The Gathered View
National Newsletter of the Prader-Willi Syndrome Association (USA)
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Chapter News

Legal Protections Won in Connecticut!

by Don Goranson, Vice President, PWSA (USA)

Members of the Connecticut Chapter of PWSA (USA) are enjoying a "victory lap" this summer, celebrating a major legislative success that will mandate services for all Connecticut residents who were born with Prader-Willi syndrome—regardless of IQ.

Governor Lowell P. Weicker, Jr., signed the measure into law on June 7, after Connecticut Chapter members and legislative co-sponsors earned nearly unanimous victories in both the Senate and House of Representatives of the Connecticut General Assembly.

The entire process, beginning with an emergency meeting of the Connecticut Chapter on an icy, below-zero Sunday night in February until the day of the governor's actual signing of the proposal into law, took less than four months.

A Single Family Pushed to the Limit

The decision to pursue legislative action was a calculated risk, but one the Connecticut Chapter decided had to be taken. This course of action was a swift response to a family that had come to the end of the road in requests for services, with denials by the Connecticut Department of Mental Retardation (DMR), appeals of DMR decisions, a public hearing, and one final and binding denial by a DMR deputy commissioner—all because of an IQ above the 70-point cutoff.

David and Brenda White moved to North Granby, Connecticut, from Texas several years ago with their son Kevin, now 25, who had been diagnosed as having Prader-Willi syndrome at age 3. All of their efforts to "crack the system" and win a group home placement for their son fell on deaf ears. At the monthly meetings of the Connecticut Chapter, the Whites regularly briefed fellow members on the bureaucratic roadblocks they were encountering.

Connecticut's DMR left the Whites and all families with Prader-Willi children having IQs over 70 with little recourse. The deputy commissioner who presided over the hearing at which the denial of services for Kevin White was appealed became quite specific in refusing to bend the department's regulations. The only way to secure a group home placement for Kevin and anyone else with PWS having an IQ over 70 would be to go to the state Legislature and try to get the criteria changed.

We were up to the challenge! We knew it was wrong to use IQ as the sole criterion. We know there are deficits in adaptive behaviors and social skills. We know persons with Prader-Willi syndrome cannot live independently without supports. We know IQ is a smokescreen with our children. The legislative route was the only remaining avenue to amend our statute.

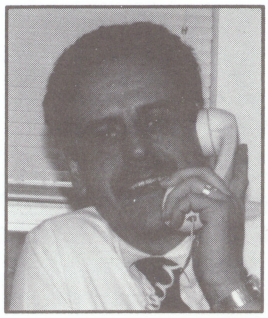
A Tight Deadline, a Simple Strategy

There was very little time. The state Legislature had been in session for about one month. The deadline for filing bills was upon us. Within a short period of time we were able to get a legislator to agree to sponsor a bill that would amend the existing "Act Concerning Services By The Department of Mental Retardation." The plan seemed to be a great one—simply add the following sentence to the existing statute: *"Notwithstanding any statute to the contrary, the commissioner (of DMR) shall also provide services to persons who have been diagnosed as having Prader-Willi syndrome."*

A small group, called together by David and Brenda White and Connecticut Chapter President June Smith and Vice President Bob Cloonan, met in Hartford on Sunday, February 27, to chart strategies. The reason for the rush: The Legislature's Public Health Committee would be conducting a public hearing on this bill in just two days. Speakers were selected, topics chosen, an information sheet prepared, a news release written, and a letter drafted to our membership asking that "urgent" calls be placed to their state representatives and senators. Within two hours, we were as ready and up to the challenge as we would ever be.

Continued on page 12

Inside



Out of the Office

by Russ Myler, Executive Director

I just returned from The ARC of Iowa State Conference where I presented two workshops on PWS. The theme of their conference was "Looking Beyond The Horizon." That theme stuck with me as I drove the eight hours home. I noticed one thing about the horizon; it only stays the same when you are standing still. As long as you keep moving, the horizon keeps changing. I thought about when PWSA first began and what the Association was facing. I can only imagine how different the horizon of PWSA (USA) is now than the one we saw in 1975 when we started our journey.

Earlier diagnosis (the office recently received a call from a parent with a four-day-old infant just diagnosed with PWS), increased frequency of diagnosis, improved treatment, added service resources, and increased support across the nation (and now the world) changes our view and presents us with a new horizon. What an exciting time for us as we move toward our new one.

Today's horizon presents us with challenges we have never had to face before. How do we evaluate, relate to, and make decisions about new developments in medicine and gene therapy? How should we work to influence directions of research and treatment? How do we make sure that we support members with younger children, with older children, with children in placement, and those members whose children have died? How do we make sure we have room for everyone on this journey?

How have our efforts changed the horizon for the persons with the syndrome? What do we do when a 6-foot-2-inch adult person with PWS announces his intention to marry a woman who lives with him in a group home? What roles do parents play in the lives of their adult children when state agencies begin working with their children as adults with full freedom to make major life decisions?

All of these issues and many others are facing us right now. The PWSA (USA) family is looking at a new horizon. As we learned from our past travels, the trip will not be an easy one. To reach our new horizon, we have to keep moving while we do the things that keep us together and strong. We must care for each other, support each other, advocate for each other, and accept new challenges the journey brings.

However, we must also remember that when we reach that horizon, a new one will have taken its place. That is the nature of the horizon.

A Helping Hand...

