Church of Spencer, Massachusetts, PW Kentucky, Sunlighters Group First Congregational Church, William & Linda Dorsey, Richard Basker Trust

Operating:
PW Kentucky

CIT:
Ed & Lois Olson (2)

Conference Grants:
Linda Keder, Doris & Nicolet Luhman, Bobby & D.J. Miller, Lamar & Barbara Paul, Curt & Marion Shacklett, Henry & Susanne Singer, Mary Shommer, Beatrice Sturm, Douglas Taylor, Marjorie Royle, Ruth Prettyman, Margaret & T. Clifford Fleming

Conference Grant for the Youth/Adult Activity Program in Phoenix, Arizona:
The Carpet Company

Contributions through the United Way:
Barbara Barney, Mr. & Mrs. James C. Hill, Mary Culver, Peat Marwick, Maurice & Jean Vermeulen

Movie Star Status

by Janalee Tomaseski-Heinemann

For the first time in my life I was compared to a movie star. No, it was not because of my eyecatching beauty or remarkable talent. It was because many years ago, our then 8-year-old daughter, Sarah, and I collaborated on a small unsophisticated booklet for PWS siblings called *Sometimes I'm Mad -- Sometimes I'm Glad*. An adult sibling came up to me at the recent national conference in Phoenix, Arizona, and exclaimed, "I feel like I'm meeting a movie star! I grew up on your book and reread it many times." How special it was to find out something I did, however insignificant it was to most of the world, meant so much to one young woman -- and perhaps others. If it hadn't been for the national conference where we exchange ideas and encourage each other, that book, along with other articles I have written, would have died on the shelves of a teacher's basement.

> *"How special it was to find out something I did, however insignificant it was to most of the world, meant so much to one young woman ..."*

This year's conference was at a beautiful resort, and the dinner/dance was such a unique and delightful experience (the setting was a western town out in the desert, complete with hayride, square dancing, and more) that next year's host committee said they wouldn't even attempt to top it. The best part was, as always, meeting old friends and new. The part I delight the most in is the touching and humorous "real life" stories I get from other families. Because space limits me here, I will just mention a few that come to mind: The young man with Prader-Willi syndrome who "borrowed" his mother's checkbook and bought a cow (for a FFA project). His justification to his mother was, "I told you I didn't like rabbits!" ... The two young men standing by the pool and arguing over whose girlfriend was whose. When the one young man's father came by, he was told, in no uncertain terms, not to interrupt because this was "girl talk." ... I met a mother who had four biological children and then adopted four children -- one with PWS, one with Angelman syndrome, one with Down syndrome, and one with a physical disability. If that doesn't humble you!!

(If anyone has any good stories from the conference, I would love to hear from you.)

Chapter Presidents' Day

The following is a report from our brainstorming session during our PWS Chapter Presidents' Day, held the day before the conference officially starts. I can't tell you how impressed I was with the group, their energy, and their wonderful collection of ideas. A more detailed report will go to the chapter presidents.

What National Can Do for State Chapters and Affiliates

1. Develop a press/media packet.
2. Develop a lobbying/legislative packet with dramatic stories.
3. Prepare a list of resources for non-English speaking families and professionals.
4. Do an article in *The Gathered View* or *Chapter President's Quarterly* on how to target medical meetings and set up displays at these meetings.
5. Have state chapter displays at the national conference to generate idea sharing.
6. Have a state chapter calendar of events in *The Gathered View*.
7. Develop a sibling packet.
8. Develop a new general brochure.
9. Share information on grant writing.
10. Don't try to screen new information -- just report it. Use qualifying statements if necessary.

Issues and Ideas for States

1. Have a "game plan" for recruitment of officers (i.e. parents of children of various ages).
2. Recruit physicians to a) become a specialist on the syndrome and have a clinic for your families, b) get on your board, and c) promote more research.
3. Give national an information sheet on services you offer, when and where meetings are, etc. -- to relay to inquiring families.
4. Have a special program for parents of young children with PWS.
5. Request meeting space and assistance with your youth group from a university or hospital. Pay $5 fee per family for someone to run the youth group.
6. Newsletters help bridge the gap in communication throughout the state. Remember to put adjoining state chapter presidents on your mailing list.
7. Have social gatherings for your families (i.e., dance, picnics, swim party).
8. Promote a professional in your group to do Grand Rounds in your local hospitals. Set up an information table at your state genetics conference or related conferences.

PWS Awareness Event

1. Should only be a week to get focused media attention.
2. It will be early October of 1994.
3. Chapters should target schools, local groups, or hospitals to help (i.e., book-a-thon, jump-a-thon).
4. Money between chapters and National should be split 50/50.
5. National will provide shirts, hats, cups, etc., with logos.
6. National will provide media packet and "how to" packet.
7. The focus should be positive and informative beyond the eating problems, i.e., "Ask Me About PWS".

Several exciting ideas for the 1994 Atlanta PWSA (USA) national conference were shared with Dottie Cooper, conference coordinator from the Georgia chapter. The Atlanta group is already incredibly organized. I am confident they will put together an outstanding conference. I hope to see you there. Be prepared to experience true "southern hospitality."

Board Elections

Elections were held for two openings on the PWSA (USA) board of directors at the national conference in Arizona. Four members in good standing were nominated from the floor at the general membership meeting. Those nominated were: Dr. Suzanne Cassidy, clinical geneticist; Sheldon Tarakan, publisher of *Prader-Willi Perspectives;* James Kane, PWSA (USA) treasurer; and Gail Overton, a dietitian from New Mexico. Dr. Cassidy was reelected for a fourth three-year term. James Kane was elected to the second seat. Subsequent to the conference, the procedures and results of this election were challenged by Mr. Tarakan, and they are now under review.

The Association extends its appreciation to Curt Shacklett, who is leaving the board, for his leadership and service, not to mention for sharing his delightful sense of humor from the podium at the national conference. As past board chairman, he will serve for one year on the board's advisory committee.

