



The *Gathered View*

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

A Shot Was Heard Around the World

by Janalee Heinemann, PWSA (USA) Executive Director

Although months ahead of our official "Awareness Week," January and February brought unprecedented publicity to the cause of Prader-Willi syndrome. The tragic death of Christina Corrigan at age 13, and the further tragedy of her mother, Marlene, being tried for her death has triggered the curiosity of reporters all over the world and banded parents together for a common cause. Their rage at the injustice and prejudice they have all faced to varying degrees has been expressed in their response to this case and the publicity it has received.

The following are just a few quotes from the hundreds of letters and calls made on Marlene's behalf:

A mother from New Zealand:

"We parents know the extreme difficulty in keeping food inaccessible to those with Prader-Willi syndrome. We all know the sadness and exhaustion that goes along with caring for the child/adult with PWS. We all know and use locks and keys, padlocks and chains to help keep food out of the reach of the food-seeker, and we all know how difficult it is to explain to others that this is necessary. It is, in fact, the only way to manage this syndrome. If only Mrs. Corrigan had this knowledge and the support of her local PWS association, her daughter would not have died in such ignorant circumstances. It is the neglect of professionals who saw and tended

to her daughter that has in the end caused this unnecessary death. Not neglect by the mother."

A mother from Washington:

"We are judged no matter what we do, and yet it is almost impossible to receive help from the state. We have an almost impossible job of trying to protect our child and living in this constant prison that must be maintained at all costs. People don't understand it unless they live it."

Every article and news coverage piece has brought some level of awareness and assistance that would not have been there without it.

A mother from Australia:

"Look what happened to Christine Corrigan when mother could not devote her attention to Christine's needs because she had to go to work. Gosh, even in Australia we know that accommodation is a serious, in fact critical, need for PWS-affected individuals and their families.

"On the very serious matter of Christine and Marlene Corrigan, I am appalled, shocked, disgusted, with the way the child and the mother have been treated

... where were the welfare workers, social workers, community health services officers when Christine was over 20 stone, and 29 stone, and gaining? Why was she not taken into critical care and treated appropriately? How on earth can they dare to even contemplate punishing the mother when there was an obvious need for critical specialized health care for the girl, and for the mother to be able to get to work?"

*

We are very grateful to our member families who wrote letters on Marlene's behalf to her probation officer and thankful for all of the publicity, which you could title "The Good, the Bad, and the Ugly."

I have had to console more than one parent about what they have read and seen on TV and remind them that in spite of incorrect, incomplete, and inappropriate information, it still gets the word out. Every article and news coverage piece has brought some level of awareness and assistance that would not have been there without it.

The worst "sin" is that of omission — for which the Oprah Winfrey Show gets the award. We heard that she was looking for parents of obese children for a show. David, our office manager, and I both contacted the station, as did many parents. Although they did not choose to respond, I was confident that PWS

(Continued on page 2)

inside

Ohio Conference News	4
Medical Alert on Gastric Problems	6
Behavior: Perseveration, Aggression	12-13
From the Home Front.....	14
A Sibling Gets Published!	15

Special Inside Section:
Making the Case for Food Restriction
PWSA Policy Statement 7-8
Article by E. Dykens et al.9-10

Policy Statement: Adults With Prader-Willi Syndrome and Decisions Regarding Least Restrictive Environment and the Right To Eat — *Continued*

The management of the eating behaviors in persons with Prader-Willi syndrome is based on similar physiologic failures and is equally medically critical. In this instance, there is a genetically based inability to sense satiety, combined with a decreased utilization of calories, resulting in an elevated production of fat tissue. A failure to experience satiety leads the person to continue eating far beyond physiologic or nutritional needs. This overeating combined with elevated fat tissue production leads to rapid and morbid obesity. This rapid obesity overtaxes the heart and leads to complications that can include sleep apnea, diabetes, hypertension, and cardiopulmonary compromise. This physiologically driven eating behavior is no more under cognitive control, nor amenable to cognitive remediation, than is the failure of the pancreas to produce insulin in diabetes. Further, there are, to date, no medical, pharmacologic, or behavioral treatments that fix or cure this biological malfunction. Bioethicists dictate that informed consent requires the capacity to consider, and fully understand, the pros and cons of both sides of a decision issue prior to making a decision. Since by their own physiology, persons with Prader-Willi syndrome cannot decide “not to eat,” therefore they cannot responsibly decide the converse: “to eat, or not to diet.” Thus, to allow such decisions under the guise of “restriction of rights” is both medically and ethically unsound. Failure of the caregiving environment to maintain a rigidly managed diet or to supervise food access leads to the previously described rapid weight gain and can easily result in cardiopulmonary compromise and death. Such a failure in a medical setting would lead to charges of malpractice. Such a failure in a certified living environment may arguably lead to equally serious legal consequences.