*PWSA (USA) welcomes **Courtney Trueblood** as part-time office assistant for this summer. She will be a senior at Ladue High School in St. Louis, Missouri. Courtney is helping the office staff with various mailings, filing, and general office duties.*

A special thanks and recognition to the following people for volunteering their time and assistance in the office of PWSA (USA):

Michelle Price ***Cari Stricker***
Dolores Miller ***Jessica Williams***
Cathy Stricker ***Cameron Christy***

A Final Message

by Janalee Tomaseski-Heinemann, May 27, 1994

Dear PWSA (USA) members,

As I told our chapter presidents, three years ago when I started my term as president of PWSA (USA), Bush was president, Los Angeles was fairly intact, Magic Johnson and Michael Jordan were at the peak of their careers—and I felt much younger! In our world of Prader-Willi syndrome, there have also been major changes in the last three years. Although some changes were hard, I hope many of them have been positive. But now my term of presidency is up, and I plan to step down to make way for newer members and fresh ideas. I have been proud, though, to be a team player for PWSA (USA). This is truly a parents' organization and one that has a bright and committed team. In the short run it is easier to carry the ball to glory all alone; however, though a team can slow you down, tackle the wrong person, and sometimes even run the wrong way, it can also go further, be fairer, and pull together to lift you up when you need it.

During the last three years, Al and I also have had many changes on a personal level. Our state chapter took on the challenge of advocating for specialized supportive living homes for all of our young adults with the syndrome. In spite of what seemed like an impossible hurdle, we have seen the opening of four new homes in our state. Our own son, Matt, has been in one of these homes for two years now. As many of our readers know, there have been many steep hills and sharp curves on his roller coaster to adjustment. During this time we held onto the words of parents who had been on this ride before us. They kept saying, "Hang on—he will adjust, even though it may take one to two years." Al and I confessed to each other that we didn't have their confidence. But, fortunately, they were right. Matt is now doing wonderfully in his placement and hasn't had an "incident" in three months.

I'd like to end my term with the national association at the same place where I began—with my personal writing. But first, I want to address a concern voiced by a parent from New York. She was bothered by a recent article I had written, which (I thought) dealt with the humorous side of PWS. She wrote, "... as president of our national organization, your personal problems should be left at home and not broadcast to the world via the organization's national newsletter." From most of the feedback I have received in the past, parents have requested that I continue to write in my own personal style, which includes honesty, humor, and the reality of living with the syndrome. On the other hand, I can see where some of our stories would be disturbing to the parents of a younger child with PWS, and I apologize to them. (It's hard to meet the needs of both the parents of the older individuals with PWS and the parents of the younger ones.)

As president and as a writer, I'm certain I have not done everything "right" or pleasing to all, but I try to remember the statement I read in an Ann Landers column: "Criticism is something you can avoid by saying nothing, doing nothing, and being nothing." So here is my last story, which will probably delight some and offend others.

Celebrating Matt's 21st

This month we celebrated Matt's 21st birthday and took him out to eat at one of his favorite restaurants, The Pasta House. (Note that Matt's weight remains excellent, so we can afford the luxury of approved special treats now and again.) Matt, of course, knew there was a free birthday dessert with the meal at this restaurant, so he had also been there for lunch by himself with the staff's permission. Matt beamed with pride when the servers recognized him and said he was the first person they had known who celebrated both birthday meals at The Pasta House!

Not surprisingly, Matt wanted an alcoholic drink to celebrate this rite of passage to manhood. Also not surprisingly, he wanted the largest, sweetest drink they had—a strawberry margarita. Due to his psychotropic medications, he is not supposed to drink alcohol, so I ordered "virgin" margaritas for Matt and myself and hoped he wouldn't catch on. When we were sipping on the drinks, Matt asked, "Does this have alcohol in it?" I told a white lie (not my first) and said, "a little," which satisfied him.

The special part of the evening for Al and myself was that, for the first time, Matt was eager to leave us to join his staff and roommates (both over 21) at a dance club. In fact, he was so eager to go that he asked for a container for the rest of his pasta, rather than eat it hurriedly. To be expected, he did digress a little while we were driving to the dance club, when he peeked in the container and said, "Maybe I'll eat it at the dance." I told him it probably wouldn't be "cool" to eat it in front of the girls there, to which he said, "Oh, okay." So we dropped Matt off and drove away, knowing we had no place in this part of Matt's world.

Having four other grown children, we can accept and rejoice in the fact that, although we are an important part of their lives, their universe does not revolve around us any more. Although we were the first to help our children stand on their own two feet, they all have different supports to hold onto now. We want for Matt what we want for all our children—that they be sincere, happy, compassionate adults. As we still do, they will work at developing over a lifetime these characteristics that are as elusive and evolving as a butterfly.

Vanja A. Holm, M.D.

The Scientific Advisory Board

A Passing of the Torch ...

Vanja Holm, M.D., has chaired the PWSA Scientific Advisory Board (SAB) since it was established, not long after the Association itself came into being in 1975. In January 1994, Suzanne Cassidy, M.D., was appointed by the board of directors to succeed Dr. Holm as chair of the SAB, and Dr. Holm was named chairperson emeritus. It is with deep gratitude for their many years of work and devotion to PWS that we wish to recognize both these women.

Reminiscence on the Beginnings

by Vanja A. Holm, M.D.

I graduated from medical school, the Karolinska Institute in Stockholm, Sweden, in 1955. Carl and I had married in 1952, when he was working on his Ph.D. thesis in economics in Stockholm. As he had grown up in Seattle that is where we settled. After an internship and residency in pediatrics and a fellowship in developmental pediatrics at the University of Washington, I joined the faculty. I am presently an associate professor of pediatrics at the School of Medicine and coordinator of clinical services for the Child Development and Mental Retardation Center (CDMRC) at our university.

