



## PWSA (USA) - 1994 National Conference

# 'Peachy' Plans for YAAP

Lots of "Peachy" activities are planned for the young people who have "Georgia on their minds" for this summer. The YAAP (Youth and Adult Activities Program) Committee is excited to be offering a "Bushel Basket" full of choices for children and adults with PWS, and their brothers and sisters, who will visit Atlanta with their families during this year's PWSA (USA) conference.



Sci-Trek will keep our minds active and engaged in some interactive learning. The Center for Puppetry Arts is appealing to all ages and includes a puppet-making workshop! For the more mature crowd, a trip to the Martin Luther King Center or the Carter Presidential Center will be offered.

In addition to the annual banquet held Friday night, the Georgia delegation will host a social (maybe poolside!) on Thursday night and a special awards luncheon before ending the conference Saturday afternoon.

Specialists in the areas of art, dance/movement, music, and social skills will be on hand to work with groups, divided by age, on our last day

together. At that time, a specialist on sibling issues will be available as well. Child care will be provided for children 5 and under in our special child care suite. Entertainers as well as experienced child care workers will be on site. Children and adults, and siblings, will travel together in age appropriate groups for the field trips mentioned above.

Many local, close-to-the-hotel activities will be available for "free time" with families (if you're not napping!) such as bowling, movies, shopping, and Chattahoochee River walks. Siblings will be treated to a little "down time" of their own--a hospitality suite will be reserved for them throughout the conference.

*"Georgia on My Mind"*

## 16th Annual Conference: July 20-23, 1994

### Wednesday, July 20th

- \* Scientific Day
- \* Residential Providers' Meeting
- \* Chapter Presidents' Meeting
- \* Conference Registration Opens

### Thurs.-Sat., July 21-23

- \* Conference Sessions
- \* General Membership Meeting
- \* Social Events

Atlanta Marriott Northwest  
I-75 at Windy Hill Road  
200 Interstate North Parkway  
Atlanta, Georgia 30339  
404-952-7900

Registration packets for the National Conference will be mailed to PWSA (USA) members around the first of April. Non-members may request packets by writing or calling PWSA (USA), 800-926-4797 or 314-962-7644.

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## Prader-Willi Syndrome Association (USA)

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Myra Miller, Assistant to the Exec. Dir.

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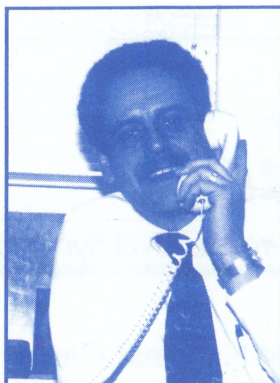
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## Out of The Office

by Russ Myler, Executive Director

The furniture, files, and history of the Association made the move to St. Louis January 7, 1994, without mishap. The office is now back in operation and in the first 20 days sent out over 45 information packets and 5 new member packets. January was spent organizing the office, preparing for the board meeting, and interviewing many excellent people for the assistant's position. The first month in St. Louis proved hectic, but very rewarding.

Myra Miller joined the staff in mid-January. Myra has extensive experience with all areas of office management, newsletter and brochure development, computer operations, conference and camp programming, and volunteer services. She is simply a wonderful person, you will like her, I'm sure. Myra moved to St. Louis from Springfield, Missouri, in the middle of the great freeze.

The next several months will be dedicated to "getting current" in filling orders, answering correspondence, and establishing office procedures. The office goal is to give fast, efficient, and understanding service to all. Communication and dissemination of information are two of the most important functions of the National Office, and your staff members are committed to fulfilling those functions.

PWSA (USA) has made a great deal of progress over the past 17 years. We should all be proud of the accomplishments, but we have a future waiting for us that is full of challenges to be met. To succeed in this future we must all act as a unified body, striving for common goals. Your national staff will be your full partners in reaching those goals.



Myra Miller

## Special Thanks to Our Contributors

*Donations received January-February, 1994*

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Mary & Jim Harrison,  
J.J. O'Grady,  
Mrs. Robert Randolph

1993-94 Angel Contributors will be listed in the next **Gathered View**. If there are names of other contributors not listed that should be, please notify the office. Please accept our sincere apology in the delay of this issue.



# Voices from Below

by Janalee Tomaseski-Heinemann

When Matt was no longer able to fit into the school system and our other children were around less to keep an eye on him, what we were going to do with Matt became more and more of an issue--thus began our battle to get a supportive living program opened.

Fortunately for us, we were able to reconvert our house into a temporary workable living situation for Matt. We have a split-level home with sliding glass doors both on the top deck and on the bottom level that go out to the back yard. We enclosed the stairway and part of the lower level into Matt's temporary "apartment." He had his own bedroom, living room, and bathroom. He had his own phone line so he could call us upstairs. We had flip locks installed on the door that prevented him from coming upstairs, so we could open it up at times, but tried to keep his place like a separate apartment. He had a small refrigerator with just diet soda. Matt could call us to check on meal times--and often did. We would have to tell Matt that, "Tomorrow is Saturday and we are sleeping in, so don't you *dare* call before 9:00 a.m.!" However, we forgot to forewarn a friend of ours who stayed at our home on Labor Day weekend while we volunteered at a camp, so she was awakened on Sunday at 4:00 a.m., 5:00, and 6:00 with Matt asking, "Is it breakfast time yet?"

