

The Gathered View

Newsletter of the
Prader-Willi Syndrome Association

JULY-AUGUST 1990

VOLUME XVI, NUMBER 4

Telephone Study Produces Results

Parents of children with PWS, ages 7-11, participated in a telephone study to determine the impact of early diagnosis on current obesity and behaviors. Sixty-seven parents were interviewed. For all patients, 26 months was the median age of onset of obesity and 36 months was the median age at the time of diagnosis. At the time of diagnosis, 60% of the children were obese. Subjects were divided into two groups, those diagnosed early (0-36 months) and those diagnosed late (>37 months). There were no differences between the groups for the variables of sex, current age, weight, and height; nor did the age of onset of obesity differ. Obesity at the time of diagnosis was significantly less common among the early diagnosis group, however.

Parental reports of current cognitive levels were higher for children diagnosed early versus those diagnosed late. In contrast, reports of I.Q.s were less statistically significant. Although most I.Q. scores fell in the retarded range, 10.6% of parents did indicate their child's I.Q. as being "average". Approximately 2/3rds were in special education, 24.6% were in combined programming, and 7.7% were in regular classrooms.

Those reported obese at diagnosis were more likely to be massively obese later. Children who were obese at the time of diagnosis were currently more belligerent about food-related activities than those not obese at diagnosis. Conversely, those who had been obese when diagnosed were less commonly described by their parents as always good natured. Massively obese 7-11 year olds were most often antisocial with their peers.

The median age of onset of unusual behaviors was 48 months. The mean age of onset and the frequency of specific behaviors were not variable with sex of patient.

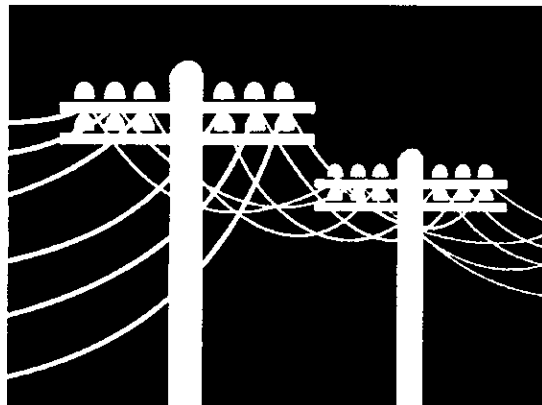
The premise of this study was that early diagnosis might lead to improved weight management and assist a child's adaptation later in life. Until recently, early diagnosis of PWS tended to focus on weight management to prevent the onset or reduce the degree of obesity. While only a few studies document the presence and impact of concurrent emotional liability and behavioral problems, parents repeatedly report that poor psychosocial skills and limited ability to adapt are

major problems that appear to function independently of weight and intensify over time.

The age of onset of obesity did not differ based on whether the children were diagnosed early or late. Children diagnosed early were less likely to be obese at the time, probably because of their younger age, which in some cases was prior to the onset of obesity.

Most of the specific behavior problems identified at a later age were not significantly related to age at diagnosis. Parents reported higher cognitive functioning in the children diagnosed early.

We thank Louise Greenswag, RN, Ph.D. and Randell Alexander, M.D., Ph.D., for their continued interest. The above information was taken from "Early Diagnosis in PWS: Implications for Managing Weight and Behavior," a paper published in the *Dysmorphology and Clinical Genetics Journal*.