Appointments and Actions

As in past years, the board of directors met a number of times during conference week to conduct business. Decisions made by the board during the July 1993 meetings include:

Viki Turner, a parent from Pennsylvania, was appointed board secretary. Jim Kane was elected board chair for the coming year. Since the board chair cannot also be an officer, a new treasurer will be appointed. Stewart Maurer leaves the position of vice president, having served the maximum two terms. The board will reevaluate the role/responsibilities of this position prior to appointing a new vice president.

Annette Ruiz was appointed chair of the nominating committee. Board members Paul Wissmann and Pauline Parent will serve on that committee, along with other members to be appointed from the general membership.

The public relations special team, chaired by PWSA (USA) President Janalee Tomaseski-Heinemann, was made a permanent standing committee. On that committee's recommendation, PWSA (USA) will commission the design of a new Association logo as part of a publicity packet being developed to promote PWS Awareness Week in the fall of 1994.

PWSA (USA) will make an unconditional grant of $2,500 toward the 2nd International Conference on PWS, tentatively scheduled for 1994 in England. A second grant of equal amount will be presented when the conference takes place.

The publications committee, chaired by Dr. Barbara Whitman, was granted $17,500 to revise designated PWSA (USA) brochures and videos. Individual projects costing more than $2,000 will require specific board approval.

Coming Soon: PWS Holiday Cards

Members who send holiday cards are in for a treat this year. Beautiful cards created by individuals with Prader-Willi syndrome are being sent to a sampling of members as a test fund raiser.

In October, active members will receive an envelope containing 10 cards, 10 envelopes, a contribution form and return envelope. Members indicate their contribution amount on the form and return it to PWSA (USA) with their donation. Details will be included with the holiday cards.

With this project in mind, drawings were collected from YAAP participants at the 1992 conference in Philadelphia. Paul Alterman, chair of the fund raising committee, had the designs printed on 8 1/2 x 5 1/2 cards. The large colorful cards come complete with a description of PWS on the back of the card.

Billy Eleazer from South Carolina created this image of a holiday scene.

Scenes pictured include a Christmas tree and presents, a lone evergreen tree with a simple Merry Christmas greeting, a Santa Claus and sleigh, a Star of David with a Happy Hanukkah greeting, and the snowman pictured here. All of the cards open to the greeting, "Wishing you a wonderful holiday season and a blessed New Year!"

The Association would like to thank Paul Alterman and the entire fund raising committee for devising such a unique way to illustrate one of the many talents of individuals with Prader-Willi syndrome.

Scientific Day Summary

The eighth annual Scientific Day on Prader-Willi Syndrome was held Wednesday, July 14, the day before the national conference, in Scottsdale and featured 12 research presentations. Dr. Suzanne Cassidy coordinated the preconference day and summarized the findings of these studies for the main conference attendees on July 15.

In addition to sharing new findings, the research professionals established collaborative approaches for future studies in several areas related to PWS – specifically, clinical testing of the diagnostic criteria that were recently set forth, psychiatric issues, and the study of brain samples to try to identify the gene responsible for the characteristics of PWS. Dr. Cassidy, who explained this last area of research in the January-February issue of *The Gathered View*, reiterated to families at the conference the importance of considering this research need when a loved one with PWS dies. (Researchers need to be notified immediately so that brain tissue can be properly preserved for study.)

Following is a brief listing of the research presented on Scientific Day:

- ***Investigation of thermoregulatory characteristics in patients with Prader-Willi syndrome compared with other neurodevelopmentally impaired children.*** M.S. Williams, B.L. Rooney, J. Williams, K. Josephson, Gunderson Clinic, La Crosse, Wisconsin; and R. Pauli, University of Wisconsin, Madison.

- ***Confirmation of parasympathetic nervous system deficiency in Prader-Willi syndrome by measuring respiratory sinus arrhythmia.*** F.J. DiMario, Jr., L. Bauer, J. Volpe, University of Connecticut School of Medicine, Farmington; and S.B. Cassidy, University of Arizona School of Medicine, Tucson.

- ***Developmental changes in sleep related breathing patterns in children with Prader-Willi syndrome.*** G. Hertz, M. Cataletto, S.H. Feinsilver, and M. Angulo, Winthrop University Hospital, SUNY Stony Brook, Mineola, N.Y.

- ***Hypoventilation, oxygen saturation levels and Prader-Willi syndrome.*** J.M. Hanchett, The Rehabilitation Institute of Pittsburgh, Pa.

- ***Photoanthropometric study of craniofacial features in Prader-Willi syndrome.*** G. Levine, J. Le, and M.G. Butler, Vanderbilt University School of Medicine, Nashville, Tenn.

- ***Aging in Prader-Willi syndrome: 22 patients over age 30 years.*** S.B. Cassidy, University of Arizona School of Medicine, Tucson; A. Devi, C. Forte, C. Mukaida, University of Connecticut School of Medicine.

- ***A five-year-old boy with clinical Prader-Willi syndrome fulfilling criteria for autism.*** V.A. Holm, M.L. Acosta, University of Washington, Seattle.

- ***Duplication of chromosome 15q in Prader-Willi and Angelman syndromes: a gene dosage paradox.*** A. Mutirangura, A. Kuwano, F. Greenberg, D.H. Ledbetter, Baylor College of Medicine, Houston, Texas; W.P. Robinson, University of Zurich, Switzerland; S. Malcolm, University of London, England.

- **Small nuclear ribonucleoprotein polypeptide N (SNRPN), a maternally imprinted gene in mouse implicated *in Prader-Willi syndrome in humans.*** S.E. Leff, M.L. Reed, E. Sanjines, T. Ozcelik, U. Francke, Stanford University, Conn.; C.I. Brannan, N.G. Copeland, National Cancer Institute, Frederick, Md.

- ***Prenatal diagnosis of chromosome 15 abnormalities in the Prader-Willi and Angelman region.*** S. Toth-Fejel, S. Olson, R.E. Magenis, Oregon Health Sciences University, Portland.