Nonetheless, in the past three years, several persons with Prader-Willi syndrome have been placed in less restrictive environments under the argument of “rights.” To date, several have died and the rest have been placed in more restrictive settings or rushed to critical care due to cardiopulmonary crises. Most have gained over 100 pounds in less than six months with the attendant acute medical complications. Clearly, restricting food is not an abrogation of rights; it is the standard of care for a person with Prader-Willi syndrome. Failure to restrict food and allowing a person to eat themselves to death is, in fact, a removal of “rights” to a protected environment.

Is it not a paradox that we would allow someone with Prader-Willi syndrome the “right” to eat themselves to death, but if someone without such cognitive limitations were to threaten suicide, the caregiver that failed to provide suicide restrictions would be found guilty of lack of protection? So we will protect those who are cognitively normal from their own self-destructive impulses, but argue that someone who has cognitive limitations and has physiologically driven eating behaviors has the right to eat themselves to death.

In planning the caregiving environment for persons with Prader-Willi syndrome, some contradictions are evident. While persons with Prader-Willi syndrome need extensive food support, they show fewer needs for support in other aspects of their lives. Indeed, many persons with Prader-Willi syndrome show competencies and decision-making abilities outside the food arena. Nonetheless, until there are medical or pharmacologic interventions for this physiologically driven eating behavior, structured environments with restricted access to, and intake of, food must be standard care for persons with Prader-Willi syndrome.

Some will argue that these recommendations conflict with concerns with choice, personal rights, and least restrictive environment. We do not take issue with these philosophical goals. Instead we assert that the appropriate frame of reference is the “least restrictive environment,” given that the individual has Prader-Willi syndrome. Indeed the concept “least restrictive environment” is meant to imply “as normal a life as possible within the framework of a given disability.” Too often it is translated as: “Despite your disability, you will live as though you are normal.” Society’s efforts to undo a previously created “social disability” may ultimately lead to a completely restricted environment when appropriate limit setting is insufficient. Persons with Prader-Willi syndrome must be uniquely considered as least restrictive goals are put into practice in order to prevent further deaths and to promote a fuller quality of life.

Eating Themselves to Death: Have “Personal Rights” Gone Too Far in Treating People With Prader-Willi Syndrome?

Elisabeth M. Dykens, Barbara J. Goff, Robert M. Hodapp, Lisa Davis, Pablo Devanzo, Fran Moss, Jan Halliday, Bhavik Shah, Mathew State, and Bryan King

This article, first published in the journal Mental Retardation, August 1997, is reprinted by the Prader-Willi Syndrome Association (USA) with permission from the publisher, the American Association on Mental Retardation, Washington, D.C.

In contrast to even a decade ago, giant steps have been made toward increased community inclusion, choice, and self-determination for persons with mental retardation. As workers put these goals into practice, they are sometimes faced with uncomfortable exceptions to the rule: persons for whom increased decision-making leads to unhealthy and even life-threatening consequences. Prader-Willi syndrome is one such exception.

Prader-Willi syndrome is a genetic disorder resulting in mild to moderate levels of mental retardation and distinctive features (Cassidy, 1992; Dykens & Cassidy, 1996; Dykens, Hodapp, Walsh & Nash, 1992a, 1992b). One of the hallmark features of this syndrome is hyperphagia, overeating and marked interests in food (Holm et al., 1993). Contrary to popular belief, hyperphagia in individuals with Prader-Willi syndrome is not due to a weak character or to a lack of willpower around food. Rather, this hyperphagia in individuals with Prader-Willi syndrome stems from altered function of the hypothalamus (Swaab, Purba, & Hofman, 1995), the part of the brain that controls appetite and feelings of satiety. Though hyperphagia's exact cause is unknown, research shows that people with Prader-Willi syndrome do not have normal feelings of fullness and have reduced metabolic rates (Holland, Treasure, Coskeran & Dallow, 1995). These features lead to overeating and to high risks of obesity in affected individuals. Even today, Prader-Willi syndrome remains a life-threatening disorder, with most deaths related to complications of obesity, including cardiopulmonary compromise, hypertension, and Type II diabetes (Hanchett et al., in press).

