

Our National Office Has Moved!

On February 1, 1993, PWSA (USA) established a new headquarters in St. Paul, Minnesota, requiring a change of mailing address and local telephone numbers. PWSA's 800 number, however, remains the same. The move was necessitated by a 100 percent increase in rent in our former quarters. This would have required monthly payments of \$1,000. Thus the Board decided by mail vote to find a new location.

Announcements have been sent to interested organizations, institutions, publishers, and others. Please help our efforts by notifying those in your area, doctors, libraries, schools. In this way if we have missed anyone in your area, the word will get out.

All PWSA brochures are now being reprinted to show the new address, and old brochures can be updated with new address stickers available from the National Office.

Be sure to update your own records with the new address and telephone numbers.

We Are Here For You

PWSA (USA)

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The Gathered View Becomes One Again

Beginning with this issue of *The Gathered View*, PWSA will return to publishing a single version of its bimonthly newsletter, rather than a separate edition "For the Younger Set."

Since 1985, PWSA has mailed a different version of *The Gathered View* to families of children age 10 and under. The front page feature story for the "Younger Set" version was on a different topic than that in the regular newsletter, while pages 3 through 10 were identical in the two versions. This special edition was created at the time in re-

sponse to parents of young children who said they did not want to read about adult issues they were not yet facing.

The times have changed, it seems, and the pendulum has swung back. Much more information is available on PWS now, and parents of young children have expressed a desire to read everything they can about the syndrome. It also came to the attention of PWSA's Publications Committee that many of our recent front-page stories would be of equal interest to parents of all age groups.

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The information needs of parents dealing with PWS will vary and change as the children grow. Issues that arise during the infant stage are dramatically different from school-age problems, and the difficulties of the teen years are unlike both adult concerns and those of the earlier years. *The Gathered View* will attempt to keep the full range of needs in mind. We invite and welcome your suggestions, comments, complaints, and compliments at any time. (If you're interested in obtaining back issues of either version of *The Gathered View*, call the National Office.)

A Gene for Prader-Willi Syndrome?

Recently, a gene has been found, and localized to the critical region for Prader-Willi syndrome, which may have an important role in causing the symptoms of PWS. The gene is called SNRPN (pronounced "snerp") and codes for small proteins in the brain involved in the structure of the genetic messenger, mRNA. mRNA acts to carry the information contained on the DNA out of the cell nucleus and into the cytoplasm, where proteins are made. Deficiency of this gene (as would occur with deletion or disomy of PWS) would decrease the amount of mRNA and, therefore, of important proteins in the brain.

However, we can't know if this is *really* an important gene in PWS, or study its effects further, unless we have cells from someone with PWS. For this, help is needed from the membership of PWSA.

Although it is a difficult issue to think about, when a person with PWS passes away, it is important that a small, fresh piece of brain be saved *immediately*, frozen, and quickly sent to the researchers who are studying this gene.

For more information, contact PWSA or Dr. Suzanne Cassidy at (602) 626-6000 as quickly as possible after the individual passes away or after obtaining a frozen sample.

References: Ozelik et al. Small nuclear ribonucleoprotein polypeptide N (SNRPN), an expressed gene in the Prader-Willi syndrome critical region. Nature Genetics 2:265-269, 1992.

Helpful Food Labels on the Horizon

The Food and Drug Administration (FDA) published its final rules on nutrition labeling of food products in January, revoking proposed regulations that had gone into effect by default in November of last year. The regulations were mandated by the Nutrition Labeling and Education Act of 1990 and will require a complete overhaul of virtually every food label now on the supermarket shelf by next year.

The nutrition section of the new labels must show, for a specified serving size: the number of total calories and calories from fat; the quantity of total fat, saturated fat, cholesterol, sodium, total carbohydrate, dietary fiber, sugars, and protein; and, for most of these nutrients, as well as for vitamins A and C, calcium, and iron, their corresponding percentage of a recommended daily amount or limit (to be called "Daily Value"), given a 2,000-calorie-a-day diet. The front of the new labels must comply with strict new definitions for use of terms such as "free," "reduced," "low-fat," and "light."