The "voice from below" became a source of irritation or humor--depending on what mood Matt was in or we were in. How many call and say, "com"? Or, "I c a n n e d were also embarrassment, such as the time Sarah brought a new boyfriend home to meet us. Suddenly there was a voice from below that said, "Can I come up?" When I went and flipped the lock, Matt came bouncing upstairs and hugged the young man with his elbows (he kept his hands in the air because he had been picking his fingernails and they were bleeding) and asked him, "Are you an Indian?" Sarah's boyfriends either came with a sense of humor--or left quickly!

*"Is it breakfast time yet?"*

*"Are you an Indian?"*

"tenants" would  
"I smell pop-  
hear Dad say  
food"? There  
moments of

Although Matt's life was "on hold" during this time, he was content. Matt would sleep all morning and then a respite provider would come at noon and take him out to lunch. (We tried leaving his lunch in his refrigerator, but found that he would eat it five minutes after we left in the morning.) Then they would go to the museum, zoo, show, or wherever Matt chose to go. The only productive thing he would have to do was water the plants in the back yard and clean his room under our supervision twice a week. Of course, he would stay up at night since he slept all morning, but usually after we went to bed he was content to watch TV, play Nintendo, or (unfortunately) pick his nails and skin. Because of the complete lack of expectations from Matt, this was a quiet period and he seldom really "lost it."

During one of those weeks when Matt was being lovable and very rational, we got an unexpected call at 2:15 a.m. from a neighbor girl, who said in a shaky voice: "Mrs. Heinemann? There is someone in your back yard hollering and talking to God." We looked at each other and our hearts sank. After we threw on our robes and went outside, we could see Matt illuminated by the security light on the patio. He was waving his arms and hollering:

*"Mrs. Heinemann? There is someone in your back yard hollering and talking to God."*

"God! You need to help your son, Jesus. He's in trouble!" We convinced Matt that if he was worried about Jesus, he could pray quietly to God and God could still hear him--and also that he was scaring the neighbors. We knew enough not to try to convince him Jesus was not in trouble. Although Matt had done worse things before, he had never done anything quite this strange. On the other hand, we have learned to expect the unexpected. As far as what the neighbors think--they never asked and we never explained. We swallowed what pride we had left long ago and accept that we are probably known as "that weird Heinemann family."

*"...what we were going to do with Matt became more and more of an issue... thus began our battle to get a supportive living program opened."*



# Board Meeting Focuses on New Directions

A reception in the new PWSA(USA) offices, hosted by the Missouri chapter, launched the mid-year meeting of the Association's board of directors, held January 14-16 in St. Louis. During the two days of meetings that followed, board members and officers reviewed and reconfirmed objectives, briefed the new office staff, and further developed plans for new undertakings.



**PWSA-U.S.A.**

Jan. 15-16, 1994  
St. Louis, Missouri

*Board member Barbara Whitman, Ph.D., created this design as a theme for the board meeting and stitched a matching banner for the new office, marking PWSA's relocation to St. Louis, Missouri.*

## Highlights from the St. Louis board meeting include the following:

- \* The Public Relations and Fundraising Committees announced joint efforts to contact and seek major funding from nontraditional sources in 1994, such as foundation grants, corporate donations, and matching funds. The PR Committee also reported separate initiatives to network with medical associations and other groups to expand the knowledge and awareness of PWS and the Association.

- \* Board members approved a conceptual drawing for a new PWSA (USA) logo and fundraising brochure, developed over the previous five months by a design team working with Don Goranson, PWSA Vice President. The new logo and brochure will be unveiled in the coming months.

- \* The PWS Awareness Day event will be a major focus of the Chapter Presidents' Day meeting in July. The PR Committee has done considerable work developing a plan for PWS Awareness Day and is now targeting spring of 1995 for the event in order to allow chapters ample time to prepare.

- \* Vice President Don Goranson was appointed to chair the Public Relations Committee, succeeding Janalee Tomaseski-Heinemann.

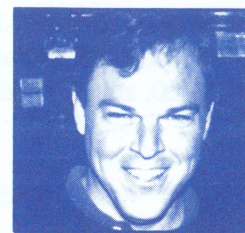
- \* The Publications Committee reported significant progress on the second edition of the Management textbook (projected to be available in July 1995) and on the remake of the Medical Overview videotape—two key educational products available from PWSA. Considerable time and effort are being committed to revising other Association products and to developing several new publications and video concepts.

- \* Progress was reported on a major effort to compile a national database of residential alternatives for persons with PWS. Pauline Parent was assigned responsibility for directing the Group Home Team through the data-gathering phase of this project. When complete, the residential alternatives database will provide a source of referrals for families seeking residential placement and an information network for providers.