The CDMRC is one of 40-plus University Affiliated Programs in the United States dedicated to training university students from many disciplines in how to provide interdisciplinary services to children with developmental disabilities. Our center opened in 1968 and is one of the biggest and, we think, the best.

My first patient with PWS showed up at our doorsteps at the CDMRC in 1969. He was a svelte 18-month-old who had just recovered from his failure-to-thrive stage. The diagnosis of PWS had been made at 7 months of age when the father was in the Armed Services. The family was directed to contact the CDMRC when they moved to Seattle.

Dr. David W. Smith was a prominent figure in dysmorphology (syndrome recognition) and a member of the pediatric faculty at the University of Washington in the late '60s. As part of the interdisciplinary team assessment, I presented my patient to Dr. Smith in consultation. He agreed with the diagnosis but noted that the boy of course would have to develop obesity for the diagnosis to be confirmed. Death from massive obesity was thought to be the inevitable outcome of this condition. Peggy Pipes, then and now one of our nutritionists, said that she would prevent this by carefully instructing the mother about the caloric needs of a child his age and height. By 2-1/2 years of age he was of course obese. After carefully weighing the boy's food intake, we learned about the reduced caloric need in PWS. Our patient lost weight after adjustments in the nutritional instructions.

As is often done, we presented this information at rounds and meetings. After he learned that patients with PWS could indeed be kept from becoming increasingly overweight, Dr. Smith referred all patients with this syndrome to us for management. He traveled around the Northwest doing dysmorphology consultations, and we quickly accumulated many patients and lots of experience in the nutritional and behavioral management of PWS. Eventually we wrote about the subject in a pediatric journal, describing the management of seven patients.

A local editor, who was available for fine-tuning of papers before they were submitted for publication, was intrigued by PWS and wrote an article about the syndrome and its associated nutritional implications and food-related behaviors in a University of Washington Health Sciences public relations paper. *Seattle Times* picked it up, and the story soon snowballed. By the spring of 1975 most Sunday edition newspapers in the country had run an Associated Press story on the syndrome, and in October it appeared in the medical section of *Newsweek*. We were deluged with phone calls and letters, but the good news is that many new cases were diagnosed as a result of the publicity.

Exactly how the Neasons, one of our families, the Deterlings in Minnesota, and Dr. Sam Beltran in California found each other, I am not sure. All I know is that we met at the CDMRC that year and the PWSA had been conceived. It was nurtured at a meeting in San Francisco a year later hosted by geneticist Bryan Hall, M.D. After a long gestation and much hard work by involved parents—Deterlings, Neasons, Beltrans, Wetts—the first annual meeting was held in Minneapolis in 1979. And, as the saying goes, the rest is history.

I am pleased to have served as a PWSA board member for the first nine years and to have been chair for the Scientific Advisory Board until 1994. Meeting so many persons with PWS and getting to know their families all across this continent and the world has been a most satisfying experience.

continued on page 5

Profile of New SAB Chair

Suzanne B. Cassidy, M.D.

Dr. Cassidy received her M.D. in 1976 from the Vanderbilt University School of Medicine, where she subsequently did a year of genetics fellowship with Dr. Eric Engel. She then did a pediatric residency and medical genetics fellowship at the University of Washington in Seattle. In 1981, she became an assistant professor in the Division of Human Genetics, Department of Pediatrics, at the University of Connecticut School of Medicine, where she became division director in 1984 and associate professor in 1988. In 1988, she moved to the Section of Genetics/ Dysmorphology, Department of Pediatrics, University of Arizona, where she was later promoted to professor. In 1993, she was appointed clinical director of the Center for Human Genetics at Case Western Reserve University, where she is a professor of genetics and pediatrics.

Dr. Cassidy is certified by the American Board of Medical Genetics as a clinical geneticist. She has been a member of the American Society of Human Genetics since 1979 and is a founding member of the American College of Medical Genetics. She is currently a member of the board of directors of the American Society of Human Genetics, is on the original Residency Review Committee for Medical Genetics of the Accreditation Council for Graduate Medical Evaluation, and is on the March of Dimes Clinical Genetics Conference Advisory Board. In addition, she has served on the PWSA (USA) board of directors since 1985, chaired the Scientific Day preceding the PWSA national conference, and became a member of the Scientific Advisory Board in 1992.

Dr. Cassidy describes her research interests as follows:

"Clinical genetics research has been one of the fundamental approaches to determining the manifestations, natural history, variability, and causes of human genetic disorders. In collaboration with cytogeneticists, molecular geneticists, other clinical geneticists, and clinicians in other areas of medicine, I conduct clinical research on a wide variety of genetic disorders, with particular emphasis on Prader-Willi syndrome. Our research focuses on better delineating the features and natural history of the condition, determining optimum management of the various problems, and collaborating to advance understanding of its genetic basis.

"Other areas of particular clinical research focus include phenotype-karyotype and phenotype-genotype correlations of congenital heart defects, tuberous sclerosis, and connective tissue disorders. Ethical issues in medical genetics and training in clinical aspects of human genetics are also of interest."