- ***Pharmacological treatment of food stealing and excessive food consumption in three persons with Prader-Willi syndrome.*** J. Jung, W. Fisher, C.M. Anderson, N.C. Grace, L.G. Bowman, L.P. Hagopian, R.O. Carpenter, Kennedy Krieger Institute and Johns Hopkins University School of Medicine, Baltimore, Md.

- ***Enhancing self-management skills in people with Prader-Willi syndrome.*** D.A. Coleman, Jr., The Devereux School in New York, Red Hook.

Two additional papers were submitted but not presented in person at the Scientific Day. These were:

- ***Changes in body composition after human growth hormone treatment of Prader-Willi syndrome.*** F. Greenberg, K. Hwu, J.M. Pivarnik, H. Henson, B. Brown, W. Klish, P.D.K. Lee, Baylor College of Medicine, Houston, Texas.

- ***Smith-Magenis syndrome (del(17)(p11.2)) and its clinical overlap with Prader-Willi syndrome.*** F. Greenberg, P.I. Patel, J.R. Lupski, Baylor College of Medicine, Houston, Texas; R.E. Magenis, Oregon Health Science University, Portland; B. Finucane, Elwyn Inc., Elwyn, Pa.; A.C.M. Smith, PRISMS, Reston, Va.

(A compilation of the research abstracts is available from the PWSA (USA) National Office for $11, including postage and handling.)

Educating the School Staff

by Linda Keder
Co-Editor, The Gathered View

Just in time for the start of a new school year, Barb Dorn, President of the PWSA of Wisconsin chapter, outlined for national conference attendees her strategies for preparing school staff to meet her child.

Barb recommends that parents do a presentation for school personnel to explain what PWS is, how it affects your individual child, and what management strategies the school is likely to find most successful with your child. She advises parents to:

- Ask the building principal or special educator to set up the meeting. (If the school is unwilling to give you the time, Barb suggests you deliver your presentation in writing and/or ask to add "staff training" time to your child's IEP.)

- Identify your *main* concerns and focus on those.

- Take to the meeting photographs of your son or daughter so no one loses sight of the individual child involved. (Barb created a portable display by hinging two bulletin boards together and arranging on them photos of her son and information about his needs and PWS in general.)

- Leave with the staff written information about PWS that is appropriate to your child's age and needs.

The Meeting Format

The following agenda is one that Barb has found successful:

1. **Welcome the staff and state the purpose** of the meeting.

2. **Describe your child as an individual,** focusing on his/her strengths and interests. Show the photos or display.

3. **Present a general overview of PWS** including the genetic cause, the two distinct stages ("failure to thrive" and "thriving too well"), and the health risks. Specifically, your child's educators may need to know about the complications of obesity, the increased risk of joint trauma due to hypotonia, increased bruising, the need for extra sun protection in those with fair skin, the high pain threshold, and skin picking.

4. **Recommend management strategies** for the specific characteristics of PWS that impede your child's learning process. (See page 7 for suggestions.)

5. **Describe how PWS affects individual teachers,** such as the regular or special education teacher, art teacher, physical education teacher, librarian, etc.

6. **Summarize your points, and thank the staff.** Encourage them to read the additional materials and to order the textbook, *Management of Prader-Willi Syndrome.*

Specific Management Tips for Educators

Barb stresses that teachers may have misconceptions about a child's ability to gain control of the food drive, the rigid thinking, or other problematic behaviors associated with the syndrome. Parents need to clear up any misconceptions right away by explaining that PWS is a brain dysfunction that prevents affected children from developing certain inner controls.

Rather than trying to change the behaviors, teachers need to find ways to adapt the environment. The key to helping kids with PWS, Barb says, is to "provide supports in their environment in order for them to function effectively and in some cases for them to survive."

Barb points out seven common PWS traits in her presentations to teachers and suggests possible management strategies (see page 7). She emphasizes that each parent's presentation needs to be tailored to the particular child involved and to recommend strategies that are known to be effective with that child.

In addition to the general concerns outlined on page 7, Barb suggests that parents tell individual teachers about the specific ways PWS might affect their child in that teacher's class. For example:

- *The classroom teacher* may need to know that children with PWS tend to be good at reading and have more problems with math, that they need concrete items to be able to understand concepts, and that they need visual cues in addition to verbal ones.

- *The art teacher* should be advised if your child needs extra assistance in drawing or other art activities. Many children with PWS need projects broken down into simple steps using simple shapes; they "can't just see and do." (An occupational therapist may need to be involved.) Ask the art teacher to avoid using food items in art projects, for obvious reasons.

- *The physical education teacher* needs to know that children with PWS tend to be weak in fine and gross motor skills, have poor joint stability (which may put them at risk for injury), and fatigue easily. The P.E. teacher may need to modify activities and to direct the child to stop before reaching his or her endurance limits. (The physical therapist can be a resource for this class.)

In order to be successful, Barb concludes, your presentation must be personal, accurate, informative, and practical.

Barb Dorn is a nurse and the mother of 8-year-old Tony, who has PWS. Barb was a driving force in organizing the PWSA of Wis. chapter, which achieved official status in May 1992, and serves as its president. She has shared her parenting experiences in previous issues of The Gathered View, her chapter leadership experiences in the Chapter President's Quarterly, and she describes her son's educational placement and related supports in the July issue of Prader-Willi Perspectives.

Barb included in her conference handout a list of articles and other information resources for teachers and therapists. Call the National Office if you would like a copy.

In addition, a handbook for educators has just been published by Prader-Willi Perspectives. The 15-page booklet, entitled "Children with Prader-Willi Syndrome: Information for School Staff," by Karen Levine, Ph.D., and Robert H. Wharton, M.D., is available for $... a copy, with a discount for multiple-copy orders. For information, call Visible Ink Incorporated, 800-358-0682.

Information for School Staff

(Adapted from Barb Dorn's conference handout, July 1993)

Common PWS Characteristics

Rigid Thought Process

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.

Perseverative Thinking

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. Perseveration can lead to loss of emotional control.