- \* Board member Paul Wissmann was elected to the office of Treasurer, effective April 1, succeeding Jim Kane.

- \* Board election policies were reviewed, and several proposals were made. The board expects to vote on policy revisions and announce any changes in the next issue of *The Gathered View*. The Nominating Committee, chaired by Annette Ruiz, has begun its work of considering candidates for the seven board seats that will be up for election this July.

- \* A conflict of interest policy governing the conduct and relationships of PWSA officers, directors, committee members, and staff was proposed and approved by the board.



**Paul Wissman**



**Al Heinemann chats with Jon Shacklett and Andy Maurer at the board reception.**



**Jim Kane, Viki Turner, and Louise Greenswag, Ph.D., at the January board of directors' meeting.**



## Extended School Year: A Need for Many Children with PWS

by Barb Dorn, President, PWSA of Wisconsin

Throughout their school years, children are expected to acquire new skills and knowledge and continually build on what they learn to meet increasing academic demands. Some loss of new skills and knowledge (regression) occurs in all children when their educational programming is interrupted. It is my belief, however, that without an extended school year program, many children with PWS lose much more than academic skills. Around the age of 6, many children with PWS start to exhibit more challenging behaviors during unstructured times. This can be seen during school breaks, summer vacation, and, in some children, even on weekends. They regress in behavior and impulse control, as well as in appropriate peer interactions, which are necessary for community living.

Parents often see the need for an extended school year in planning a more successful summer for their child with PWS. Due to budget constraints, school districts may require more documentation and justification in order to provide these services.

### An Example from Wisconsin...

According to the Wisconsin Dept. of Public Instruction guidelines: *"a handicapped student is entitled to an education program in excess of 180 days per year if regression caused by interruption in education programming, together with the student's limited recoupment capacity, render it impossible or unlikely that the student will attain the level of self-sufficiency and independence from caretakers that the student would otherwise be expected to reach in view of his/her handicapping condition."* Remember, each state may define this differently, but the general concept will be the same.

Wisconsin has identified certain factors which make it likely that a child will meet the standard described above:

The purpose of this article is to share some strategies that may help parents secure extended school year (summer school) services for their child with PWS. Although each state will have minor differences in defining and documenting the need for an extended school year, I think the underlying needs and justification will be quite similar.

### Start with the State Requirements

First of all, parents should contact their state's Department of Education (DOE) or Public Instruction (DPI) and ask for information or guidelines on an extended school year for a child with special needs. State education offices often have a bulletin or paper that specifically addresses this topic.

The next step is to prepare a justification, examples, and proof of the child's need for an extended school year, following the format of the state document. For example, if the document first addresses

the nature of the disability, the parent should do this too. This approach helps to organize thoughts and ensures that each of the school system's considerations is addressed. I would also suggest that a copy of the state guidelines be included in the justification presented to school staff. Parents are advised to solicit help from teachers, therapists, social workers, physicians, and any other professionals who may be familiar with their child or with the syndrome. Having written documentation from professionals tends to be very influential in convincing school personnel.

If the characteristics or factors outlined in the extended school year guidelines apply to a child, the preparers of the IEP *must* take this into account and address those needs in the child's IEP. Parents may also request that these needs be evaluated and documented in the initial eligibility committee process--known in various states as the Multidisciplinary Team (M-Team), Placement Advisory Committee (PAC), or Admission, Review, and Dismissal (ARD) committee.

#### 1. Nature of Handicapping Condition

Children with certain disabilities require consistent, highly structured programs and are predisposed to regression when their programs are interrupted. Autism is given as an example of such a disability. Prader-Willi syndrome can be another.

We know that children and adults with PWS require consistent, structured programming. Parents seeking extended school year services need to include articles that support and document this aspect of PWS as well as providing specific examples of how their child reacts to interruptions in his school year (e.g., daily tantrums, increased skin picking, increased perseveration). In addition to published information, such as that available from PWSA, the child's physician could be asked to write a letter explaining how the nature of PWS necessitates an extended school year for the child.

#### 2. Severity of Handicapping Condition

Typically, children with severe emotional disturbances (or severe behavior problems) are more likely to revert to lower functioning levels or exhibit inappropriate behaviors such as withdrawal, anxiety reactions, or fetishes which interfere with learning when their program has been interrupted. For these children, an interruption also reduces their motivation and trust and may impede progress toward self-sufficiency.

Many children with PWS have severe behavior problems. They often exhibit inappropriate behaviors (e.g., crying, stubbornness, perseveration, skin picking, and temper tantrums) that interfere with learning, and they exhibit all kinds of "anxiety reactions" as a response to changes in their environment, routine, and program personnel. Children with PWS tend to feel a lack of trust in new personnel, resulting in constant questions and the need for continual reassurance. All these behaviors can be exacerbated when school is interrupted. Parents and teachers need to document which behaviors are prevalent in a particular child. Parents are often the best judges of the behavior regression that occurs during vacations, holidays, and on weekends and must be prepared to give specific examples.

Continued on p.8



# The Sibling Survey...

## What Siblings Have To Say About Having a Brother or Sister with PWS

by Lota E. Mitchell, M.S.W.