Because of "undue economic hardship" to food manufacturers, the original compliance date for the new labels has been delayed one year, until May 8, 1994. In the meantime, "FDA, USDA, health professional organizations, and the food industry are developing a comprehensive consumer education program that will ease the transition to the revised nutrition label and help consumers to use the label to make well-informed dietary choices," says the FDA. (Final rules appear in the *Federal Register*, Vol. 58, No. 3, Wednesday, Jan. 6, 1993.)

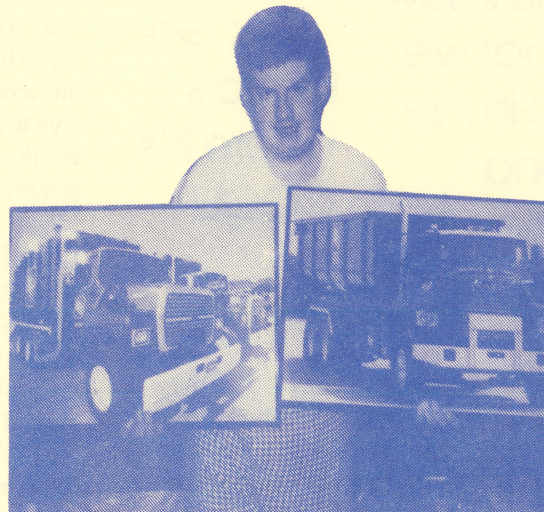
by Janalee Tomaseski-Heinemann

The word perseveration and Prader-Willi syndrome go hand in hand. According to the dictionary, "to perseverate" means "to continue or repeat an action after the stimulus or need for it has passed." Children or adults with PWS (and others with certain forms of brain damage), get started on some pattern of behavior or speech and are not able to stop. I remember our son, Matt, was "stuck" on the topic of the cupcakes he was going to take to school for his birthday. After bringing up the subject for what seemed like every few minutes all day, he said, "Mom, I'm so excited 'bout my cupcakes I can hardly stand it!" I had to sigh, roll my eyes at Al and say, "I know Matt, I can hardly stand it either!"

I think my favorite example of perseveration is the story of Timmy and the trash men. Timmy O'Leary, one of the young men in the newest Missouri PWS group home, has been obsessed with trash collection for several years. His parents state that Timmy begins to get excited about the trash man coming the night before pickup and has a hard time getting to sleep. He immediately wakes to the rumbling sound of a garbage truck coming down the street. Timmy not only watches them pick up the trash, but makes friends with the garbage men, and if there is not enough garbage for them, he will find items to add—such as family clothing and good furniture. Once, Timmy called every

garbage collection agency in the city of St. Louis and ordered pickup for his parents' home. Of course, then he felt a need to have trash for each truck that came, so he started collecting neighbors' cans and putting them in the O'Leary driveway. His parents didn't catch on until they started getting bills from each collection company!

As time went on, Timmy became a unique critic of the trash men's on-the-job performance. He would



personally "hire and fire" workers based on how they were dressed, how hard they worked, and how friendly they were. Becoming well known among the companies, he was responsible last year for seeing that one person actually was fired. Timmy saw him drinking a beer in his truck, and knew immediately that it was AGAINST THE RULES. (People with PWS tend to see rules as black and white—especially for others!)

One of the most exciting days of our lives almost turned into a disaster for Timmy and his parents due to his

perseveration. We had just walked out of the house that Timmy, Matt, and Robert would soon be moving into. It was finally completed and furnished. We all breathed sighs of satisfaction and had smiles on our faces when suddenly, Timmy asked what agency would collect the trash. When he heard their subdivision used BFI, Timmy said "I hate BFI! I had them before and they were no good!!!" Logic was useless in consoling Timmy, and he left insisting that they had to change companies.