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Reminiscence...

continued from page 4

Some additional personal notes ... An enduring marriage to a most supportive man has sustained me over the years. I also readily admit that I get a charge out of learning from our daughter, who is a pediatrician with specialty competence also in genetics and endocrinology at the Boston Children's Hospital, that she gets consultations about growth hormone treatment in PWS from families who know she is my daughter. Knowledge is not inherited. Maybe attitude is.

The Role of the Scientific Advisory Board

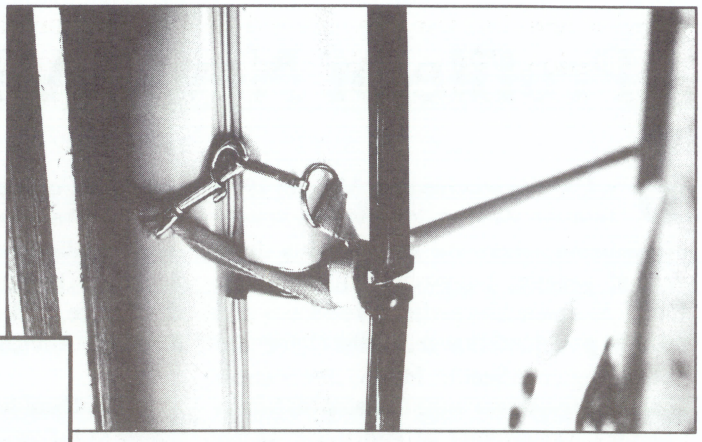
The Scientific Advisory Board has several functions in its role as an advisory body to PWSA (USA):

* to keep the Association informed and current on research on PWS,
* to further knowledge about the syndrome through encouragement of research,
* to review grant proposals to PWSA (USA) requesting Research Funds and to forward its recommendations to the Board of Directors for approval or rejection.
* to serve as expert resources to PWSA (USA) in preparing publications and in answering questions which cannot be answered at the national office, and
* to spread information among parents and professionals about the syndrome

The members of the SAB are highly-qualified professionals from various disciplines who have demonstrated interest in both research and Prader-Willi syndrome. The chair of the SAB has the authority to structure the Board and its membership to best carry out its various responsibilities.

Prader-Willi Syndrome in

Black and White

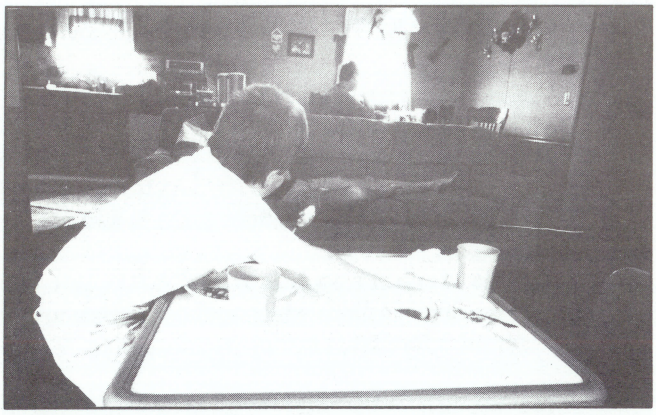


A strap on the refrigerator door is a symbol of a way of life in a family with PWS.

"The first thing you notice when you walk into the Oakleys' kitchen is the refrigerator door; it is strapped shut. At second glance, you see the cupboard is padlocked. There are no fruit bowls, no cookie jars, not even a sugar dispenser in sight. The only time food appears is during strictly scheduled meal times, three times a day."

So began a feature article by Steve Exum in the Moore County (North Carolina) *Citizen News-Record*, which also published some of the pictures (reprinted on these pages) from his award-winning photo story on Steven Oakley, a little boy with Prader-Willi syndrome. While there have been many newspaper articles about PWS, none that we're aware of has captured the syndrome on film the way Exum's did. Here's the story behind the story.

The end of August 1992 found Exum, then a journalism student at the University of North Carolina at Chapel Hill, looking for a subject for the photojournalism competition that he planned to enter. He had heard about Steven and the syndrome and thought it sounded both interesting and unique, in that most people had never heard of PWS. So he dropped by the Neurodevelopmental Nutrition Program (the PWS clinic at UNC) one day when Doris Oakley, president of the PWSA of North Carolina, was there with her son Steven, then 5-1/2.



Thinking no one is watching, Steven steals food from his brother's plate ...

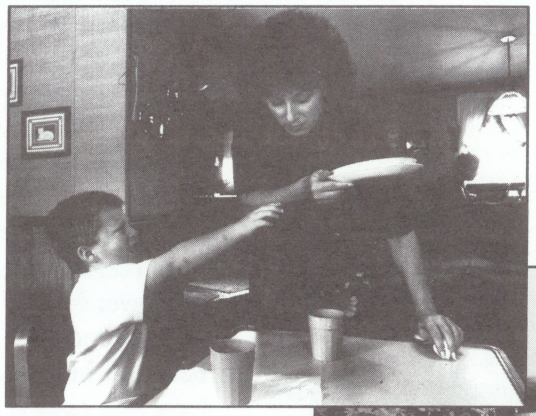
That was the beginning of many, many hours spent by Exum at the Oakley home over the course of an entire year, getting Steven used to his presence so he would be able to get the candid shots he needed. Staging pictures was not permitted. The novelty of a new person wore off, and Steven's concentration went from Exum back to food.

Having completed his photo story in the fall of 1993, Exum entered it in the William Randolph Hearst Photo Journalism Contest, competing against 45 other schools, each of which, like his own U. of N.C., had selected and entered two of its journalism students. Exum's story on Steven—the essence of Prader-Willi syndrome in stark black and white—won first prize in the picture story category and 10th overall.