For four months the mailbox has yielded up treasures in the form of sibling surveys from 18 states, Italy, Canada, The Netherlands, and South Africa - 38 in all. Brothers and sisters of all ages shared their feelings about having a sibling with PWS and their parents' handling of the situation. Some of their opinions reflected individual situations; others represented feelings common among siblings of those with developmental disabilities. Attitudes ranged from angry (*"She's a pain!"*--girl, 9, of her sister with PWS, 8) to very loving (*"Other than the other things ... he's a great kid, and I love him."*--boy, 11, about his 7-year-old brother).

Perhaps the three words that appeared most in the responses were "patience" (both their own and their parents' or the lack of it), "yelling" (nobody liked it, regardless of whether the sibling with PWS or the parents were doing it), and "attention" (who was getting it).

### BENEFITS...

#### of Having a Sibling with PWS

The benefits most frequently mentioned at all ages had to do with attitudes toward people with disabilities. Many brothers and sisters felt that, as a result of having a sibling with PWS, they have more tolerance, awareness, understanding, compassion, and/or appreciation toward others who are different, and they seemed to feel good about themselves for having these attitudes. (*"She has taught me compassion towards people who don't have all the advantages I do."*--brother, 15, of sister with PWS, 13). Several cited

learning patience, being a more sympathetic person, being able to help others, and learning about the syndrome.

Of the four who felt that there is no positive aspect at all, three were in the youngest age group (8 to 12), where they are "on the front lines," so to speak, living at home and dealing not only with the problems of the syndrome but also with normal sibling rivalry.

Some in the older age groups reported they felt they have matured faster than their peers and are more responsible as a result of having a sibling with PWS. (*"I learned before a lot of my friends did that it's okay to be different and to be my own person."*--sister, 22, with 24-year-old sister with PWS) Gratitude, new perspective, realizing the importance of life, and wanting to live every day intensely were also cited as positive outcomes.

Some had very individual benefits, like the woman whose career choice was influenced--and enhanced--by her experiences with her sister with PWS, and the young man who, when little, had learned not to be afraid of the monsters in the dark because his sister with PWS was not.

### NEGATIVES...

#### of Having a Sibling with PWS

Yes, there are plenty of those!! Topping the list for the younger children were yelling and having to keep food locked up. In addition, they said that the child with PWS asks too many questions, is annoying, has tantrums, fights, gets too much attention, is bossy or mean, complains, doesn't listen, chooses the biggest piece of food, and/or gets into and takes the belongings of the other! Not being able to eat candy in front of the child with PWS and having to give in too much were also mentioned.

Not surprisingly, those age 13 and up commented on many of the same negatives, especially the locks--on valuables as well as on food--and temper tantrums. By the teens, some recognized perseverative behavior, the fact that it's impossible to live like other families, and

the uncertainty of the future. A couple of teens in two-child families grieved the absence of a little brother to goof off with or a big brother to set an example. One man noted his own regret in adulthood at having no "normal" sister as well as the distress of his parents. A few commented on stress put on the parents' relationship.

In all age groups, there was concern about others making fun of the sibling with PWS and the need to stand up for them, problems of embarrassment relating to their friends' reactions, and having to watch the sibling with PWS when the parents weren't present--an even greater problem when the sibling with PWS is the older one.

Those 20 years and older expressed some feelings common to many siblings of disabled persons--feelings that are normal, although neither logical nor justified. One is guilty for being "normal" when the sibling is not and even feeling somehow to blame (*"Part of me has always wondered if I were somehow responsible and why it happened to her and not me. I always gave up a lot to keep her happy."*--sister, 31, of sister with PWS, 38). Another common tendency is desiring attention--then feeling selfish for wanting it.

A problem experienced by some was the lack of a diagnosis during the growing up years. (*"The biggest negative is that no one knew what was wrong with her--only that something was wrong. As a child, I could never understand why she wasn't the kind of sister everyone else seemed to have ... She was not my friend--she was my problem; yet I defended her to the hilt if she was ever threatened in any way."*--sister, 33, of 30-year-old sibling)

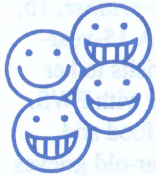
### WHAT ABOUT PARENTS'... Handling of the Situation?

There was a relatively high level of satisfaction with the way the parents handle(d) things. A number said there was nothing they disliked, and a dozen would make no changes at all.

What do they like? At least eight, at all age levels, mentioned in some way their approval of parents treating all the children equally and making the child with PWS an



The following is excerpted from a letter from Mark Berry, which came in response to the sibling survey and is published with his permission. Mark is a 23-year-old man with PWS who wanted to express some feelings and to offer advice to others with PWS.