According to his mother, Judy, Timmy perseverated for days on how they needed to get a different trash collection agency. Finally, creative desperation led Judy to call BFI and ask if Timmy could come talk to them. BFI not only agreed, but in their attempt to win him over, they gave Timmy a hat, shirt, pen and paper with their logo. The crowning touch was when the owner of the company took his two framed pictures of BFI garbage trucks off the wall and presented them to Timmy! So now when you visit Timmy in his new home, he often has his BFI hat on and proudly shows the pictures on his bedroom wall. He even took his new staff for a tour of BFI, and they reported he was greeted warmly by every worker there and treated like the president of the company. BFI competitors, though, have not given up on winning Timmy over. Since he had given them all his new address, they have reportedly stopped by at his request and given him gifts with their own companies' logos.

Stay tuned for the sequel:

—TIMMY AND THE
TRASH MEN II

Our generous supporters again this year helped us continue to maintain a National Office. We asked in a past *Gathered View* for your continued support for our MBNA credit card fund raiser. I reported only 27 cards had generated \$47.10. The next royalty check we received, jumped to \$1327.30, from 63 card holders. I personally thank each and every one of you who opened an account and continue to use your PWSA MasterCard.

Many thanks, also, for those who sent photos for our publications—they are marvelous!! I look forward to using them in updating our materials, which is the plan for this coming year.

Long Range Planning and Fund Raising were the themes of the mid-year meeting of the Board of Directors and officers. From the hard work and soul searching of that meeting has come a preliminary long range plan for PWSA. Its purpose is to guide the Association's course of action and to determine the resources which are necessary to accomplish what we want to do, what we need to do, and what we are able to do. The next issue of *The Gathered View* will report fully on these plans.

Calendar of Events

If you have a date and event you want posted in the Calendar of Events, please let the National Office know by Friday March 19 to be included in the next issue.

April 30 - May 1 New York-New England Conference, Albany NY

July 14 National Conference Scientific Day, Scottsdale, AZ

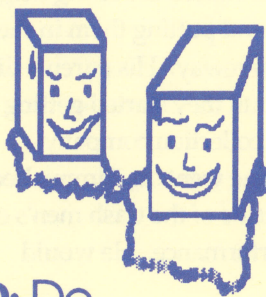
July 14 National Conference Care Providers Day, Scottsdale, AZ

July 14 National Chapter President's Day, Scottsdale, AZ

July 15 - 17 15th Annual PWSA (USA) Conference, Scottsdale, AZ

June 30 - July 3rd, 1994 Second International PWS Conference, Cambridge, England

Hot News from Arizona about the 1993 PWSA Annual Conference



Q: Do you know what we do in Arizona when it gets hot?

A: Go indoors where it's COOL!

Information will be HOT at the National PWSA conference this summer, but what about that other kind of heat? Members from gentler climates seem concerned about July in Arizona. Could anything be hotter than past conferences in Houston or San Diego? Remember, we are people who deal with PWS — you can never say "never" to us!

Our 1993 conference co-chairs, Vanessa Russell and Annette Ruiz, want to assure you that Arizona's summers *are* manageable, assuming you plan your outdoor activities for the right time of day, and that there are, in fact, lots of attractions nearby that might entice you to extend your stay in their fair state.

Fascinating Facts and Vacation Ideas

Did you know that Arizona has more mountain country than Switzerland, more national parks and monuments than California, more sun than

Florida, more golf courses than Scotland, and more forest land than Minnesota? (We didn't think you did.)

Annette and Vanessa suggest just a few of the vacation spots you might like to visit in conjunction with your conference trip: The Grand Canyon is just a four-and-a-half-hour drive from Phoenix. Or you could drive to Nogales, Mexico, to shop for those beautiful Mexican blankets and baskets. If you'd like to improve your odds (and not just abuse your PWSA MasterCard!), Lofflin, Nevada — only four hours away — is a gamblers' mecca. (But please remember PWSA when you hit that jackpot!)

You're encouraged to call Arizona's state tourist office at 1-800-842-8257 for more information on vacation travel and attractions. If you want their comprehensive tourist information packet, you need to allow five weeks for delivery, so don't put off calling too long!