Prader-Willi Syndrome Association

6490 Excelsior Blvd., E-102
 St. Louis Pk., MN 55426
 (612) 926-1947
 Office Hours: 8:30 to 4:30

Executive Director: Marge A. Wett

Board of Directors*Chairperson:*

Lota Mitchell, M.S.W., Pittsburgh, PA

Richard J. Wett, M.D., Edina, MN

Mildred Lacy, Louisville, KY

Stewart Maurer, Buffalo Grove, IL

D.J. Miller, Doylestown, PA

June W. Smith, Enfield, CT

Suzanne Cassidy, M.D., Tucson, AZ

Louise Greensawag, Ph.D., Davenport, IA

Janalee Heinemann, M.S.W.,
 Maryland Heights, MO

W. Rhett Eleazer, A.A.L., Columbia, SC

Penelope Park, Oklahoma City, OK

Barbara Whitman, Ph.D., St. Louis, MO

Officers*President:*

Delfin J. Beltran, M.D., Portola Valley, CA

Vice-President:

Stewart Maurer, Buffalo Grove, IL

Secretary:

Fausta Deterling, Orono, MN

Treasurer:

Harvey H. Bush, Vista, CA

Scientific Advisory Committee*Chairperson:*

Vanja A. Holm, M.D., University of Washington

Bryan D. Hall, M.D., University of Kentucky

Peggy Pipes, M.P.H., University of Washington

David Ledbetter, Ph.D., Baylor Molecular Institute

Stephen Sulzbacher, Ph.D.,
 University of Washington

Louise Greensawag, Ph.D., University of Iowa

Barbara Whitman, Ph.D., St. Louis University

Message from the President

Delfin J. Beltran, M.D.

In my last letter I tried to indicate how fear of the unknown and the incompletely understood can cause us to seek explanations for the reasons that our child was born with a disability. This may cause us to seek the "fault" for the occurrence. Thoughts keep filling our mind; "if only I knew why; if I could just understand the reason for my child's problem." The whys and reasons force the concept of fault. "SOMETHING IS AT FAULT!" We may look inside ourselves to find that fault or if that is unacceptable, we may turn against another. Either can result in family dysfunction when this support system is needed the most. Some become paralyzed into inactivity, lost in self-pity, or alienated from the love and support of those who can help. Others become immersed in seeking answers to the unknown and the unknowable. Fear of the future unknown can produce just as great an anxiety and sense of hopelessness. Just remember, it's Sam's fault! Diffuse and defuse. All of that anxiety and wheel-spinning with the unknown will not alter the needs your child has to survive a disability.

The response to such advice at this stage of awareness speaks of seeking help. Okay, so I am *not* supposed to wallow in self-pity. I'm *not* supposed to be fearful of the unknown. I'm *not* supposed to spin my wheels finding fault. What *should* be done? The key step at this point is to become educated in what is known about the problem of Prader-Willi syndrome. That is how our organization came into being. That is how it has grown and developed. The discovery that someone else has had the same problem frequently relieves considerable feelings of personal guilt and inadequacy. Our meetings are marked by sharing. They are further marked by a return to home with a new sense of energy and hope. The knowledge gained by learning from others who have had successes and have survived the problems of daily life with Prader-Willi syndrome, combined with the good fellowship of the PWSA conferences has helped many parents to new success in the caring of their PW person.

Continued on Page Eleven

12th Annual PWSA Conference is Just Around the Corner

The dates: July 18th to 21st, 1990. *The place:* Salt Lake City, Utah. The dates are rapidly approaching (summer is really flying). It's not too late to attend, but the national office would appreciate a phone call from you if you haven't pre-registered, rather than have you just walk in.

Wednesday, July 18th is pre-conference day, an opportunity for researchers to present papers and talk with each other, a chance for the group home people to get together and compare programs and problems, and a time for the Chapter Presidents to meet.

The regular conference starts bright and early on the Thursday, July 19th with a keynote speaker we are certain you will enjoy. Then come three full days of excellent presentations. Our children will enjoy a full program to keep them occupied during the meeting hours.

Come, enjoy, learn. Get recharged for another year.

PWSA Offers a Wide Range of Informational Brochures

If you've seen any of our new brochures you'll recognize the considerable effort expended in their development. Thanks to Janalee Heinemann, Lota Mitchell, Marge Wett, Susie Cassidy, Louise Greenswag and Barbara Carter, these brochures will be of benefit to all of our members.