Tenuous Emotional Control

Any combination of life stressors can lead to emotional "discontrol" as evidenced by tantrums – yelling, swearing, aggression, destruction, self-injury. Recovery of control takes time and is often followed by sadness, remorse, and guilt.

Difficulty with Peer Interactions

While our children want and need other children and value friends, it may be difficult for them to be exposed to the unpredictability of others for long periods. The need for order also translates into fairness issues and comparing themselves to others, often resulting in anger.

Food Craving and Diet Restrictions

For people with PWS, the craving for food is unusually strong, and food is metabolized at a rate that causes extraordinary weight gain. Food must be monitored and the individual supervised.

Poor Stamina

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.

Scratching and Skin Picking

These two behaviors are often seen in individuals with PWS and may be worse during stressful times. Combined with the higher pain threshold, these behaviors can result in tissue damage if not controlled.

Possible Management Strategies

- Give advance notice of change and allow for discussion
- Use lists and schedules (but avoid stating exact times)
- Tell parents in advance of changes in staff
- Break procedures into specific, orderly steps
- Don't make promises you can't keep

- Coax child to a resolution or another topic
- Defer discussion to a future time
- Avoid power struggles and ultimatums

- Prevent loss of control by encouraging communication, acknowledging feelings
- A child who is losing control should be removed from the sight of other children and taken to an empty classroom (not a closet)
- Provide soothing activities until the child has recovered
- Try talking about how the child might handle a situation better the next time
- Provide reassurance and positive closure

- Try the child in small groups rather than large ones
- Verbally rehearse planned social/play times; limit the length of social encounters
- Consider whether social difficulties are too great for full-day placement in a regular education classroom

- Supervise the child at lunch, and follow the parents' guidelines if the child is seen "trading" or "stealing" food from others
- Address the problem in private; don't embarrass or punish the child for taking food
- Let the child take a toy or activity to the lunchroom or eat lunch with a friend in the classroom, if either would reduce stress and temptation
- Follow the parents' guidelines on special snacks and holiday treats
- Don't put the child in a situation where he is alone with food
- Don't delay snack or lunchtime without warning

- Follow parents' suggestions for heading off sleep (e.g., send child on an errand, send to health room for a breathing/blowing activity to expand lungs)
- Plan a high-energy activity for the period following lunch

- Provide activities/materials to keep hands busy (some parents have found a particular object their child likes to handle during stressful times and can carry in a pocket)
- Provide supervision; explain that you are trying to help the child learn self-care
- Reward and praise for not picking
- If behavior increases, consider what may be adding additional stress in the environment; is there too much free time?

1993 PWSA (USA) Conference

"This gathering helps us to understand many issues better. You sometimes seem to forget why your child has certain behaviors. Talking and listening to other parents helps a lot."

Allison Mahan, cowboy, and Dayne Slade pose before exploring the western town of Rawhide.

Attendees enjoy a hayride to their barbeque in the desert.

"I will try to be more patient and understanding of my child's behaviors. Even though I have always known they were a result of the syndrome, it helped to hear so many others who have been through the same thing."

"I learned many techniques and basic information essential to the duties of my position as a case manager."

"I liked the moderated sessions where we learn from the true experts—each other."

Al Heinemann prepares to swing his partner, Sheila Evetts.

Shawn Cooper, Kristen West, and Ricky Lacy take a breather.

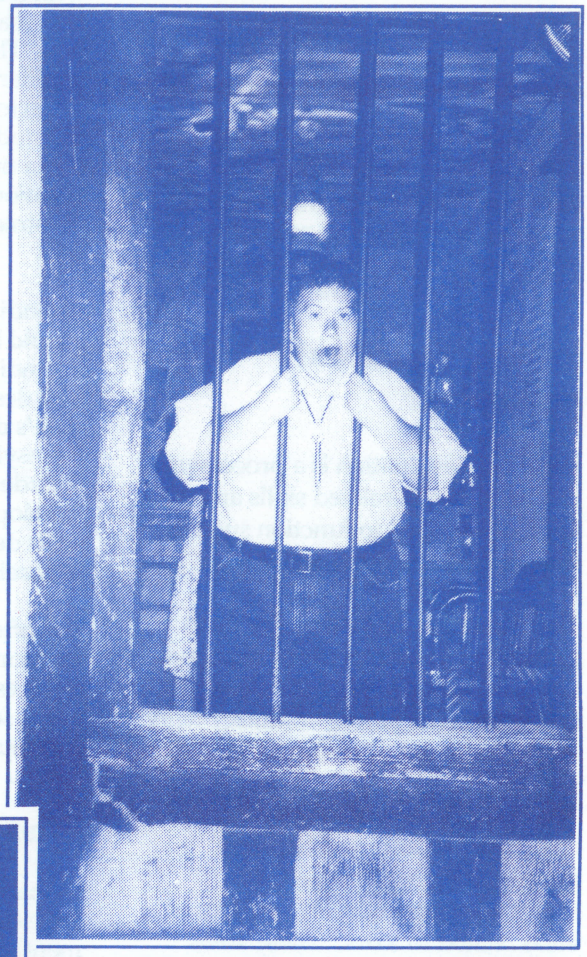
in Photos

"The conference gave us a greater awareness of the effects of PWS. I have a clearer picture of the dysfunctions and how to more effectively cope with them in the future."

"The meeting offers good family support and a well planned youth program."

"Just super. This was our first, and we will be annual participants and hope to become more involved to help our organization."

"The meeting gave me renewed energy and renewed strength."



Jerad Styer gives his prisoner impression.

Adam Sporbert and Kristen Ewish practice a tricky move.

"I now have a greater understanding of Prader-Willi syndrome and what are realistic expectations for a person with PW."

"The meetings help to prepare for the future."

Vocational Evaluation

What It Means for People with Prader-Willi Syndrome

Second in a series of articles on work

by Anna Marie Saporito, M.Ed., C.R.C., C.V.E., and Melanie Grace, B.S., C.R.C., C.W.A.