Conference Details

Full details on the conference itself will be arriving in your preregistration packet next month. If you're eager to start your planning, here are a few things you should know:

- The conference will be held at the Registry Resort (1-800-247-9810) in Scottsdale, Arizona, Thursday through Saturday, July 15-17. Pre-conference sessions will take place on Wednesday, July 14.

- A special travel offer has been arranged by an individual member of PWSA, and details will be included in your conference packet. (Please note, however, that PWSA cannot take responsibility for arrangements made with this or any other travel agency.)

- You can arrange child care for evenings out by contacting Vanessa Russell for reservations before June 15. There will be a fee for this special service, but the caregivers have offered to donate 25 cents of each dollar earned to the Deterling Research Fund of PWSA. If you are interested in this service, please call Vanessa Russell directly at 602-945-0512 or write to her at 8708 E. Rancho Vista Dr., Scottsdale, AZ 85251.

Chapter Notes

New York-New England Conference

The Prader-Willi Alliance of New York, Inc., announced its Third Annual Conference to be held April 30 and May 1, 1993, in Albany, N.Y. The conference will be sponsored jointly with the Prader-Willi Syndrome Association of New England.

Featured speakers at the conference will be Dr. Moris Angulo of Winthrop University Hospital in Mineola, N.Y., speaking about the medical management of Prader-Willi syndrome in children and adults; Drs. Ivy and James Boyle of Shaker Heights, Ohio, presenting a parent perspective on the use of growth hormones; Roy L. Smith, Esq., of Stonington, Conn., speaking about estate planning for families with developmentally disabled children; and Karen Levine, Ph.D., and Robert Wharton, M.D., of Spaulding Rehabilitation Hospital in Boston, Mass., speaking on teaching educators about Prader-Willi syndrome. Pat rmier, Program Director of the Gilbough Center in Brewster, Mass., will coordinate a program about residential care, including group homes, their development and management. *For further information, contact: Prader-Willi Alliance of New York, 40 Holly Lane, East Hills, N.Y. 11577. Telephone (516) 621-2445, Fax (516) 484-7154.*



The Rehabilitation Institute of Pittsburgh announces the opening of a Prader-Willi syndrome out-patient clinic. It will be held on the fourth Tuesday of each month with the purpose of confirming or disproving the diagnosis of PWS and providing consultation for the management of progressive obesity and behavior problems. For further information, write or call Jeanne Hanchett, M.D., The Rehabilitation Institute of Pittsburgh, 1603 Northumberland, Pittsburgh, PA 15217, 412-521-9000.

Mental Retardation Redefined

The American Association on Mental Retardation (AAMR) has revised its definition of "mental retardation," eliminating classifications based on level of IQ and focusing instead on an individual's adaptive behavior. The new definition states:

"Mental retardation refers to substantial limitations in present functioning. It is manifested as significantly subaverage intellectual functioning, existing concurrently with related disabilities in two or more of the following applicable adaptive skill areas: communication, self-care, home living, social skills, community use, self-direction, health and safety, functional academics, leisure and work . . ."

The AAMR's definition of mental retardation has been internationally accepted since 1921. This is the ninth revision of the definition, the last having been made in 1983.

To be eligible for services under the new definition, a person must have an IQ below 70-75, with age of onset prior to 18, and have disabilities in at least two of the areas listed above. Along with the new definition, a process is outlined for creating a program of supports, based on the specific functional areas of disability.

The former Association for Retarded Citizens (ARC) participated in the development of AAMR's new definition. In its official newsletter, The ARC explains what will happen once eligibility for services has been established under the new definition:

"The next step in the process is to describe various aspects of the person's condition in order to identify needed supports. Aspects screened are psychological and/or emotional traits, the person's overall physical health and effects of the condition, and the person's current living situation and what would improve the person's life.

"These first two steps lead to

the third step which is to identify the level of support a person needs. Four levels of support are defined:

"Intermittent: does not require constant support, but may need support on a short-term basis for special occurrences, such as help in finding a new job.

"Limited: requires certain supports consistently over time, such as with handling finances, or may need time-limited support for employment training.