Although not curable, hyperphagia can be successfully managed through a reduced calorie diet, a regular exercise program, and frequent weigh-ins. Appetite suppressants and behavior modification techniques aimed at reducing overeating have not proven particularly helpful, though pharmacotherapy and behavioral techniques are often successfully used to treat psychiatric and other problems in people with Prader-Willi syndrome (e.g., Dykens et al., 1992a; Dykens, Leckman, & Cassidy, 1996). Widely used approaches for managing hyperphagia in Prader-Willi syndrome rely on external controls, such as locking the refrig-

erator and other food sources; supervising clients in cafeterias, grocery stores, and community settings; and limiting client access to spending money because money is often used to buy food (e.g., Alexander & Greenswag, 1995).

Does it violate the rights of clients with Prader-Willi syndrome to lock up their food and restrict visits to friends, community travel, or spending money, especially given the relatively high mean IQ of 70 in this population? Many states have answered yes to this question. In these states, agencies and group homes that specialize in Prader-Willi syndrome are increasingly criticized as being too restrictive and have been cited for violating client rights. Further, these group home practices are viewed by many states as being overly protective and as denying people with Prader-Willi syndrome opportunities to learn from the “natural consequences” of their behavior (Goff, 1995). Subsequently, program personnel have been ordered to increase client access to food, move clients into less restrictive settings, and give clients control of their diets (Cormier, 1995; Goff, Greenswag et al., 1995).

Unfortunately, the natural consequences that stem from lesser restrictions include increased health risks and premature deaths related to complications of obesity. To date, we know of at least a dozen cases of deaths related to state policies and mandates to relax the food restrictions or living situations of clients with Prader-Willi syndrome. Other natural consequences include job losses due to food-seeking or poor work performance secondary to obesity, repeated arrests for stealing, physical harm while panhandling for food or money, and even trading sexual favors for food (Cormier, 1995; Goff, 1995). Indeed, an individual's drive for food can easily overcome their good judgments about the high risks of stealing, pan-handling, or becoming more obese.

Future tragedies such as premature death or exploitation can be avoided by appreciating certain unequivocal rules in treating hyperphagia in people with Prader-Willi syndrome. One unequivocal rule is that hyperphagia has a physiologic as opposed to a motivational basis. As such, external controls are needed to curb obesity, such as locked food sources and close supervision. Though people can

learn specific food techniques that are helpful to them, interventions should not rely solely on this learning or on strategies such as increasing willpower or self-control around food. A second unequivocal rule is that food restrictions in Prader-Willi syndrome need to be life-long. Hyperphagia does not disappear if people achieve their ideal weight, though the drive for food may wax and wane in any particular individual over time. Long-term dietary restrictions in Prader-Willi syndrome are just as necessary as the life-saving diets used to treat other chronic medical disorders, such as phenylketonuria or diabetes.

Other treatment approaches to hyperphagia in Prader-Willi syndrome are less clear-cut. Specific food practices, for example, often differ between as well as within people over the course of time. One person may tolerate an unsupervised coffee break at work, another may use this same opportunity to steal food. Some individuals do very well helping plan the group home menu; others become overly anxious doing so. One person may readily handle making his bag lunch with minimal supervision, another may not. The same person who successfully bags his lunch may forage through the garbage at work. Food supervision may need to increase in new environments and during transitions or stressful times and return to baseline when activities and places are more routine and familiar. Most professionals readily agree that specific food practices need to be tailored to the changing needs of each individual with Prader-Willi syndrome.

In spite of this agreement, however, the hyperphagia issue remains controversial among professionals, policymakers, families, and clients. This is because many "best practices" in treating Prader-Willi syndrome conflict with mandates for increased choice and personal rights and the movement to place people in least restrictive environments. As workers strive to increase the independence of their clients with Prader-Willi syndrome on the job or in the community, they run into the immediate obstacle—food is everywhere. As people with Prader-Willi syndrome often exploit this fact with remarkable ingenuity, careful planning and coordination are needed across all settings where people live, work, and play.