"I wished that I was normal like everyone else because I hate having Prader-Willi syndrome, especially when I do bad things that I know are wrong like stealing other people's belongings ... [and] destroying things ... dumb stuff, you know. And the bad thing about it is no matter how hard I try to get away with doing something I know that is wrong, I eventually get caught some time or another. So it just doesn't pay to get into trouble, and if I just realize that I won't be able to get away with nothing, the quicker I can get my life straightened out. I will be able to live a more enjoyable life. So all you PWs out there, take my advice [and] stay out of trouble. You'll stay out of jail longer, and you'll be able to live a good long enjoyable and happy life, because getting into trouble doesn't pay."

## Extended School Year Continued from p.5

### 3. Areas of Learning

Certain behaviors or skills are thought to be essential to meeting the goals of self-sufficiency. For example, basic self-help skills such as toileting and feeding are essential for minimal independence; developing stable relationships, impulse control, and appropriate peer interactions are necessary for community living. Behaviors or skills that have not been mastered or cannot be generalized--transferred from one situation to another--are more easily lost, and the child is likely to regress if his/her programming is interrupted.

For older children, the focus of a summer program might be self-help skills, especially impulse control and peer interactions. For younger children, the interruption in exposure to reading, math, and other academics could result in regression with limited recoupment (ability to regain lost skills). Many children with PWS are receiving services through a learning disabilities program or cognitive disabilities program. Even with daily exposure and instruction, these children struggle to understand and master grade-level expectations. For such a child to go without instruction for two and one-half months would most likely result in significant regression.

### 4. Capacity of Family to Prevent Regression

A parent or guardian may be unable to maintain a child's skill level during a break in programming because of the complexity of the program, time constraints, lack of expertise, or for other reasons. Some school districts may provide parent training programs or refer parents to support agencies. School districts are free to utilize these resources as long as there is no cost or financial liability to the child's parents or guardians.

Children with PWS often drain parents of the energy and patience that is needed to manage them 24 hours a day. The need for assistance is not a result of our lack of expertise but instead a desperate need for support. It is next to impossible to provide the amount of structure in a home that a school is able to provide. These children thrive on structure and generally do well in the school environment.

## Siblings Continued from p.7

negative effects, such as strain on the family and feelings of guilt for not seeing the sibling more. Two reported better relationships with their siblings in the adult years--one enjoys spending time with a brother, the other being able to "do things like real sisters."

Many of the adult respondents are no longer living at home, and five said they weren't being affected much at present. This is in contrast to the oldest respondent (age 54), whose parents both died in recent years: "It affects my

life. Since then I have had to try to fill in for them at times for my brother with PWS, 29." Others, too, were concerned about responsibility. ("As an adult, I feel obligated to take a greater role in his caregiving; I have accepted the responsibility of caring for him when my parents are no longer able to, which is a lifelong commitment."--sister, 20, with brother with PWS, 23)

A sibling in her 30s, three years older than her sister with PWS, expresses both the love and the burden: "I think I feel more responsible for her than 'normal' siblings

## Tell Us How You Exercise!



For an upcoming issue of *The Gathered View*, we invite those with PWS who have found a regular form of exercise to write and tell us about it--and send a photo of yourself doing the exercise, if possible.

Most Americans find it hard to fit exercise into their daily lives. For people with PWS, it's more difficult to be active, yet it's even more important to do some form of exercise in order to burn calories and stay healthy. Whether you walk, swim, use exercise equipment, climb mountains, or ski down them, your story just might inspire others to work harder at exercising. Tell us about your routine and how you got started, and we'll spread the word!

Parents of small children with PWS are urged to write, too. Tell us what kinds of motor activities your little one likes and how you fit movement and exercise into their day.

Send your exercise stories to the PWSA National Office, 2510 S. Brentwood Blvd., Suite 220, St. Louis, MO 63144.

would feel for one another. The responsibility for her wellbeing is on my shoulders. I do not resent it; I love her deeply, but I often feel the weight of the responsibility."

**Author's Note:** I wish I could write and thank every single person who took the time and trouble to respond. There were even parents and grandparents who sent along notes. Watch *The Gathered View* for a follow-up article with advice on some of our common sibling issues in parenting a child with a disability.--L.E.M.



equal part of the family. Fairness is valued. (*"They always know what to do. They don't treat her like she's different. They treat her like they treat me."*--sister, 12, with sister with PWS, 10) Most children, however, really want to be special in their own family, and several responding siblings admitted they would like more attention for themselves or remembered that they felt the sibling with PWS was getting the lion's share when they were growing up. Some prized special consideration directed to themselves, such as being allowed sweets the sibling with PWS can't have, or the family going out sometimes without that sibling.

Parents' efforts to get information about the syndrome and to impart that information to the other siblings was valued by many. The importance of information about PWS, or the lack of it, seems to increase from the youngest to the oldest age group. Approved also are the parents' efforts to help the sibling with PWS, such as maintaining the diet and controlling food, not giving up, being patient, not yelling. Several in the older age group felt their sibling with PWS had turned out better because of their parents' hard work. Into the teens and beyond came increased understanding by several that their parents had done or were doing the best they could under the circumstance.

What do they dislike about their parents' actions? Unequal treatment, of course. Not enough attention for themselves. Giving in to the sibling with PWS. Impatience. Yelling.