"Extensive: needs daily support in some aspects of daily living, such as long-term job support.

"Pervasive: requires constant, high-intensity support for all aspects of life."

Parents are advised to find out from their state service and benefits providers what definition of mental retardation is currently being used for eligibility determinations, whether state officials are aware of the AAMR's revised definition, and, if so, whether they expect any changes in state policies as a result.

According to AAMR Executive Director Doreen Croser, adoption of the revised definition should result in more individualized program planning and "help eliminate rigid and automatic placement based on IQ" as well as preventing the prejudice and erroneous expectations that arise from the use of terms such as "educable" and "trainable".

The new definition is published in a manual entitled *Mental Retardation: Definition, Classification and Systems of Supports, 9th Edition*.

For more information, contact:

**The American Association
on Mental Retardation:
1719 Kalorama Road, N.W.,
Washington, D.C. 20009.
Telephone: 800-424-3688.**

Feeding the Young Child with PWS

by Margie Morris, R.D.

University Hospital, The Milton S. Hershey Medical Center

Mealtimes with babies and young children are usually happy occasions for families. Even with hectic lives, meals tend to be a time during which we relax, converse with each other, teach the young ones table manners and nourish our bodies. Unfortunately, this is not always the case when PWS is involved. For at least two years, and sometimes longer, the simple task of sucking, chewing, and swallowing is hard work for the child as well as the caregiver. The result may be poor weight gain and an enormous "burnout" for moms and dads.

Understanding Poor Feeding Skills

Hypotonia and poor saliva production place children with PWS at high risk for not having a safe, efficient swallow. The neurological process of coordinating breathing, sucking, and swallowing is a complex process involving many areas of the brain. Literature has frequently documented that children who have breathing problems at birth are especially susceptible to this problem. We now are aware that diminished saliva flow may make eating more difficult for these children as well.

Good feeding skills are a prerequisite for speech. The same tongue, lip, and jaw motions necessary to suck from a bottle or to take from a spoon also provide preliminary speech development. It is unlikely that children will demonstrate articulation skills without first demonstrating quality feeding skills.

The young child with PWS may need a "feeding specialist" to help him or her advance to eating more challenging food textures. A physician also may need to be involved, if medication is required to increase saliva flow.

When to Look for Help

Sometimes the signs of poor feeding skills are subtle. Any one or several of the following "red flags" may mean that a child with PWS needs to be evaluated by a trained feeding specialist:

- ◆ excessive drooling
- ◆ coughing while swallowing
- ◆ a change in voice or cry quality
- ◆ slow intake (people normally take half a second or less to move chewed food or liquid from the front of the mouth to the stomach)
- ◆ multiple swallows needed to clear mouth
- ◆ difficulty in advancing to new textures (such as going from baby food green beans to fork-mashed green beans)
- ◆ irritability during feeding
- ◆ excessive wet burps or frequent vomiting
- ◆ frequent respiratory infections, bronchitis, or pneumonia
- ◆ chronic constipation
- ◆ poor weight gain in infancy
- ◆ excessive weight gain from age 3 up attributable to texture sensitivity and, consequently, a limited variety of foods in the diet
- ◆ family fatigue from special food preparation and excessive time spent feeding the child.



Feeding specialists might be found through:

- ◆ early childhood intervention centers
- ◆ Easter Seal programs
- ◆ pediatric rehabilitation facilities
- ◆ pediatric occupational therapists
- ◆ pediatric speech-language pathologists
- ◆ United Cerebral Palsy.

(Editor's note: Margie Morris is a dietician and an affiliate member of PWSA through Penn State University. Her interest and efforts contributed greatly to the formation of The Prader-Willi Syndrome Association of Pennsylvania, and she continues to be a committed member and a vital force in that chapter.)

Research

Two research studies on oral motor deficits in young children with PWS were recently conducted by Margie Morris and the Pediatric Feeding Clinic in the University Hospital Rehabilitation Center. She summarizes some of the results below. For more information, contact Margie Morris, R.N., University Hospital Rehabilitation Center for Children and Adults, P.O. Box 850, Hershey, PA 17033.