To date, then, no single or simple solution exists to the autonomy versus food control controversy in Prader-Willi syndrome. Though we do not take issue with ideas of independence or personal rights, persons with Prader-Willi syndrome need to be uniquely considered as workers put these ideas into practice. In particular, we ask professionals and policymakers to keep in mind that hyperphagia in Prader-Willi syndrome has a physiological as opposed to a motiva-

tional basis that results in a life-long need for external food controls. Appreciating these simple facts will go a long way toward meeting our most fundamental goal—the prevention of even more unnecessary deaths.

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Letters to the Editor

A Clarification on "Free Legal Help"

As staff attorney at the National Organization of Social Security Claimants' Representatives (NOSSCR), I read with interest the short article entitled "SSI Alert" in the November 1997 newsletter *The Gathered View* (p. 8). It is very important to inform families that they should appeal the denials or terminations of children's SSI benefits. However, the article is a little misleading. The phone number listed (1-800-772-1213) is the Social Security Administration's toll-free phone line. People can call that number for information about filing for SSI or for other information about Social Security, but it is not the number to call for free legal help. SSA will provide the caller with the number of a local legal services office.

The American Bar Association Children's SSI project has compiled a list of hotline numbers for families to obtain free legal help. As of January 15, there is at least one toll-free number for families to call in 47 states and the District of Columbia. Families can call the specific number for their state for a referral to free legal assistance, including legal services offices and private attorneys who are working for free on these cases.

Barbara Silverstone
NOSSCR
Midland Park, N.J.

Editor's note: Our thanks to Ms. Silverstone for bringing these resources to our attention. The latest available listing by state of these hotline numbers appears to the right. A good Web site for more information on SSI appeals is the Bazelon Center for Mental Health Law: www.bazelon.org.

Children's SSI: Tips on Appeals

As a result of the 1996 welfare reform legislation, more than one-fourth of the children under age 18 who were receiving Supplemental Security Income (SSI) payments became subject to "redetermination" because of a new set of eligibility criteria. Since more than half of 80,000 children with mental retardation have lost their SSI under the new rules, it is likely that some children with rarer conditions such as Prader-Willi syndrome, have been similarly denied SSI in the past year, according to the National Organization for Rare Disorders. Because many families did not appeal the original denial, and because of concerns about the fairness of the decisions, SSA is reviewing some 45,000 cases and is offering a second chance to appeal.

The following tips on SSI appeals are provided by the Parents' Place of Maryland, a federally funded parent information and training center:

- Read all letters from Social Security carefully.
- Go to your local Social Security Administration office or return the information by the requested due date. Promptly deliver or mail all the records showing your child's disability. Provide the SSA with the names and addresses of people who have records documenting your child's disability, even if you don't have copies of your own (e.g., hospitals, doctors, therapists, schools, social or mental health agencies). Make copies of ALL information forwarded to SSA.
- If you miss the 10-day appeal limit, you can still appeal—and continue your child's SSI benefits—if you have a "good cause" reason for missing the deadline.
- Even if you missed the 10-day appeal deadline without good cause, you can still appeal the SSI cutoff within 60 days of receipt of the letter—the benefits will not be continued during the appeal process but would be restored later if the appeal wins.
- If you appeal your child's SSI cutoff, be sure to check the box that says, "I wish to appear at the disability hearing."
- Be sure to ask for a written receipt from your local Social Security office that verifies the day you appealed your child's cutoff from SSI.
- If your child hasn't been to the doctor or therapist recently, be sure to start going again.
- If you do lose SSI benefits, your child may still qualify for Medicaid and other help.

American Bar Association's Children's SSI Project Toll-Free Telephone Numbers for Free Legal Assistance

(Numbers are statewide, unless otherwise noted)