Steven's History

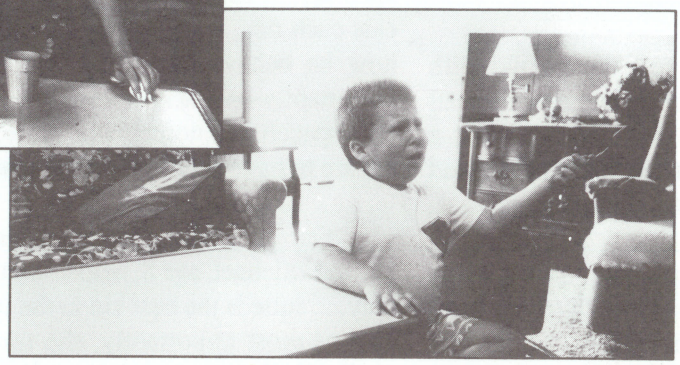
Steven's mother, Doris, had first sensed something was wrong while she was pregnant with him because he did not kick or move like her first child. She asked doctors about this

lack of activity, but they offered no explanation. Four weeks overdue, Doris took the labor-inducing drug Pitocin in the delivery room. The forced labor did not push the infant out; instead, it shut off his oxygen with every contraction. A Caesarian section was done immediately.



Steven was born a "floppy baby." Typical of the syndrome, he could not swallow and showed no appetite. Doris fed him liquids through a gavage tube. It was not until after much uncertainty, more than a year and three pediatricians later, that she finally received the correct diagnosis. Even then, it took pressure on the pediatrician to agree to test for PWS, which Doris had heard about from a nurse who helped her with Steven. The test, showing a deletion on the 15th chromosome, verified her suspicions.

After his second birthday, Steven catapulted into the typical second phase of PWS, with boundless appetite, food stealing, hoarding, bingeing, and weight gain. Doris says that Steven began helping himself to food as soon as he could walk. One night he got up and raided the refrigerator, sucking chocolate syrup right out of the squeeze bottle. So they pushed a chair up against the refrigerator as a temporary blockade—and he simply used it to reach the freezer and get a 4-pound bucket of ice cream, which he was working on when they caught him.



... but he's caught by his mother and denied the rest of his lunch.

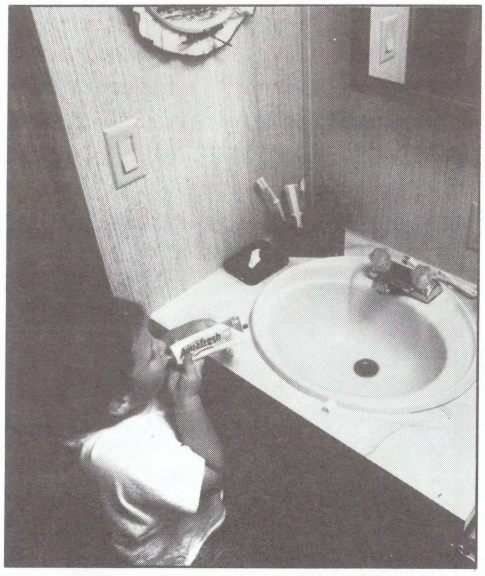
As he's grown older, Steven's temper tantrums and explosive behaviors around food, as well as his obsession with it, have worsened.

Exum captures PWS again, this time in words, in concluding his article:

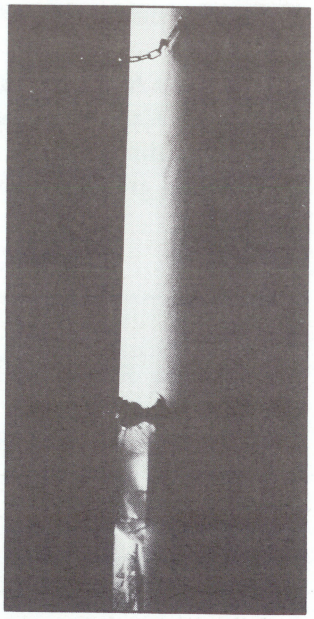
"During a typical lunch, Steven's mother strategically prepares his meal ... Steven sits across from his five-year-old brother Richard. Their mother places identical plates before each boy. Richard is indifferent, almost bothered by the interruption as he sits captivated by a TV game show.

"Steven scrutinizes both plates, comparing them for any possible reason to throw a tantrum. Satisfied, he eats. With a deliberate jerk he stabs a morsel of beef, brings it to his mouth, and chews. He chews without haste, without hurry, with a look of sheer ecstasy. His eyes roll back in his head unconsciously as he relishes each bite.

"It's more than just satisfying, more than giving in to simple hunger. Steven is getting his 'fix.' He's satisfying an uncontrollable addiction until the next craving."



Even toothbrushing provides opportunity—Steven eats toothpaste from the tube.



Bedtime means another lock—on Steven's room—to prevent nighttime foraging.

(Lota Mitchell wrote the above story, based on interviews with both Steve Exum and Doris Oakley. Our thanks to both for permission to use the photographs and retell Steven's story.)

How Lucky To Be a Prader-Willi Parent

by Brian Norton

Do we ever stop and think how lucky we are?

I mean, sure, to have a child with Prader-Willi syndrome—who would call that luck? People must think: What a poor soul, to have raised that handicapped child. What a burden that must be!

But, you know, other people may not ever have the chance to think of someone other than themselves. And, you know, I really think that makes us better people.