Study #1

The University Hospital Rehabilitation Center Pediatric Feeding Clinic in Hershey, Pa., has worked with several children with PWS ages 2-5 years. Frequently these children are overweight, have a prolonged dependency on the baby bottle, and demonstrate minimal chewing skills. We were curious if we were seeing problems typical of this syndrome or if only the more severe cases were coming to us. In fall 1991, 105 families in the United States and Canada received questionnaires from our Feeding Clinic. The response was overwhelming. Sixty-three people answered questions we specifically asked and added additional paragraphs about their child's personal struggle with eating. The chart shows the percentage of children with PWS who are delayed in moving on to specific food textures.

Study #2

At the 1992 Prader-Willi Syndrome Association Annual Conference in Philadelphia, the feeding skills of 11 children with PWS (18-60 months in age) were observed. It appeared that marginal saliva secretion was a major factor in contributing to the delayed chewing skills in 10 cases (91 percent). A moist binding agent (applesauce) added to a challenging texture enabled children to chew half-inch pieces of raw celery, carrots, apple with peel, and chicken.

The recommendation to parents is to add a moist binding agent to foods a child is rejecting. Applesauce, plain unflavored yogurt, low-fat gravy, low-calorie mayonnaise, or low-calorie salad dressing are all possibilities to use without adding excessive calories.

Standard	Age of Child with PWS	Percent Not Meeting Standard
Normal infants take commercial baby food by 6 mo	6 mos. 12 18	50% 10 2
Normal infants eat soft table foods by 12 months	12 mos. 18	33% 10
By age 60 months, children eat any texture:		
meatloaf	60 mos.	8%
raw carrots	"	42
raw celery	"	62
apple with peel	"	41

We conclude that many children with PWS have an extended period of poor eating skills characterized by delay in transition from infant- to adult-textured foods. The survey results suggest the presence of oral sensory deficits and persistence of infantile oral motor skills. Unusual dentition (eruption and/or arrangement of the teeth), poor trunk control, and/or behavioral problems also contribute to oral motor deficits. These problems make it difficult to provide appropriate, nutritionally balanced meals.



A Christmas Letter

Dear Folks,

It's 6:00 a.m., but I just wanted to take this opportunity before I head out for work to write a quick note to you all at this holiday season.

We have an almost 12-year-old son, Ben, with PWS. We received a diagnosis from Dr. [Suzanne] Cassidy during the first annual International PWS Conference in Calgary in 1989. For Ben's first nine years, we had alternately fought with doctors about the possibility that there was something very wrong or that I was just an unfit mother. What a release to finally realize at that conference that we had not lost our entire set of marbles, or that we were not alone! Although experiencing the conference was very difficult, it was also a great relief!

Since that time, we continue to have some very difficult times with some of the medical community regarding Ben's diagnosis. Because his PWS doesn't show up chromosomally (to this date, with the tests that have been done), each appointment seems to begin by our having to verify/defend the diagnosis. And, of course, that means we must educate them on the symptoms of PWS.

One of our visits to the Children's Hospital began with the endocrinologist walking in and telling me that Ben could not possibly be PWS because he had never seen a PWS child with his weight less than in the grossly obese range or this well behaved. He then spent the next five days trying to refute the diagnosis. At the end of numerous appointments with a barrage of specialists, he apologized and told me the diagnosis stands. I told him that it took us nine years to get a diagnosis and, although this is definitely not what we had hoped for Ben, the label opened up a world of hope and help. And, by arbitrarily taking away that label, we have nothing again. Funny that we must



Ben Willms
Age 11 1/2

fight so hard for a "label" and then spend the next how long trying to down-play the same label.

But thank you so much for your wonderful continuing efforts to provide valid, up-to-date information for those of us parents who have, by necessity, had to become lay authorities on PWS. The bimonthly newsletter is always a much-needed breath of fresh air for our family!