Alabama 1-800-392-5660
Alaska 1-800-478-1234
Arizona 1-800-852-9075
Arkansas 1-800-950-5817
California 1-800-532-2888
Colorado 1-800-955-9173
Connecticut 1-800-453-3320
Delaware 1-800-494-1913 (Kent & Sussex Cos.), 478-8850 (New Castle Co.)
District of Columbia 1-202-824-1869 (English), 1-202-783-9402 (Spanish)
Florida 1-800-694-6361
Georgia 1-888-224-2669
Hawaii 1-808-536-4302, 1-808-528-7046
Idaho 1-800-242-4785, 1-800-632-5125, 1-800-221-3295
Illinois 1-800-471-9300
Indiana 1-800-869-0212
Iowa 1-800-532-1275
Kansas 1-800-723-6953
Kentucky 1-800-200-3633
Louisiana 1-800-922-3425
Maine 1-800-442-4293
Maryland 1-800-236-5641
Massachusetts 1-800-872-9992
Michigan 1-800-288-5923
Minnesota 1-800-292-4152
Mississippi 1-800-682-0047
Missouri 1-800-233-3959
Montana 1-800-666-6899 or 406-543-8343 (Western), 1-800-999-4941 or 406-248-7113 (Eastern)
Nebraska 1-800-742-3005
Nevada 1-800-522-1070, ext. 200
New Hampshire 1-800-562-3994
New Jersey 1-888-576-5529
New Mexico 1-800-524-5176
New York 1-800-724-0490
North Carolina 1-888-756-1986
North Dakota 1-800-932-8880 or 701-255-1506
Oklahoma 1-800-299-3338 (Eastern), 1-800-421-1641 (Western)
Ohio 1-888-601-5437
Oregon 1-800-452-7636 or 684-3763, except Multnomah County: 503-417-0199
Pennsylvania 1-800-598-1989
Rhode Island 1-800-339-7758
South Carolina 1-800-521-9788, ext. 175
Tennessee 1-800-899-6993
Texas 1-888-281-6511, except Houston: 1-888-224-0401
Utah 1-800-662-4245, except Salt Lake City: 328-8891
Vermont 1-800-889-2047
Virginia 1-888-615-2126
Washington 1-800-878-9383
West Virginia 1-800-834-0598

In states not listed, contact local legal services. Even if there is a toll-free legal assistance telephone number for your area, you may still call the Social Security Administration's toll-free telephone number (1-800-772-1213) if you need assistance.

Perseveration and the Child with Prader-Willi Syndrome

by Fiona Whyte, Hon. Speech and Language Advisor to the Prader-Willi Syndrome Association (UK)

(Reprinted from the PWSA-UK News, March 1997, No. 52)

There is at present a debate whether the phenomenon of perseveration is a behavioral problem or a linguistic one. Has the child just not understood the answer, not listened, or is s/he "stuck" on that particular utterance? Is the child seeking reassurance, or is it that s/he cannot move on to the next step in the conversation?

Some parents and teachers feel that it is almost a personal attack when they have answered the same question six, or even 16, times already, and the child repeats it again. Whatever the cause, there are some strategies that are worth trying to see if your child will respond to some, all, or a combination of them.

Some Practical Suggestions

1. **Diversion.** Can you change the subject for him/her? "We've talked about that, let's talk about this now." If the problem the child is experiencing is "topic shifting," this should help.
2. **Reassurance.** It may be that s/he needs the answer confirmed, or to hear an answer more than once before s/he can process it and compare it to the original question. When you have answered again, try confirming that s/he has the answer right by asking, "You know that. I told you. You tell me." In this way, s/he can confirm that the answer is what s/he thought, and also you are indicating that further repetition is not necessary.
3. **"I don't know"** can be an answer too, but some children may have to be taught this concept. They may think that you are just not bothering, or that if they ask again you will know next time.
4. **Wait.** If you cannot answer for some reason, but could find out, put a time limit on. Say, "I'll tell you in 10 minutes," or if s/he can't tell the time, "I'll tell you when the hands on the clock get to ..." and point to the clock, but do make sure you answer then, or this will not work again.

5. **Demonstrating knowledge.** Sometimes the repetition is just to let you know, or to get you to confirm, something s/he is not sure of, e.g.: "It's sewing today." "Is it sewing today?" "It's sewing because it's Thursday." All it should take to solve this one is to say that "Yes, it is Thursday and that means sewing today" — although, as above, this might take more than one exchange.

If this problem is regarded as a linguistic, higher-language difficulty, although diversion and behavioral management may well help to control it,

ultimately the only way to bring about any lasting change in the way the person operates in conversation is to teach the correct, acceptable social exchange patterns, so that a reference is available the next time their linguistic "set" lets them down. It is not enough to explain that it is upsetting to have the same question repeated; if they are stuck on an idea there is no way they can go on to another subject without our help.

There are other techniques for teaching social and conversational skills that could be helpful, but these would have to be specific to the individual's presentation.

When the Neurological Valve Is Stuck Open:

An Analogy and Management Strategy for Perseveration

by Barbara Y. Whitman, Ph.D.