Survey respondents were concerned not only about how they are treated but also with the discipline of the sibling with PWS. Views about discipline varied. Some wanted more strictness and didn't like parents giving in. Others were troubled by yelling and physical punishment. A few were bothered by differences in parental approaches, which in a couple instances had contributed to divorce.

Being given responsibility for the sibling with PWS can be a problem. One boy wishes he would never have to watch his little brother with PWS outside because he likes to run away. Having older siblings watch younger ones can be problematic

even in families with no disabilities, but the difficulties increase even more when it is a younger sibling trying watch out for an older one with PWS. (*"I don't like it when I get told over and over that no matter what whenever I try to tell her she can't have something she'll still yell and scream because I'm her LITTLE sister."*--girl, 11, with sister with PWS, 16)

### HOW IS IT AFFECTING THEM. In the Present?

In the youngest group, some felt it is affecting them negatively, like the 9-year-old who wrote tersely, *"Not good."* In addition to the attention issues mentioned above, several respondents in this age group described how the sibling with PWS interferes with their lives--e.g., temper tantrums disrupting music practice, problems having friends over. Two others, however, felt that it is developing in them qualities which they value, like sympathy, patience, and better understanding of disabilities. Several expressed warm

sibling with PWS affecting them at present. However, several teens, too, are living with the need to stand up for their sibling with PWS when he or she is made fun of. (*"It gives me a basis for my standard lecture when I hear kids call other kids 'retard' or 'retarded' or making fun of the kids in special ed."*--brother, 16, of 6-year-old boy with PWS) A 15-year-old is bothered by nocturnal visits to her room by her 24-year-old sister with PWS, who searches her drawers for food and wakes her up. Another 15-year-old grieves the "loss" of his 13-year-old sister, who recently moved into a group home.

As in the youngest group, a number of teens felt they were probably better people for the experience of growing up with a disabled sibling. (*"Through growing up with a handicapped child in the home, people acquire such traits as patience, understanding, goodwill, and love. All of these qualities aided me to flourish into a finer, well rounded individual."*--brother, 19, of sister with PWS, 23) A couple valued the education it gives them. (*"I consider having a sister with*

### About the Siblings Who Responded

	All Respondents	By Age		
		(8-12)	(13-19)	(20-54)
Total	38	10	12	16
Female	25	7	6	12
Male	13	3	6	4
Same Sex as Sib w/PWS	23	9	6	8
Eldest Child	16	5	4	7
Older than Sib w/PWS	24	7	7	10
From two-child family	12	5	2	5

Three responding siblings were from the same family. Of the 36 siblings with PWS, 22 are female, and 14 are male; their ages range from 5 to 38.

feelings toward their sibling. (*"It isn't affecting me. I really like having her."*--sister, 12, of 10-year-old girl with PWS) An older brother, 11, troubled by friends sometimes making fun of his little brother with PWS, 6, says, "but I love him."

The focus in normal teen years is outside the home, which may account for the relative mildness of how they saw their

PWS a definite asset. *It's a constant learning experience."*--sister, 19, with sister with PWS, 13)

In the 20-and-older group, a few commented on good qualities they have developed like compassion and a greater ability to deal with people. A few felt

Continued on p.8



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*Learning that your infant or young child has Prader-Willi syndrome can turn your world upside down. A letter recently sent to PWSA President Janalee Tomaseski-Heinemann describes eloquently one family's reaction to the news and to our newsletter. We'd like to share parts of that letter with our members.*

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# The Early Years

Dear Janalee:

We have just received our second issue of The Gathered View and felt compelled to put some thoughts down on paper. ... In reading the newsletter, I go through a full range of emotions, from hope to despair, but I read it from cover to cover many times and then make copies for family and friends who have been such a great help to us in the last six months.

Our son, Robbie, was born on April 11, 1993, after a very difficult pregnancy and delivery. The first few weeks were very difficult as Robbie was born with classic PWS symptoms—completely floppy, severe feeding difficulties, undescended testicles, etc. We were fortunate that the pediatrician at his birth recognized the symptoms and suspected Prader-Willi syndrome almost immediately. Robbie's initial chromosome tests came back as normal and although we were all very hopeful, it was very obvious that there was something seriously wrong with our son. DNA studies were done on my husband, myself, and Robbie that diagnosed PWS. I was devastated by the final diagnosis although my husband and other family members had already accepted the situation and felt relieved that we could put a name to the problem.

We brought Robbie home from the hospital when he was eight weeks old with a NG (nasogastric) tube to feed him. It was a very difficult time, trying to encourage him to drink from a bottle but almost always having to use the tube to get nourishment in him. He pulled the tube out after about six weeks and we decided to try him on a bottle alone and see how it went. The first few weeks were exhausting but Robbie got stronger daily and by about four months of age was taking formula from a bottle normally. Now at almost seven months old, he is taking pabulum and formula completely normally and is thriving well. From just under five pounds at birth, he now weighs in at a healthy 14 pounds.