For fun I wanted to share a few "vital stats" about Ben: His favorite TV show is *Cookin' USA*. When he grows up, he wants to be a zoo-keeper. When my husband questioned him further, Ben told him that he didn't want to be the person to actually care for the animals, but that he would be the animals' chef in the zoo kitchen, preparing all the food. Ben has learned to count, subtract, and add by the negotiations we have over how many carrot sticks he may have. Ben's first words when he gets in my car after school are: "I'm hungry! That won't fill me up! Can't we stop at a store? I'm tired! Turn the radio up/down! It's too hot! What time is supper? What's for supper? That won't fill me up!"

In spite of the tantrums and stubbornness that are just a part of life at this house, Ben has a marvelous sense of humor (when he is not hungry, tired, or focused on something else)! He has made us all stretch and grow in a thousand ways.

During this holiday season, I wanted especially to thank you for how much your hard work and dedication mean to the average PWS family like us. We seldom write to express this, but during this season of Christ's love to the world, our family says "Thank you, and may the Christ of Christmas bless you all!"

Yours truly,
Donna Willms, Kelowna, BC, Canada

A Bewitching Tale

Pat La Bella reported to her Wisconsin PWSA chapter how she recently visited her daughter's fifth grade classroom to explain what PWS is and to answer any concerns her classmates had. She arrived with a display explaining PWS. But that's not all . . .

To grab the attention of a tough audience for her brief presentation, Pat arrived *wearing a witch's costume!* With the fifth graders captivated by her appearance, Pat explained that, while a costume can easily be removed, there are many things in real life that we cannot change about ourselves. She then led the class in a discussion about how each of us has characteristics and traits that we may not like but can't change. This set the stage for her explanation of PWS and the things over which her daughter has little or no control. The class asked Pat and her daughter a lot of questions — Is PWS contagious? Why do people with PWS gain weight so easily? How much food can they eat? What can classmates do to help their friend?

Kudos to Pat for a bold move that could make a real difference in the lives of a roomful of children!



Q: *How do you handle the constant repetition of questions or requests? Are there any good strategies to head the conversation onto a new track?*

Ask the Parents

A: The most effective way I have found for excessively repetitive questions (and I answer the same one many times before I even notice) is to ask our Hannah (she's 11) what she thinks. She nearly always has a good answer. Then I point out to her that she doesn't need to ask me, because she already knows! Usually, too, this leads her to change the subject. I have to be really quick if I want to introduce a topic of my choice! I guess there is something about Hannah finishing the thought that makes it relatively easy in this situation to move to the next topic.

—Kathy Switzer, Japan

A: I find the most effective method of changing subjects is to merely change the subject. With our son, he is so energetic and has such a short attention span that changing the subject is easy. However, there are many times that, somewhere along the line, he recalls that we never completed the discussion. In this case we agree to discuss the topic again for five minutes, and then the discussion ends and I always let him have the final word.

—Parent of a 7-year-old

A: One response I have come up with for the repetition of questions is to say, "What was the answer I gave you the last time you asked?" If the answer can be repeated, then we know communication has been successful, and I inform my child I will not answer the question again. I further assure communication and acknowledgement of feelings by explaining my anger when questions are asked again and again. I also explain to my child that I understand this is part of the behavior of PWS, and through acknowledging this, there are never any feelings of shame or guilt.

—Parent of a 19-year-old

Ask the Professionals

Since this is such a pervasive problem within the syndrome, we invited a professional to respond to the same question. Barbara Whitman, Ph.D., family therapist, and member of both PWSA's Board of Directors and Scientific Advisory Board, answers.

A: These repetitive verbalizations are perhaps one of the more disconcerting behaviors that persons with Prader-Willi exhibit, particularly as they grow older. 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.

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.

(Continued on the bottom of page 10)

Ten Commandments for Parents of Children with PWS

by Wanda Collins
President, PWSA South
Carolina Chapter

I. Join the National Prader-Willi Syndrome Association. You will need all of the up-to-date information you can get, and National is the place to get it. Then join your state PWSA chapter or support group[if one exists].