These repetitive verbalizations are perhaps one of the more disconcerting behaviors that persons with Prader-Willi syndrome exhibit. They are disconcerting for several reasons: 1) they are annoying to the listener; 2) they usually occur after the person with PWS has been told "no" about something, so that the verbal repetitions seem to keep alive an issue that the listener would like to drop; and 3) they can, if not sidetracked, escalate to tantrums. Consequently, the issue of how to handle them is not a minor one.

In order to understand how to change these behaviors, let's first understand more clearly what they are. To do that, we must start with what they are not. First and foremost, they are not a psychiatric problem—they are not obsessions, nor are they indications of any deep-seated emotional problem. Secondly, they are not an indication of a rotten kid, or even a rotten personality. However, if not managed well, a rotten personality can result. They are, however, a function of a nervous system whose off/on switch often gets stuck in the "on" position.

To use a mechanical analogy, it's like a shutoff valve that occasionally gets stuck in an "on" position, allowing the liquid inside to keep pouring when you are trying to turn it off. Or, to bring it back to human workings, it is similar to the erratic motor behaviors of a newborn infant whose arms and legs begin to flail at will. The solution for the newborn infant is to wrap the blanket tightly around them so that the flailing stops and their nervous system calms down, a technique known as "bunting." The solution for repetitive verbalizations is a grown-up version of bunting.

**"The
primary
rule ... is
DO NOT
ARGUE."**

(Continued on page 13)

Ask the Parents

Dealing with Aggression

In response to the aunt of someone with PWS who says he is experiencing aggressive behaviors (*Gathered View*, November 1997), I am responding with much compassion because I do realize how difficult life can become.

Our son Paul, age 34, started manifesting aggressive behaviors at the age of 21. The behaviors occurred frequently while he was attending an adult activity center. After seeking out several professionals, I was directed to a psychiatrist, who has been treating Paul for the past 10 years.

To alleviate much of the stress and frustration that some persons with PWS suffer, we have Paul exercise on his stationery bike and walk, which he enjoys tremendously. Exercise is the one activity we stress even to the staff at the group home where Paul has been living for the past four years.

Paul has been on several medications over the years. It is a matter of trial and error to see what medication works best for him. Just a few months ago, Paul needed a change in his medication. Our doctor prescribed "Depakote," which calms the central nervous system along with the medication "Risperdal." By the grace of God, Paul is now much calmer and is able to function daily without being drowsy.

I do hope the above information will help and guide you in the direction you need to take. My prayers are with you.

—Dolores R. Brindisi
Franklinville, NJ

Neurological Valve—Continued from page 12

If we accept that these repetitions are a failure of a neurological off/on switch, it follows that the content of these verbalizations is not the major issue. No matter how much you reason or counter these verbalizations with logical arguments, it will have no impact. Therefore, the first rule of management is: "Don't get hooked into arguing or explaining." If these behaviors have not become a major issue in your family, and you are just beginning to handle them, you can establish a routine of not arguing and stating: "The discussion is now over. If you need to calm down for a while, please go to your room until you are ready to change the subject." Alternatively, for the older child who needs to feel some participation in the decision to stop, you can set a time limit: "We will discuss this only five more minutes, at which time I must do ____." As you say that, note to the person the current time and what the time will be when five minutes are up. At the end of the time, remind them that this is the time to quit and end the discussion. If necessary, send the child to another room or go to one yourself.

Let us not forget that your children are children first and PWS-affected second. So an occasional "because I said so" or "Knock it off!" is not out of line. The primary rule, however, is DO NOT ARGUE. Remember, the content is immaterial once the "neurological valve is stuck open." At that point, you want to verbally and behaviorally "bunt" the verbalizations with structured limits. The more you can present an area of choice within those limits (e.g., "You can have five more minutes now or we can talk about it again for five minutes after supper"), the more you place the decision for stopping with your person with PWS.

For many families, these behaviors have become a major part of family dynamics. For these families, additional behavior management efforts will be needed before the above techniques will be effective.

(Reprinted from *The Gathered View*, January-February 1993, p. 9-10)

The Use of Psychiatric Medications in PWS

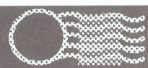
According to a recent article in *Prader-Willi Perspectives* by Dr. Ivy Boyle, a child psychiatrist and the mother of a young boy with PWS, medications "can help take the edge off difficult behaviors" in individuals with PWS and may "allow other, more appropriate behaviors to emerge."