General Brochure: Description of PWS, PWSA & Application for Membership.

Medical Alert: A diagnosis & reference guide for professionals.

Shelly Brochure: A photo of a young lady who died of complications due to obesity; descriptions of symptoms & characteristics.

Scotty Brochure: A photo of a young man, symptoms, description of our association, and request for donations.

Behavior & Weight Management: Do's & Don'ts in these two very important areas, stages of development.

Education for Parents: What every parent should know about education.

Education for Educators: What educators should know about PWS.

My Child has PWS. Now What?: Designed for those who have just received a diagnosis of PWS or have a young child with PWS.

Late Teens/Young Adults: Guidance in many areas for parents whose children have entered this age group.

We are offering these brochures *without charge in small quantities*. Bulk cost: 51-100 for a \$2 fee; 101-300 at 10 cents each; 301 up at actual cost (cost varies from 8 to 21 cents each). Call for more information.

Ode to a Volunteer

Many will be shocked to find when the day of judgment nears that there's a special place in heaven set aside for volunteers. Furnished with big recliners, satin couches and footstools, where there's no committee chairperson no group leaders or carpools. No eager team that needs a coach no bazaar with a bake sale, there will be nothing to staple not one thing to fold or mail. Telephone lists will be outlawed, but a finger snap will bring cool drinks and gourmet dinners, and rare treats fit for a king. You ask, who'll serve these privileged few and work for all they're worth? Why all those who reaped benefits, and not once volunteered on earth. -- *author unknown*

It's Election Time

I am a pediatrician and medical geneticist who has been interested in PWS since my training with Dr. Vanja Holm in 1980. Since that time, PWS has been the major area in which I have conducted research while an Assistant Professor, then an Associate Professor, at the University of Connecticut and presently at the University of Arizona, in the Department of Pediatrics. I have been a member of the Board of the PWSA for two terms (6 years), and would be interested in continuing to remain on the Board.

I have written a number of review and medical articles about original research of PWS, primarily with regard to the clinical manifestations. I receive more and more telephone calls from all over the world from physicians, other professionals, and parents. I am in the process of planning an international conference on PWS, which will be held in May 1991 in Holland. This is intended to improve communication and collaboration among scientists and to educate people, particularly those in Europe, concerning the various aspects of care.

I feel that it is important to have a member of the Board of Directors who is a physician with major research interest in PWS, adding contacts with the medical community and in the development of educational materials. I would like to continue my involvement and I feel that the best way to accomplish this is by being a member of the Board.



Suzanne B. Cassidy, M.D.

Vacation Resort -- Sounds Ideal!

Platform tents for a family of four rent for \$16 per night or \$100 per week; Cabins for a family of six rent for \$25 per night or \$150 per week; swimming, boating, hiking, arts & crafts, recreational sports are all available for individuals with developmental disabilities and their families.

For further information: Joseph Kuhn, Camp Director, The Bancroft Camp, Lighthouse Rd., Owls Head, ME 04854, (207) 594-5022.

The Importance of Publicity

Over the years the Prader-Willi syndrome has received publicity, mainly in the form of newspaper articles, radio and TV coverage. Naturally, when an article is written by a reporter we do not have control over what ultimately appears in print. We have occasionally been extremely unhappy with articles that exploit the syndrome. Most of the time these articles include a few factual errors, although some accurate information appears as well.

Most of our TV exposure has been on a local level, although we have had a few short segments on national shows. Our most recent exposure was on the Joan Rivers Show of May 29. Featured in the 15-minute segment were PWSA members Henry Singer and his 19-year-old son, Joshua; Sharen and Sheldon Tarakan and their 7-year-old daughter, Emily; and Dr. Jack Sherman of Nassau Medical Center.

Unfortunately, control over what is aired on TV usually presents the same problems as with the print media. On this show the guests were not given the opportunity to mention there was a national organization. Therefore the exposure resulted in only a few calls from people seeking information. However, we did succeed in getting the Prader-Willi name across on national TV.