II. Join a local support group — if you can't find one, create one. Strength is found within when you offer support to others.

III. Take care of your marriage — it will take both of you working together. Don't place blame! Accentuate the positive, not the negative.

IV. Teach or train your family and anyone who has contact with your child about the syndrome. It will be much easier if all of you are singing off the same sheet of music. The importance of family can never be underestimated. It may take longer for some family members to get out of the denial stage than others. (It is hard to accept the fact that you have a child with a disability.) Be patient and be there for each other.

V. Start a weight management program now. Lock up all food and don't forget pet food! Exercise *with* your child.

VI. When it comes to education, find out what services are available — schools must provide early intervention programs as well as special education until your child reaches the age of 21. Please remember that some of life's most important lessons are not measured on a report card. Stay involved with your child's education!

VII. Grow a hard shell to protect you from the darts others will throw because they think you are a mean mother when you say, "Yes, one little piece of candy *will* hurt." Just remember you are doing what is best for your child. This hard shell also comes in handy when your child uses you as a verbal punching bag and stomps your heart flat. To ease tension, maintain a sense of humor.

VIII. Be as creative in your thinking of alternatives as your child is in his/her food-seeking behaviors.

IX. Pray continuously for guidance and the strength to outlast your child when he/she argues about food.

X. Constantly seek out greenhouses where your child can grow and be nurtured. Self-esteem and coping skills are most important — enough tearing down is done by peers — you must be their cheerleader!

(Continued from page 9)

Let us not forget that our 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.

Next question for Ask the Parents: "What do you do when your child with PWS has destroyed property — either their own or someone else's — in a tantrum?"

Write or phone your responses, questions for parents or professionals, by March 19th to the National Office at 800-926-4797 or write: 1821 University Ave W. #N356, St. Paul, MN 55104-2803.

*Your generous response will really help us fly
and continue to be the angels needed by those
with PWS.*



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(Leo Collins): Beckman, Landeen, Boy Scout Troop 805.

Publications

The *American Journal on Mental Retardation* has published a "Special Issue on the Relation of Communication and Language Development to Mental Retardation" (Vol. 97, No. 2, September 1992), which may be of interest to speech and language professionals and others working with children affected by PWS. Single copies of this bimonthly professional journal may be purchased for \$15 from AAMR, 1719 Kalorama Rd., N.W., Washington, D.C. 20009, toll-free 800-424-3688.

Prader-Willi Perspectives has published its "First Annual Summer Camp Directory" in the January 1993 issue (Vol. 1, No. 2). Data are reported on 37 camps that responded to a nationwide survey of 300 camps and indicated that they either had met in the past, or were able to meet, the unique needs of campers with PWS. *Prader-Willi Perspectives* is a new quarterly journal for professionals and parents that began publication in October 1992. Many PWSA members have received promotional copies of the journal.

For more information, contact: Sheldon Tarakan, Editor and Publisher, Visible Ink, Inc., 40 Holly Lane, Roslyn Heights, N.Y. 11577, toll-free: 800-358-0682.

(For information on camps in your area that may not have responded to the survey for inclusion in this directory, call the PWSA National Office.)

Exceptional Parent's January 1993 issue includes its "Annual Guide to Products & Services." The directory lists sources of everything from augmentative communication to wheelchairs, including computer equipment and software, estate planning, feeding aids, incontinence products, clothing, toys and recreational equipment, schools, camps, and residences. *Exceptional Parent* is published nine times a year, and subscriptions are \$18 for individuals.

For information, contact: Exceptional Parent, P.O. Box 3000, Dept. EP, Denville, N.J. 07834, toll-free 800-247-8080.

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. Duplication of this newsletter for publication is prohibited. Quotations may be used upon credit given to PWSA. Membership dues are \$21 for an individual, \$26 per family, \$31 per agency/professional. Send dues, change of address, or letters to: 1821 University Ave. #N356, St. Paul, MN 55104. Questions or comments regarding this publication or PWS call: 800-926-4797 or 612-641-1955 or Fax 612-641-1952.

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