A "vocational evaluation" is a valuable and necessary tool used to assist people entering the work world by determining their work capabilities. This article will provide useful information for people considering entering the world of work and for their families. In addition, specific issues related to evaluation of individuals with Prader-Willi syndrome also will be discussed.

What is vocational evaluation?

Vocational evaluation is a process used to identify work-related skills and aptitudes needed to function successfully in the world of work.

The evaluation process begins with gathering information on educational/technical background, previous work experiences (including volunteer work), and interest areas (work and leisure).

The evaluation continues by administering a series of tests and/or work samples designed to assess the individual's aptitudes in a variety of areas related to work. These areas include academics, motor skills, cognition, and work behaviors. As the information is reviewed and synthesized, a profile of the individual as a worker is developed.

In addition to determining a person's present vocational level, recommendations are generally developed to ensure success in the workplace. The need for special accommodations, use of strategies, behavioral management techniques, and adaptive equipment are just some of the areas that may be addressed.

Who decides if and when an evaluation should occur?

Most state offices of vocational rehabilitation strongly recommend that students with disabling conditions open a case file two years prior to high school graduation. This allows the state to become familiar with the student and family and begin to explore possible resources for the student. The student or family can begin the process by phoning their state vocational agency, scheduling an appointment, and completing the application.

An individualized transitional plan (ITP) should be established and included in the individualized education plan (IEP) by a designated age, as dictated by your state's department of education (in Pennsylvania the age is 14.5). The ITP includes a written plan for life skills development, community awareness skills development, prevocational counseling, and vocational evaluation.

Too often students leave school and wait for months or years to receive vocational services. Families should become aware of the services that are available in the community and initiate appropriate action.

Who does the evaluation?

In many cases, a vocational evaluator, rehabilitation counselor, or work adjustment specialist conducts a vocational evaluation. These individuals are found in many settings: state offices of vocational rehabilitation, public and private rehabilitation facilities, and some vocational/technical programs.

Much of the information provided by teachers, psychologists, occupational therapists, physical therapists, speech-language therapists, and previous work supervisors can prove to be valuable in establishing an accurate profile of a worker. Using relevant and recent information can save time and energy for both the evaluator and the person being evaluated.

Who pays for the evaluation?

For individuals with developmental disabilities, funding for evaluations can be made by the state office of vocational rehabilitation, community mental health/mental retardation agencies, some private insurances, and self-payment.

What is the difference between formal and situational assessments?

Types of evaluations may differ from agency to agency. They may provide formal assessments, situational assessments, or a combination of the two. Formal assessment utilizes standardized tests. Scores can be compared with specific norm groups. Most formal tests are paper-and-pencil type and are generally timed. Other formal testing instruments are standardized work samples, or pieces of work, that determine skills, aptitudes, and level of interest for specific working situations.

Simulated work activities are utilized during situational assessment. These work activities are also pieces of various types of work. However, the evaluator has the opportunity to view the learning process and make other observations that may be missed by formal tests.

Information from both types of assessment is very important in determining the vocational strengths of an individual and in matching these strengths to jobs that actually exist in the community.

Who receives the results of the evaluation?

All vocational evaluation results are considered confidential and should be treated as such. Information release consent forms should be signed before any confidential records are sent.

The agency that provided the funding for the vocational evaluation generally receives the results. They are reviewed

with the person who was tested and, in most cases, with the family. Copies can be made available upon request. If the individual is planning on attending a training or work program in the community, it is recommended to forward a copy to avoid unnecessary testing.

What are some important things to consider when evaluating individuals with Prader-Willi syndrome?

(It is suggested that parents share the following information with the person doing a vocational evaluation on their son or daughter.)

Many individuals with PWS exhibit difficulty in concentrating while sitting for long periods of time. Distractibility and, at times, sleepiness may have a negative effect on test results. Selecting tests that are not lengthy may provide more reliable results. Also limiting distractions in the evaluation area is extremely helpful.

Allowing the individual to stand for short periods of time while being tested may prevent sleepiness from interfering with testing. It is important to caution evaluators that very obese persons may have swelling or infections in their lower extremities that will require them to elevate their legs while seated.

Individuals with PWS may present unique expressive and receptive language skills. It is common that expressive skills are much higher than receptive language. This means that a person with PWS may say that he/she understood an instruction but, in actuality, may have missed some of the details or misinterpreted the instruction. This can often lead to argumentativeness or excessive question asking.

There are some useful techniques to minimize potential problems in this area. Limiting the amount of information presented at one time, asking the person to repeat the information, clarifying any discrepancies before the person begins testing, and maintaining a supportive, positive interaction style will ensure a successful evaluation.

Motivators or behavioral intervention techniques that have been successful with the individual in other settings, such as in school, work placements, residential arrangements, home, and volunteer work, can be utilized to minimize problem behaviors resulting from the normal stresses that come with being tested.

It is important for evaluators to keep in mind that, although people with PWS possess many similar characteristics, each individual may also possess unique aptitudes, skills, or learning abilities that are atypical of the syndrome. Evaluators should look beyond generalizations and strive to maintain objectivity in spite of frequently unflattering reports about the person they are testing.

We hope that you find this information useful in obtaining vocational evaluations, dealing with evaluators, and utilizing evaluation results. We welcome any comments or questions you may have about the information. *(The authors can be contacted through the PWSA (USA) National Office.)*

Chapter Notes

Pa. Mini-Conference – Local support group Prader-Willi Western Pennsylvania Association and The Rehabilitation Institute of Pittsburgh (TRI) will host a conference at TRI on Saturday, Oct. 2, for members of PWSA of Pa., vocational and group home providers, and base service unit personnel in the region. Contact: Sandy Immekus, 412-831-9291

Chapter Calendar

PW Kentucky Association – general meeting Sept. 18
Contact: Willie Lacy, 502-968-2626

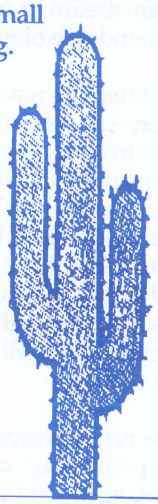
PW Northwest Association general meetings – Sept. 11 and Nov. 6
Contact: Powell and Joane Underwood, 206-641-9452

PWS Assocation of Ohio – general meeting Nov. 6
Contact: Jim Boyle, 216-932-3587

PWSA of Wisconsin – picnic Aug. 28, group home tour Sept. 18, business and support meeting Nov. 6
Contact: Barb Dorn, 608-838-9535

Thank you, Arizona!