Not to say we are better than anyone else. It's just that we have to think of our son or daughter first. Their life depends on us. To control their diet, schooling, social life, health, and well-being.

My son Andrew is now 3 years old. He attends Wentzville special education classes four times a week. How lucky we are to have a great program in our school district.

His teacher understands his way of life very well. She sends a note home each day. It tells of his progress, what he eats each day, what games they played, how he behaved that day, and—most importantly—what new things he can do with speech and motor skills. How lucky we are to have such a caring teacher!

Our other two children, Julie, age 13, and Michael, age 8, help us considerably. ... Julie is the best kid in the world! ... And most importantly, she takes no stuff off of Andrew. Julie makes him behave and treats him like any other child she babysits. And I know she loves him very much. How lucky we are, to have a great kid like Julie to help us raise Andrew.

... Michael is as athletic as they come, and a great soccer player. ... But most importantly, he treats Andrew like a normal brother. They fight like cats and dogs. Running, jumping, screaming, laughing, crying, and fighting—never a dull moment with those two. With Michael around, Andrew has no chance to lay around and become inactive. How lucky we are to have Michael as our son.

And then there is Andrew. How lucky am I to have him. He wakes up every morning about 5 a.m. He stands at his gate (keeping him in his room) and hollers, "Mom, Mom, Dad?" So either Cheri or I get up and get him. How lucky we are to have him hog the covers! And then every morning Andrew gets to pick which cereal he wants. And I can tell you it is never the first box you get down. He also reminds you every day he wants a banana in his cereal. How lucky we are to have him remind us each day.

And when I leave for work, Andrew always tells me "Goodbye, Daddy." The same is true when I come home from work. Andrew is always there to greet me with a big hello, and sometimes a hug and a kiss. How lucky I am to have someone care about me so much.

And, you know, no matter how much his medical bills were, or how much his corrective shoes are, or how hard it is to make sure he only gets a certain amount to eat—nothing makes me feel any luckier than to hear Andrew say: "I LOVE YOU, DADDY."

(Adapted with permission of the author from The Missouri View, newsletter of the Missouri Chapter of PWSA)

'Pull-Ups' Now Made for Big Kids!

In the last *Gathered View*, we listed sources of larger size diapers and absorbent pants for children who had outgrown the products that are readily available in stores. Since then we've learned that the manufacturer of "Pull-Ups" brand training pants has launched a new line of disposable absorbent underpants for children who weigh 45-85 pounds. The new product is called "GoodNites" and is offered in medium (45 to 65 lbs.) and large (over 65 lbs.). If you can't find "GoodNites" in your local stores, you can call Kimberly-Clark Corp. at 1-800-544-1847 weekdays.

The Sibling View

by Lota Mitchell, M.S.W.

In every family with more than one child, sibling relationships are a highly important—and sometimes problematic—part of growing up. For better or worse, brothers and sisters learn from each other in this, their first social network, such interactions as competition, sharing, resolving conflict, who will dominate and who will be submissive, who will take care of and who will be taken care of, and a host of other patterns and behaviors absorbed through time and proximity. Indeed, birth order (whether one is oldest, youngest, or in the middle) is a contributing factor in the development of personality.

When one child has a disability, such as Prader-Willi syndrome, parents are understandably and justifiably concerned about both long-term and short-term effects on their other children. The sibling surveys that were returned to me last fall repeated what research has told us, that there can be positive effects on siblings of a disabled child. They can become more mature, self-confident, independent, responsible, and tolerant; they may develop more patience, more sensitivity and understanding for others with disabilities and their families; and they may even choose to go into, sometimes becoming leaders in, the "helping" professions.

The surveys also demonstrated that there are negative feelings and effects, too, especially when one is dealing with the exceptionally difficult behaviors of PWS. The many surveys that were returned and the strong desire of parents at the national conference in Phoenix to have their children fill them out are evidence of the considerable interest among PWSA members in sibling issues.

The Sibling View will appear in *The Gathered View* from time to time, addressing some of the specific issues that were raised in the surveys. I invite parents, and siblings as well, to write in your comments, questions, observations, examples of what has and hasn't worked for you, and so on, to contribute to this column. There is quite a bit of research on the effects of having a sibling with mental retardation or chronic illness, but almost none on siblings of those with PWS, so your expertise is needed.

For starters, the following are a half-dozen general recommendations for parents on dealing with siblings. These are appropriate for any family and certainly applicable for the family of a child with PWS.

Parenting Tips

1. Don't compare. *Instead focus on what is good and positive about the individual. Comments like "Why can't you behave like Susie?" or "Billy always takes his brother with him!" are definitely not motivating.*

2. Set aside special time for each child. *This is not always easy in today's busy world, but even 15 minutes a day solely devoted to that child can make him or her feel important and secure. The time may be spent playing a game or going someplace together or just talking, but no other child should be allowed to intrude. This may be especially important to the sibling of a child with PWS, who often feels that all the time and attention goes to the one with the disability.*

3. Provide a safe place for belongings. *Separate rooms are best, if that can be managed. In addition, have a cabinet for each child with a lock on it (and several spare keys) where he or she can keep treasured possessions. Sometimes children are expected to share too soon; a child must feel he truly owns something before he can share it. Younger brothers or sisters may break the more fragile things of an older sib, and when the brother or sister has PWS, the need for a secure spot is likely to continue indefinitely.*