No one in our family was familiar with PWS ... We contacted associations in the U.S., in eastern Canada, and talked to as many families, doctors, and professionals as we could find. Our approach was that the more information we had, the better off ourselves, our families, and Robbie would be. The initial information was very bleak and we couldn't believe the problems we were facing. The more we researched and talked to people, the better

we felt and realized how much we would benefit from people like yourself who had raised a PWS child. ...

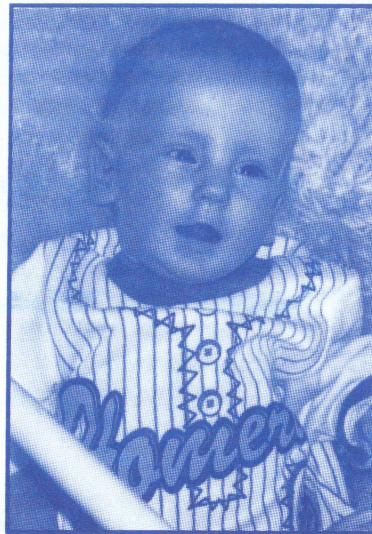
Robbie has grown from a completely unresponsive infant into a very happy little boy who laughs and plays very well. He is getting stronger daily and can now hold his head up unaided and will be sitting on his own before Christmas. He has recently discovered that his feet are actually part of his body and laughs very heartily whenever he is able to get hold of one of them. We enjoy every moment with him and feel very thankful that he is part of our life. A close friend of ours told us to appreciate every little thing that he does because, although his milestones may come a little later than a typical child, they are milestones just the same, and we have taken this advice to heart. ...

We realize that we have many difficult times ahead of us although at this time it seems so impossible as he is the easiest baby to care for. ...

... We appreciate having the advantage of learning from what other families have gone through ... The article about Matt in the recent issue of The Gathered View really upset me when I first read it, but in reading over it and in reading the rest of the newsletter, I realize that although things will be difficult, with planning, organization, and a sense of humour, we will survive the good times and the bad times. ...

We have been fortunate to have friends, family, and professionals provide us with support and information to enable us to deal with a very difficult situation. As with all new parents, we never expected anything like this would happen to us but it did and we have had to learn to handle it, and we appreciate being able to benefit from information and advice from people like you who have been down that road. ...

Yours truly,  
Cindy and Larry Armstrong  
Vancouver, B.C.  
Canada





# Research Projects and Clinical Services

*PWSA (USA) is sometimes asked to publicize to families the availability of medical and professional services either through an ongoing clinic or a specific research project. With this issue of The Gathered View, we are launching a regular column that will provide professionals a means to reach families either to inform them of services or to seek participants for research studies. Announcements for this column should be sent to the PWSA (USA) National Office in St. Louis.*

*We are pleased to publicize these opportunities, and we encourage families to investigate any that might benefit their children and/or the greater understanding of Prader-Willi syndrome. It should be noted, however, that the listing of a research project or clinical service in this column does not indicate that it is the only such service or study available, nor does it imply any sponsorship by PWSA, unless specifically stated.*

## Case Western Reserve: Offers Diagnostic Testing, Seeks Anatomical Donations for Research

**The Center for Human Genetics at Case Western Reserve University in Cleveland, Ohio, now offers complete testing to confirm or refute a diagnosis of Prader-Willi syndrome. For information, contact Dr. Suzanne Cassidy or Dr. Stuart Schwartz at (216) 844-3936.**

Dr. Cassidy also asks that families be reminded of the need for anatomical donations for research in the event of the death of a family member with PWS. Specifically, researchers need to examine fresh brain cells to determine the role of a recently discovered gene called SNRPN, which may have an important role in causing the symptoms of PWS. Families willing to support such research with an anatomical donation should be prepared, when the loved one dies, to have a small piece of brain removed immediately and frozen for delivery to the researchers. This is a difficult action to carry out at the time of a loss but a critical one that may lead to new breakthroughs in PWS research.

Dr. Cassidy should be contacted as quickly as possible after the death and can be reached through Case Western Reserve at any time by calling one of these numbers:

**(216) 844-3936 (days) or (216) 844-1000 (eve/wknds)**

## Dental Study Seeks Participants

As a human geneticist in the Department of Pediatric Dentistry at Eastman Dental Center in Rochester, New York, Dr. Suzanne Hart is interested in the oral manifestations of PWS. While there have been many reports of associated dental abnormalities with PWS, to date no comprehensive study has been conducted. By clarifying dental characteristics of PW individuals, we hope to better prescribe their dental management.

Dr. Hart has designed a questionnaire to help identify the incidence of various dental findings, including caries, gum disease, and problems in salivation. To date, 49 questionnaires have been returned. Most parents reported that it took 5-10 minutes to complete.

In addition to the questionnaire, Dr. Hart has an ongoing saliva study to evaluate the usefulness of saliva as a diagnostic indicator of PWS (Stephenson, *American Journal of Diseases in Children* 146:151-152, 1992). If individuals are interested in participating in this study but are unable to travel to Rochester, it may be possible for her to come to them.