We would like to offer our warmest heart-felt thank you to the Arizona chapter for making July in Phoenix an experience many of us would be happy to relive. Many of the Board of Directors, Officers, attendees, and participants in the Youth/Adult Activity Program have expressed gratitude to the host committee for their hours of dedication to make the 1993 National Conference a "cool", unusual, and educational experience. There's no doubt that the banquet among bright stars, majestic mountains, and cactus galore was truly breathtaking and memorable. We would specifically like to thank Vanessa Russell for her leadership. Vanessa's adventurous spirit led her and a sprinkling of others into the maze of planning a conference. Even with their small committee, they produced a smooth, creative, and fun meeting. In addition, we would like to recognize the efforts of: John and Jeanne Ewish, Annette Ruiz, Teresa Kellerman, Jean Alley, Christy Montgomery, Leona Smith, Tony and Marie LaPenta, Charlie and Rita Enlow, Rosemary and Joe Grout, Eleanor Ewish, and Tony Ellis, for their host committee work. We would also like to commend Marge Wett for her long hours at the registration desk.



Special thanks also go to Gail Overton for organizing the Group Home Day, Suzanne Cassidy for moderating the Scientific Day, Dana Bintz for managing the Youth/Adult Activity Program, and Kira Weiss for managing the Child Care Program.

Normalization! Independence! Mainstreaming!

How Much Is Enough and How Much Is Too Much?

Perspective from Parents of a Prader-Willi Adult

by Ruth Chausow, M.A., Social Sciences, and Hymen M. Chausow, Ph.D., Executive Vice-Chancellor for Academic Affairs, Emeritus, City Colleges of Chicago

We did not have a diagnosis for our daughter until she was 31 years old. Had we received a correct diagnosis when she was very young, our lives and hers would have been easier. Behaviors that were unexplainable by everyone suddenly became understandable in light of the syndrome. But everyone's inability to respond appropriately to her problems -- family, siblings, friends, teachers, doctors, etc., made life very difficult for her, as well as for everyone with whom she came into social contact.

The present climate, both nationally and statewide, is to mainstream *all* disabled individuals, regardless of the level of disability. Theoretically, the goal is excellent, but it is not realistic for everyone across the board.

The basic assumption of the theories of normalization and mainstreaming is that, given the opportunity, a normal independent life can be achieved. The new Americans with Disabilities Act (ADA) provides that disabled individuals be given this opportunity -- a most laudable goal! For disabled individuals capable of making choices, removing obstacles can work. People in wheelchairs would need ramps, wider doorways, etc., to be able to mainstream in our society -- a very commendable objective.

Very often we read that *all* disabled persons should have the same rights as others to take risks and chances in order to improve their lives. It is said, in conjunction with these thoughts and ideas, that people learn from their failures. But it is also true that people build upon their success, which motivates them to further efforts for more successes. We call this positive reinforcement.

Let us now consider "normalization," "independence," and "mainstreaming," as they apply to Prader-Willi individuals.

We know that most persons with PWS can be and usually are insatiable eaters if food is available; if left unsupervised they can eat themselves to death, in the literal sense. Most are moderately or mildly mentally and/or functionally retarded and have difficulty in keeping up with their peers and being accepted by them.

If the food access is not supervised and limited, the inner controls for self-control in this aspect of their lives are not present, no matter how much we would like to hope and wish otherwise. Their own awareness that they cannot exercise self-control, and the reactions of those around them to this lack of control, often set off behavior problems and tantrums. The continuous feelings of hunger never fully satisfied also affect their behavior.

Just as you need ramps and other environmental changes for persons in wheelchairs, persons with PWS need changes in their environment called "food access control." Consistent environments must be in place -- restricting access to refrigerators and kitchens at home and supervised controls as much as possible in the community and workplace. Educating all persons in their environment about the syndrome, which should lead to changes in *their* behaviors, is also necessary. Most PWS persons, if left completely "independent," will orient much of their behavior in the long run to food-seeking.

We believe that most of our people will not be able to be completely mainstreamed, to the degree that our social welfare agencies would like. The assumption that self-control in eating is attainable and will not often overwhelm other more desirable behaviors, will not work in the long run for most, if not all, persons with PWS.

We should always keep in mind that Prader-Willi syndrome is a genetically determined disturbance in chromosome 15 for which there is no cure or effective palliative as yet. It is an unyielding presence which thus far drugs and behavior therapy can only partially modify.

Certainly there are different ability levels among persons with PWS, and some can be more independent than others. But in many cases, *where they have the environment limited for them* related to food access, they feel less stressed because these overwhelming temptations are not ever present. They often can be *more* productive citizens in other aspects of their lives when some controls are in place.

To maximize the avowed goals, programs should be developed to allow more choices *within limits* to develop, with help, improved social skills and encourage community involvement that will encourage successes.

We should make sure that necessary prerequisite skills are learned before we move our PWS persons into "mainstreaming" and "independence." Many of our PWS persons have guardians because they are not able to make rational choices at all times.

Let's not consider the goals on an either-or philosophy (dependent vs. independent) but rather on a continuum with the goal of maximizing the normalization, independence, and mainstreaming of each individual.

(The authors welcome comments from readers of The Gathered View.)

On a Sad Note. . .