Dr. Boyle reports there has been an "explosion of new psychiatric medications" that are easier to take and often more effective than earlier drugs, but she cautions that there is no way of predicting exactly how a given medication will affect an individual and that some medications can have serious side effects. She describes five classes of medications that may be helpful to people with PWS: antidepressants, antipsychotics, antianxiety agents, mood stabilizers, and psychostimulants. "Your doctor must work closely with you to try to find the best combination of medications and behavior therapy to help the PW patient function most effectively," Dr. Boyle concludes. Her article can help families become more knowledgeable in working with psychiatric professionals.

"Psychiatric Medications in Prader-Willi Syndrome," by Ivy R. Boyle, M.D., *Prader-Willi Perspectives*, Vol. 5, No. 4, July-Sept. 1997. Published by Visible Ink, Incorporated. Phone: 1-800-358-0682. E-mail: perspectives@sprintmail.com.

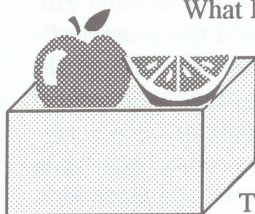
Behavior Support & Solutions

A "functional assessment" of problem behavior is a foundation of behavioral support and is now required by federal law for Individualized Education Plan (IEP) teams addressing behavioral concerns. The objective of functional assessment is not just to define and eliminate undesirable behavior but also to understand the structure and function of behavior and to teach and promote effective alternatives, according to the Beach Center on Families and Disability. The Beach Center has teamed with other organizations to create "The Family Connection," a contact point for sharing of new research on challenging behaviors and strategies for support. For more information, call 1-800-854-4938, or E-mail: dposton@dole.lsi.ukans.edu. The Beach Center's Home Page also has a new section on Positive Behavior Support: <http://www.lsi.ukans.edu/beach/pbs/pbs.htm>



A Snack Box Fills Independence Urge

Dr. Ramasamy Manikam* spoke recently at a New Jersey PWSA meeting. One of the things he mentioned that has worked with some of his patients with PWS is a snack box: You prepare a box of low-calorie snacks and put it in the refrigerator. You tell your child with PWS that this is his/her box. They can take from it whenever they want. Before each meal, you check the box to see what is missing and modify the meal accordingly.



What I have been doing is to only put in the box the snacks that my daughter, Yael, age 10, would ordinarily get in a day.

That way I don't have to modify meals. Her snack box usually includes two baggies of vegetables (carrots, celery, cucumbers, cherry tomatoes, peppers, etc.—whatever is in season) and two other small snacks, such as sugarless jello, a low-calorie yogurt, some pretzels, a piece of fruit, a low-calorie granola bar. There are never more than 150 calories total in the box and usually less. It is working wonderfully. She loves being able to take food whenever she wants, like her brother and sister can do; she likes having her own box that no one else can touch; and she likes being able to choose what she eats and when.

It is usually the first thing she goes to in the morning—she picks out a snack or two to take to school. Usually she takes the most fattening thing (which often is an apple) and leaves the vegetables for her afternoon snack when she comes home from school. Occasionally I will put in a Hershey's Kiss and that usually gets eaten with breakfast. I have found that now that she knows that she can take food whenever she wants, she does not try to sneak cookies but will go to her box and take vegetables.

—Judy Livny
Manalapan, NJ

*Dr. Manikam is assistant professor of psychiatry and behavioral science at Virginia Commonwealth University and previously worked with PWS patients at the Kennedy Krieger Institute in Baltimore.

Happy New Year,

We want to thank you for all the work that you have put into *The Gathered View* this past year.

Our daughter Lauren (PWS) was born April 19, 1997—twin to Shaun, her brother. We have found your newsletters informative and interesting. We also feel as if we belong somewhere—thank you!

—Joanne & Gordon Taylor and Family
British Columbia, Canada



Lauren Taylor at 8 months.

Too Soon To Attend a PWSA Conference?

If you have a young child diagnosed with Prader-Willi syndrome, you may have wondered if you're ready for the experience of going to a national conference—of seeing many children and adults with PWS and perhaps hearing things that you don't want to hear. Of course, each of us must decide for ourselves when we're ready for new information. The following comments from Teri Douglas, the mother of a 2-year-old boy who attended last year's conference in Orlando, might help you make up your mind:

"... all of the sessions I attended were not only insightful and interesting, but enjoyable as well. ... I found myself reliving the anger with some of the 'new' parents, and then finding solace in the strength of those farther down the road. Even though we have read and studied seemingly everything about PWS in the last 1½ years, to see the parents and children and young adults with PWS was definitely a reality check. ... Our fears are great. Still, we gain strength in our faith and, again, from those who are there for us, living those challenges today. ... To see the young adults with PWS out in the varied and challenging surroundings was inspirational. ... I guess if I were to put one word on it, I would say I left the conference with a *bittersweet* feeling. I realized [my son] really is different and special, and will probably always need help. At the same time the love I saw there and the love we have for our children tell us we can survive."