In association with Dr. Tim Wright of the University of North Carolina, Chapel Hill, Dr. Hart is conducting a tooth study to characterize the enamel defects frequently observed in PWS. For this study, they are requesting exfoliated teeth (either baby or adult teeth) from individuals with PWS and, if possible, their siblings. The teeth will be sectioned and examined using an

electron microscope. Parents can also participate by having copies of their children's dental x-rays forwarded to Dr. Hart. Measurements of the various parts of the tooth can be obtained from the x-rays.

If you are interested in obtaining a copy of the questionnaire, sending material (teeth and/or x-rays), or learning more about these research studies, please contact:

**Dr. Suzanne Hart  
Department of Pediatric Dentistry  
Eastman Dental Center  
625 Elmwood Avenue  
Rochester, NY 14620  
716-275-0945  
(collect calls accepted)**



## **Ninth Annual Scientific Day on Prader-Willi Syndrome**

### **CALL FOR PAPERS**

The PWSA (USA) National Office is now sending out information and the annual call for papers concerning the Ninth Annual Scientific Day, which will take place Wednesday, July 20, 1993, in Atlanta, Georgia, immediately preceding the national conference. The Scientific Day is devoted to the presentation of new research results on Prader-Willi syndrome and is open to all interested professionals.

**If you know of any professionals who should be added to the mailing list for Scientific Day information, please call the National Office: 1-800-926-4797**

### **Australasian Prader-Willi Syndrome Conference**

**South Australia  
October 1st & 2nd, 1994**

#### ***FOCUSING ON FAMILIES***

**Enquiries to: Mrs. E.J. Matthews, PO Box 529, Virginia, South Australia 5120. Phone & Fax #: (08) 280 8044 or Sharon Freer, Program Coordinator (088) 373201**

### **PWSA of Pennsylvania Conference**

**Saturday, April 9 1994  
8:30 am - 4:00 pm  
Woods Services, Langhorm, PA**

***Alternative Living Options  
for Individuals with PWS***

**For information call: Deborah Demko, 717-624-2977**

*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: 2510 S. Brentwood Blvd., Suite 220, St. Louis, MO 63144-2326. Any questions? Call us at: 800-926-4797 or 314-962-7644 or FAX 314-962-7869.

### **Prader-Willi Syndrome Association (USA)**

**2510 S. Brentwood Blvd., Suite 220  
St. Louis, MO 63144-2326**

**Address Correction Requested**



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# Wisconsin Seeks Participants for Growth Hormone Study

Geneticist Dr. Richard Pauli and pediatric endocrinologist Dr. David Allen of the University of Wisconsin Hospital and Clinics in Madison, are interested in studying the benefits of using growth hormone in children and young adults with PWS. They are specifically interested in the degree to which growth hormone increases muscle mass (which increases strength and activity level), increases metabolism (which allows a higher calorie intake), decreases fat distribution (improving appearance), and prevents osteoporosis. All benefits and side effects would be monitored.

The study would be open to individuals between the ages of 6 and 15 with a genetic confirmation of PWS; the researchers, however, might consider taking participants up to age 18. There are no height requirements, since height is not a focus of the research.

Proposed is a double blind study, in which neither the participants or the researchers are aware of which subjects are receiving the growth hormone. Two-thirds of the participants would initially receive growth hormone injections while one-

third receive a placebo injection of sterile water or saline. *After approximately one year, all participants are guaranteed to receive growth hormone.*

Participants would be required to travel to the clinic in Wisconsin three or four times a year, paying their travel expenses and a \$30 clinic fee each visit. All diagnostic tests, physician fees, and medication costs would be covered by the study.

Since the research will only proceed if there are 30-50 participants, families with any possible interest should contact the Wisconsin chapter immediately. Those who do so will receive more information and are under no obligation to participate in the study.

Send or call in your name, address, telephone number and the age of the family member with PWS to:

**Barb Dorn, PWSA of Wisconsin, Inc., 5403 North Pass, McFarland, WI 53558, Telephone (608) 838-9535.**

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## PWSA (UK) Dental Data

Our British counterpart--Prader-Willi Syndrome Association (UK)--reported in their December newsletter some of the results of a 1992 member survey on dental and medical problems. Based on 222 questionnaires completed by parents or guardians of children with PWS (121 males and 101 females), PWSA (UK) reports the following findings:

**Teeth eruption** - 70 percent of the parents said their child's teeth had come through normally, 26 percent reported that teeth eruption had been slower than normal, and 4 percent did not know or did not answer the question.

**Grinding teeth** - 26 percent of the parents said their child grinds his or her teeth, 68 percent said their child did not, and 6 percent did not know or did not respond.

**Crowded teeth** - 40 percent reported crowded teeth, 56 percent said their child's teeth were not crowded (although some may have had teeth removed previously), and 4 percent did not answer.

**Thick saliva** - 58 percent said their child had thicker than normal saliva, 39 percent did not think their child's saliva was thicker than average, and 3 percent did not respond.

**There were also reports of poor dental hygiene, tooth decay, poor tooth enamel, worn down teeth, discoloration, and need for bracing. However, some parents reported their child's teeth were in good condition.**