Hy and Ruth Chausow's daughter Diane, age 45, died suddenly in early July while on a nature outing with her group home companions and staff. Diane was a quiet, serious, intense little lady who struggled every day of her life to respond to internal needs over which she had no control. Her parents left no stone unturned seeking answers and help that eventually led to Diane's move to a wonderful group home program in Glenview, Ill., where she lived for the last eight years of her life. The Chausows had submitted the above article several months before their daughter's death and kindly allowed us to proceed with publication of it. There is no way to diminish the pain of their loss; we can only share a portion of their sadness.

-- Louise Greenswag

Toilet Training Made Easy

After several years of anxiety about when and how we would ever potty-train our preschooler with PWS, I'm delighted to report that we just had great success in a very brief time, using a unique procedure from a 1974 book called *Toilet Training in Less Than a Day.* The authors, Nathan Azrin and Richard Foxx, are psychologists who created the technique originally to teach profoundly retarded adults, then developed it further to teach children of all intelligence levels.

The approach requires careful planning and your full attention for at least half a day. It employs many different teaching tools and reinforcement techniques in a short period of time to teach the child every step of the toileting process from getting to the potty on time to emptying the pot and flushing the toilet.

Several key ingredients in the process are: having the child first teach a doll that drinks and wets, continually offering the child drinks to provide many opportunities to practice urinating, rewarding dry pants and successful trips to the potty with edible treats (for the first few hours only), and responding to accidents as something requiring cleanup and practice. There's more to it than I can describe in this space, however. You really need to read the book; in fact, I found I needed to read through it twice before starting our "potty day."

As predicted, my daughter was independently using her potty in a matter of a few hours, and the process was fun overall. The roughest spot was when she wet herself for the first time and cried inconsolably; having a teaching plan kept me from giving up at that point. Lesley averaged one or two accidents a day for the first week, then took a weekend trip with us accident-free, and it's been relatively clear sailing since. She still needs a diaper or Pull-Up at night, but at age 4½ she's crossed the big hurdle.

I know of several other families who've used this toilet training method (including another family in our area who trained their 3-year-old daughter with PWS), and all swear by it, whether or not their children have disabilities.

A few more tips:

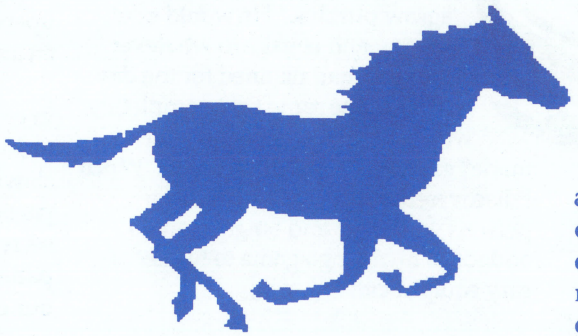
Because I wanted to ensure the best chance for success, I put off the training until I knew Lesley was physically able to remove her pants and get on and off a potty chair. I waited a little longer until summer so she didn't have to cope with heavy clothes and long school bus rides. I packed away summer clothes that she couldn't remove independently and added more dresses to her wardrobe. The last stumbling block was our toilet handles, which Lesley was too weak to flush. We discussed the problem with an occupational therapist, who then extended the handles with a moldable substance called "orthoplast" -- it works like a charm! We now have an independent little girl who's quite proud of her new skills -- and all her new underpants. Here's hoping our experience can help someone else.

Linda Keder
Silver Spring, Maryland

The Benefits of Horseback Riding

Several families at the national conference advocated therapeutic horseback riding for children with PWS. They cited benefits both in improved muscle tone and improved self-esteem in their children who've been engaged in this activity. Therapeutic riding is a specialized horseback riding program that is designed to meet the needs of individuals with disabilities, usually with the guidance of a physical therapist. It is sometimes referred to as "hippotherapy," or therapy on a horse.

According to an article in the June/July 1993 issue of *Parenting* magazine, which publishes a periodic column on disabilities, therapeutic horseback riding can "enhance coordination, balance, and muscle tone" for kids with various neuromuscular problems, and "children with mental retardation or learning or other cognitive disabilities, can improve their concentration and attention span."



Most riding centers that offer this kind of program are accredited by the North American Riding for the Handicapped Association (NARHA), which requires instructors to have at least 120 hours of experience teaching riders with disabilities. Facilities vary widely, however, and should be checked out firsthand.

According to the *Parenting* article: "The special equipment that's needed -- riding helmets, safety stirrups, and customized saddles or sheepskins (for better stability) -- is provided or lent to riders by some programs. Full-time physical therapists are on duty at some riding centers, while others can afford only part-time or volunteer consultation by therapists. The costs differ, too. Instruction can be free or as much as $25 per hour. Many stables offer scholarships or arrange subsidies through other organizations; some parents are reimbursed by insurance."

For more information, call NARHA at 800-369-RIDE. You should also consult your child's doctor or physical therapist to make sure that they would recommend horseback riding for your child.

NICHCY:

Information for Everyone

If you are not familiar with NICHCY – the National Information Center for Children and Youth with Disabilities – you are missing a great source of free information that can be very helpful in raising a child with PWS or any other disability. A federally funded information and referral clearinghouse, NICHCY publishes a number of excellent booklets and fact sheets on disability issues and provides personal telephone assistance to families and others who need such information.

The first time you call NICHCY's toll-free number, you should ask for: their publications list; their "State Resource Sheet" (publication no. GR6) for your state, which lists officials in your state's key agencies and organizations serving the developmentally disabled; and their "Public Agencies Fact Sheet" (GR4), which describes the various types of state agencies and their functions.

If your child has recently been diagnosed, you might also want to request "You Are Not Alone" (PA1), "Children with Handicaps, Parent and Family Issues: A Guide to Readings" (ND3), and "Parents' Guide to Accessing Programs for Infants, Toddlers, and Preschoolers with Handicaps" (PA2).

NICHCY has published many excellent booklets on education of children with disabilities. Available are separate publications on IEPs, early intervention, psychological testing, least restrictive environment, related services, procedural safeguards, vocational education, sexuality education, transition, minority issues in education, and an overview of special education laws. NICHCY also offers a number of publications on family issues, one of the most recent being a 1992 booklet on estate planning for families of children with disabilities.