(Excerpted from the *Missouri View*, newsletter of the PWSA Missouri chapter)

The Sibling View

Views from Our Shoes

Growing Up with a Brother or Sister with Special Needs

Edited by Donald Meyer
Woodbine House, 1997
(paperback, 114 pages)

So many wonderful books have been published in recent years to help families cope with a child's disability, but few include a mention of Prader-Willi syndrome. It was a joy then to learn about this wonderful little book for siblings that includes an essay by one of PWSA's own: Lorielle Fiedler of Santa Rosa, California, whose brother Andrew has PWS.

Views from Our Shoes is a collection of 45 short essays by brothers and sisters of children with various disabilities. The authors range in age from 4 to 18, and the essays are arranged in age order so that a reader can quickly find and read the thoughts of a peer. The book was conceived and edited by Don Meyer, who is director of the Sibling Support Project, which operates "Sibshops"—workshops for siblings of kids with



Lorielle Fiedler, top; her brother, Andrew, above, with a birdhouse he and his dad built.

Lorielle's Essay ...

(written two years ago, when she was 10 and her brother, Andrew, was 8)

One good thing about being an older sister of a child with special needs is that I am one of my brother's best friends and he loves me very much. The bad thing about having a brother with special needs is that when he gets mad, he blames me for everything and my parents always believe him. He also learns slower and you have to teach him everything.

Kids at school used to say things about my brother like "he can't run fast" or "he talks funny." We're homeschooled now, and nobody in our neighborhood teases him.

The advice that I would give to a brother or sister of a child with special needs is to be kind, patient, and to teach them everything that you know.

I have noticed that my brother acts differently with different friends. For instance, my brother is obnoxious when my friend Hannah comes over, because Hannah is not very nice to him. But when friends

come over who are nice to my brother, he acts sweet and calm.

Although my brother has problems, he is very good at reading my thoughts! For instance, if I was thinking "hurry up!" he would say "Stop hurrying me!"

My brother shares a lot with me—he likes things to be fair. One time when we went to Toobtown he got a free soda and shared half of it with me—he even got my favorite flavor—rootbeer. He also likes to pretend to be twins with me, which means the same food, same colors, and the same drinks.

Another thing about my brother is that when he has his mind set on something he usually gets it! Once he saved his money for a radio he wanted. After he got the radio, he wanted me to sing with him. I never thought that it would be so much fun! We pretended we were teenage rock stars!

(Reprinted from *Views from Our Shoes* with permission from Woodbine House.)

disabilities—around the United States.

The book's appendix gives brief descriptions of each disability mentioned, addresses for some disability organizations (unfortunately, PWSA was not included), and information on how siblings can get more information both from the Internet and from a sibling newsletter.

Although the book is not all about PWS, there are so many common themes and experiences expressed that any child reading them will

identify with many of the other children and feel less alone. What better gift could you give the sibling of a child with PWS?

Views from Our Shoes can be purchased directly from Woodbine House, 6510 Bells Mill Road, Bethesda, MD 20817, (800) 843-7323. The price is \$14.95, plus \$4.00 shipping and handling. You might also find this book on the shelves at Barnes & Noble bookstores, which feature a "special needs" section and carry many of the Woodbine House books on disability topics.

—Linda Keder

The site is set for the 1999 PWSA (USA) National Conference:

San Diego, California

The Westin Hotel, downtown

July 7-10, 1999

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Prader-Willi syndrome (PWS) is a birth defect first identified in 1956 by Swiss doctors A. Prader, H. Willi, and A. Labhart. There are no known reasons for the genetic accident that causes this lifelong condition which affects appetite, growth, metabolism, cognitive functioning, and behavior. The Prader-Willi Syndrome Association (USA) was organized in 1975 to provide a resource for education and information about PWS and support for families and caregivers. PWSA (USA) is supported solely by memberships and tax-deductible contributions.