4. Set limits. *While families and situations may vary, the two basic rules always are: 1) no hurting and 2) no destruction of property. The rules—these and others in a particular family—should be the same for the nondisabled child(ren) and for the child with PWS. It takes parental energy, but the rules must be enforced—every time.*

5. When it is possible, avoid taking sides with either child. *It is not always possible, and certainly not when one of the above rules is being broken, but if the parent is constantly drawn into sibling conflicts, children learn that superior force decides everything. The weaker one also learns to use his weakness to manipulate Mom or Dad. Nondisabled siblings may be expected to give in to keep (or get back) the peace. Or, less commonly, the child with PWS may become the scapegoat for everything.*

6. When necessary, use graduated discipline. *The first level of discipline is to express disapproval plus understanding. If that does not bring the desired effect, isolate the offender or combatants. Next in the hierarchy is deprivation of privileges. Physical punishment—i.e., a swat on the well-padded bottom—might be used in the early years, but only as a last resort and very sparingly (Many experts advise against spanking at any age.). When the smoke has cleared, a discussion of the feelings behind the misbehavior is important. Humiliation damages self-esteem and should never, ever be used as a means of discipline.*

The next Sibling View will focus on the two-child family. And, don't forget... there will be a special session for siblings of those with PWS at the national conference in Atlanta. Hope to see you there!

Behavioral Techniques for Management of an Adult with Prader-Willi Syndrome Within a Work Program

Fourth in a series of articles on work

by Melanie Grace, B.S., C.R.C.

Adults with Prader-Willi syndrome present unique challenges for staff within a work program when they exhibit explosive behaviors. Most work programs are not equipped to deal with these behaviors, which at times can appear from nowhere.

Several reasons that work programs have difficulty managing behaviors are:

1) The staff are unaware of the behaviors.

This happens for numerous reasons. An important one is that the behaviors may be glossed over because management of the diet is given priority. What they don't understand is how interconnected diet control and behaviors are.

The staff-to-client ratio is also a factor in management. Most work programs have anywhere from a 1:8 to a 1:20 ratio. This ratio makes it difficult to monitor frustration levels that can cause the explosive behavior.

A third reason is lack of education about the syndrome beyond the immediate supervisory staff. Because of the lack of information, the rest of the staff can sometimes escalate behaviors unknowingly.

2) Management of the behaviors requires consistency and teamwork on the part of the staff.

Communication is critical here. Some of the suggested tricks that can be used (And feel free to use them!) are:

a) *Speak in a calm, somewhat slow, quiet voice (Thanks, Anna Marie.).*

b) *Establish eye contact prior to giving directions or when talking a person down.*

c) *If possible, remove the person from the situation; have them take a walk with you.*

d) *As you are walking toward the person, remove items that could be used as a weapon against you or someone else.*

e) *Walk quietly over to the person; don't run or react in an excited manner.*

f) *If someone's behavior does escalate, have a designated staff person move in and control the situation while the rest of the staff maintains a sense of normalcy within the work area (Thanks, Tom S.).*

g) *Have a cool-down area available, where the person can go to calm down.*

h) *Once the person is calm, allow them the choice of going back out to the work area or remaining in the cool-down area to work. This lets them know that they are expected to work but that they do have some choice within the situation.*

i) *If the person becomes unable to work, but can calm down without being destructive, allow them to remain at work.*

j) *If the person becomes destructive, return them to their group or family home.*

k) *Ground rules that have been established prior to the person coming into the program are then put into action, depending on the severity of the outburst.*

Variations of these suggestions can work within any program. One concept that staff need to realize is that these outbursts are a part of the disability and are uncontrollable for the most part.

After an outburst occurs and the person has calmed down, they are usually sorry and promise not to do it again. It's okay to acknowledge that the person is having a tough day. Review what happened, why it happened, and what can be done to help avoid the same situation again. Finally, review what consequences will occur.

Consequences for action is a controversial topic.

There are two schools of thought concerning this. One holds that adults with Prader-Willi syndrome have no control; therefore they shouldn't be held accountable for their actions. The other perception is that, in striving for normalcy, they should have some type of consequences—as would anyone else—for unacceptable behavior. This can be an apology or whatever else has been prearranged so that the person is aware of the consequence. Whatever the program is, it needs to be administered consistently by all staff.

There is no one right way to handle any of the behavioral outbursts that can occur. The trick is found most of the time by trial and error, knowing about your client, and being aware that at times—no matter how well you plan—he or she will be one step ahead of you.

Author's Note:
I'm in the process of trying to put together an informational flyer concerning behavioral tricks that work. Many people involved with the Arizona conference were able to contribute great ideas, but I'm always looking for more. If you have any more ideas and would like to share them, you can send them to me and I will forward the ideas I already have. Thank you.