Single copies of NICHCY's publications are provided and mailed free of charge, and there is no restriction on photocopying. Most publications include references and reading lists for further exploration of the topic.

NICHCY can be reached by mail, phone, or electronic mail:

NICHCY
P.O. Box 1492
Washington, D.C. 20013-1492
(800) 999-5599 toll-free
or (703) 893-6061 in the D.C. area
SpecialNet or SCAN User Name:
NICHCY

Calling all Siblings . . .

The survey questionnaire on page 15 was distributed to a limited number of attendees at the national conference. Because there were insufficient copies of the survey for all those who were interested, the questionnaire is included in this issue so that any sibling who would like to participate will be able to do so. (Please feel free to make copies of the survey if more than one sibling wants to complete one.)

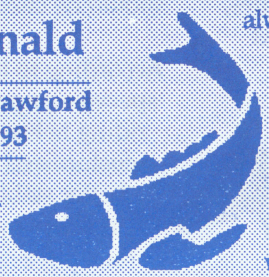
The purpose of the questionnaire is to provide information for one or more articles that Lota Mitchell plans to write for *The Gathered View*. Lota, who has a Master's degree in social work and has conducted workshops about siblings of the mentally retarded, wants to explore what special issues exist in growing up with someone with PWS. Siblings of all ages are encouraged to respond.

Names and addresses are not required on the survey, but would be helpful in case clarification of an answer or more information is needed. No names will be use in any articles written about this survey, and the questionnaires will be kept confidential.

I Will Miss Donald

A Tribute to Donald Crawford
July 6, 1952 - July 13, 1993

by Barbara Y. Whitman, Ph.D.



My personal association with persons with Prader-Willi syndrome and their families began in late 1981, when Janalee Heinemann asked the Knights of Columbus Developmental Clinic to provide a home and support person for the newly forming Missouri PWSA. Within a few months, the support group consisted of several families with children ranging in age from 4 to Donald who was 34.

Many of the "young'ns" were typical in their behavior, mercurial in both mood and actions. Donald, however, was a rock. He always brought his supply of interests to our monthly meetings – crossword puzzle books full of word searches, scissors and crayons, jigsaw puzzles. He would take his seat and engage in whatever activity I had planned for the day while at the same time completing word searches and making "Donald-made" greeting cards for everyone. When I discovered the love affair between persons with PWS and bingo, Donald added his own bingo game to his necessary equipment.

Donald, unlike the rest of our group, was extraordinarily quiet. He always participated but never talked unless directly asked a question. His quiet ways attracted the "little guys," and over time I learned to seat the littlest ones next to him so he could quietly shepherd them and help them with their crafts.

Only twice did I see Donald bristle – once when someone "borrowed" his word search book and once at a weekend camp when someone touched his fishing equipment, upending his cup of worms. Donald was an avid, if underaccomplished, fisherman.

In recent years, Donald's equipment included his rolling oxygen tank, but it slowed him not one bit nor kept him from participating in any activity. I always secretly saluted his mother, Marian, for her patience as traveling with Donald to even our meetings was a major packing job.

On July 13, 1993, Donald's heart finally gave out. I will always remember him fishing at camp and as my anchor at the head of the table in our group. Over the years I have collected Donald-made Christmas cards, valentine cards, Easter cards, potholders, trinkets, and memories. I will miss him.

Questionnaire for Siblings of Persons with PWS

Please return to Lota Mitchell at 844 Foxland Drive, Pittsburgh, PA 15243, by September 30. Please feel free to add a page if you want more space. Thanks for your help!!

NAME_____ PHONE_____

ADDRESS_____

Your age at present_____ Your sex Male_____ Female_____

Number of siblings in your family (including you)_____ Number of brothers_____ Number of sisters_____

Your birth order position (e.g., 1st of 3)_____

Age of your sibling with PWS at present_____ Sex of your sibling with PWS Male_____ Female_____

Birth order position of your sibling with PWS_____

1. What are/were the benefits, if any, to you of having a sibling with PWS?

2. What are/were the negatives, if any, to you of having a sibling with PWS?

3. What do/did you like about the way your parents handled things?

4. What do/did you dislike about the way your parents handled things?

5. What would you like/have liked them to do differently?

6. How is having a sibling with PWS affecting you in the present, if at all?

7. Other comments?

Get Ready to Read the New Labels

Although food manufacturers aren't required to comply with the new federal food labeling regulations until May of next year, we can expect many of the new labels to start appearing in the coming months. Several free publications are available to help consumers learn how to read and use the new labels.

"How to Read the New Food Label" is a new government booklet offered by the Food and Drug Administration (FDA) and the American Heart Association (AHA). Request a free copy from: Consumer Information Center, Dept. 79, Pueblo, CO 81009.

"Focus on Food Labeling, May 1993" is an informative and attractive FDA special report that covers everything from the history of food labeling to how the new labels relate to the food pyramid. Single copies of the 64-page report, published by the monthly magazine *FDA Consumer*, are available free of charge. Ask for the "FDA Consumer Special Report on Food Labeling." Send requests to: FDA, HFE-88, 5600 Fishers Lane, Rockville, MD 20857.

The Gathered View is the official newsletter of the Prader-Willi Syndrome Association and is sent to all members. The opinions expressed in *The Gathered View* represent those of the authors of the articles published, and do not necessarily reflect the opinion or position of the officers and Board of Directors of PWSA (USA). Duplication of this newsletter for distribution is prohibited. Quotations may be used if credit is given to PWSA (USA). Membership dues are $21 per year Individual, $26 per year Family, $31 per year for Agencies/Professionals (U.S. Funds). Send dues, change of address, and letters to: 1821 University Ave., W., Ste. N356, St. Paul, MN 55104-2803. Any questions? Call us at: 800-926-4797 or 612-641-1955 or Fax 612-641-1952.

Prader-Willi Syndrome Association (USA)
1821 University Avenue W., Suite N356
St. Paul, MN 55104-2803

First Class Mail