Melanie Grace, B.S., C.R.C.
119 Brallier Ct.
Pittsburgh, PA 15236

Research Update

Growth Hormone Study in Wisconsin—Update: *Barb Dorn spoke recently with Dr. Pauli about the possible growth hormone study (described in the January-February issue of The Gathered View), and Dr. Pauli shared that the proposal has been submitted to various committees. While he didn't anticipate any problems, Dr. Pauli stressed that there are no guarantees until the proposal has gone through all the committees. Barb Dorn promises to keep us informed about the status of this research proposal.*

* * *

Elisabeth Dykens, Ph.D., of the Yale University Child Study Center, *who surveyed a number of our families at last year's national conference concerning their children's behavior, reported some of her findings at the regional PWS conference held in Albany, N.Y., this past April. Based on 86 completed questionnaires, Dr. Dykens observes that certain characteristics, such as self-esteem and relationships with peers, are "highly correlated with age" in the children studied (ages 4-12) and that all of the behaviors were more commonly reported for boys than for girls. Dykens found no differences in behavior by IQ level, supporting the argument that services need to be provided regardless of IQ, and she reports that there is no clear indication that behavior problems worsen in the adult years—rather, they "wax and wane." Complete results of this study are expected to be published in the future.*

Meanwhile, Dr. Dykens and colleagues have launched a new series of studies related to PWS—on repetitive and other undesirable behaviors, on family stress, and on how people with the syndrome develop an awareness of self and others. For more information or to participate in the new research questionnaires, contact: Elisabeth Dykens, Ph.D., Yale Child Study Center, 230 South Frontage Rd., P.O. Box 207900, New Haven, CT 06520-7900. Telephone (203) 432-2353.

* * *

Suzanne Hart, Ph.D., of Eastman Dental Clinic in Rochester, N.Y., *also reported at the Albany conference findings to date of her dental study (described in the January-February of The Gathered View). Among 49 individuals with PWS on whom she had received surveys, Dr. Hart found that the most common oral/dental characteristics are a high, arched palate (91 percent) and abnormal saliva (87 percent). Also reported by more than 50 percent of survey participants: microdontia (small teeth), crowding of teeth, and enamel defects. No significant difference was found in the extent of caries (cavities) among the subjects with PWS and the control group. (A more extensive report and review of the literature is published by Dr. Hart in the April 1994 issue of Prader-Willi Perspectives.)*

Hart plans to continue the questionnaire study as well as the study of exfoliated teeth (or dental x-rays) and hopes to gain more information on individuals with maternal disomy (Molecular testing may be provided as part of the study.) and on black individuals. In addition, she has received a grant from the National Institute of Dental Health to study saliva flow rates and composition. For more information, contact: Suzanne Hart, Ph.D., Eastman Dental Center, 625 Elmwood Ave., Rochester, N.Y. 14620. Telephone (716) 275-0945.

Spotlight

A Walk With Multiple Benefits

A group of patients with PWS at The Rehabilitation Institute of Pittsburgh took a noteworthy walk around the "TRI block" this spring, Jeanne Hanchett, M.D., informs us. The group was participating in the "Walk for Cure" and raised $302 for breast cancer research through their efforts. According to TRI's newsletter, '*Tute*, participants in the walk included: Michele Noernberg, Ray Holmes, Tim Strawbridge, Susan Measles, Don Carroll, and Jason Speer. Our compliments to all for their effort and achievement. As Dr. Hanchett points out, "It is extra special when persons who must have care themselves can reach out to others."

Workshop...

"Addressing the Residential Needs of Persons with PWS"

Co-Sponsored by:
PWSA (Wisconsin) and Department of Health and Social Services Bureau of Develop. Disab. Services

September 26, 1994
8:00 a.m. - 4:30 p.m.

InnTowner Hotel
Madison, Wisconsin

For more information, call:
Marilyn Neesvig
608-267-7379

Looking Ahead...

New PWSA (USA) logo debuting in our next issue!

Special Thanks to Our Contributors

Memorials received May -June 1994

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Due to limited space, other donations from May-June will be acknowledged in the July-August issue of *The Gathered View*. It is our desire to acknowledge all contributions as an additional expression of our thanks.

Legal Protections, continued from page 1

Early Success, But a Near Loss

March 1, 1994, was public hearing day. Our cause would become sandwiched between dozens of other bills and literally hundreds of speakers. Our time came at about 4 p.m. that day. Fourteen families made time to get to the state Capitol in Hartford. Our eight speakers were ready. The 23-member committe was very attentive. Not only did they listen, they also asked questions. We left the building believing we had done well. About two weeks later the results were known. Our bill not only was approved by the Public Health Committee, but we had won a 23-0 vote of confidence.

Now it was on to the Appropriations Committee to determine financial impact. We had done our homework here also, with assistance from provider organizations. The waiting game was on once again, but we were not prepared for what was to happen. The deadline for action by the Appropriations Committee had passed and our bill didn't make it. We would have to wait another year—unless friendly senators and representatives could get our bill to the floor of the two chambers by attaching it to a bill that did make it through Appropriations. We turned up the political pressure and, believe it or not, were able to have amendments filed under separate House and Senate bills. Now we had two more chances.

The clock was ticking toward adjournment of the Connecticut General Assembly's 1994 session. One of our last two chances failed because it became attached to a budget issue that was not acted upon. But another bill—Senate Bill 362—was approved unanimously by the state Senate and then by a House vote of 145-1. The new statute requires that Connecticut's Department of Mental Retardation provide "comprehensive services" for persons with mental retardation "and, on and after July 1, 1995, persons medically diagnosed as having Prader-Willi syndrome."

The Connecticut Chapter pulled it off! The cause was a worthy one! And we were thrilled!

(Any chapter contemplating similar measures is invited to contact representatives of the Connecticut Chapter. To locate a knowledgable spokesperson or receive copies of testimony or legislation, please contact our PWSA national office in St. Louis.)

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). Annual membership dues: $21 Individual, $26 Family, $31 Agencies/Professionals (U.S. Funds). Send dues, change of address, and letters to: PWSA(USA), 2510 S. Brentwood Blvd., Suite 220, St. Louis, MO 63144-2326. Any questions? Call: 800-926-4797 or 314-962-7644 or FAX 314-962-7869.