The Joan Rivers staff contacted Kathy Wyka, one of our members from New Jersey, because of her appearance on another show. Kathy then contacted us and gave us the opportunity to make arrangements with the staff for the people to appear. Here too, decisions are made by them, and in this case they wanted people from the New York area only. If we are fortunate enough in the future to obtain a full hour show on national TV it is very important to have a wide representation of experts appear because even with this advantage some misinformation can still be relayed by the show's moderator. We do hope if any of our members are contacted they will contact the national office so we can do our best to take advantage of this publicity opportunity.

In the meantime, it is important for all of our members to learn more about the syndrome and we feel the 19 Basic Facts from the new *Medical Alert* brochure is a good starting point. You'll find a condensed version of this brochure on the next page.

Medical Alert: Prader-Willi Syndrome

1: PWS is an uncommon non-inherited birth defect, lifelong and life-threatening, affecting all races and both sexes. Prevalence is estimated to be 1:15,000.

2: Characteristics include hypotonia, insatiable appetite, obesity if food intake is uncontrolled, hypogonadism and incomplete sexual development, developmental delays, variable degrees of mental retardation or functional retardation, short stature (adult), small hands and feet, mild dysmorphism, and behavior problems which can be severe.

3: Cause is unclear. Approximately 60-70% have a chromosome 15 deletion (high resolution, prometaphase analysis).

4: Risk of recurrence in a family is estimated to be less than 0.1%.

5: Diagnosis of infants is difficult. (Doctors well acquainted with PWS will not make a diagnosis before the age of 3.)

6: Motor development is delayed.

7: Speech and language problems are common.

8: Average I.Q. is around 70, with a range from 40 to 105.

9: Behavior problems range from stubbornness to violent temper tantrums, increasing with age. True depression and psychotic episodes are reported.

10: Compulsive eating and obsession with food begin usually between the ages of 2-4. Locked food control is essential. (Pica, eating non-food items, is seldom reported.)

11: Obesity occurs in 95% if there are no environmental controls. (Problem is compulsive eating as well as gain on less calories.)

12: Sexual Development is deficient. (underdeveloped genitals, lack of normal menses)

13: Sport activities are limited (poor muscle strength, coordination). Best exercises are walking, swimming, stationary equipment.

14: Dental problems are common.

15: Conditions associated with PWS include strabismus, myopia, scoliosis, and diabetes. (Excessive sleepiness is common, respiratory problems reported, high pain threshold, lack of vomiting, and skin picking are also reported frequently.)

16: Medications so far have not been found generally useful to control appetite or behavior. Effects with PWS are not comparable to normal population. Drug treatment is essential at times but not recommended for continuing usage.

17: Adolescents and adults can function well in group living programs if they have adequate calorie control and structured living. Sheltered workshops generally provide the only employment possibilities.

18: Life expectancy may be normal if weight is controlled.

19: Stress on families is often extreme. The child with PWS can become a tyrant who rules the family and with age become more difficult to handle. Awareness of these stresses and dealing with them early on is essential to the family structure.



Mollie's Story

Mollie has maintained competitive employment since last July. She is 31 years old, and has been in a series of group homes since the age of 11. Two and a half years ago, Mollie became one of the first residents of a new home designed for PW adults and was able to achieve weight and diabetes control for the first time.

Mollie's story continues; she worked eight hours a day in housekeeping with the assistance of a job coach, provided her own transportation by taxi and walking, and at the same time, maintained her weight. After Mollie's hours were reduced, she found a new job by applying to a factory she walked past.

We certainly congratulate Mollie and recognize what she has accomplished – and there are other Mollies out there who have done well too. But as the editor of a newsletter for PWS, I feel I would be lax if I did not do a little editorializing on the subject of competitive employment. It is terrific when we hear about those

who have entered competitive employment and have still managed weight control because that usually is the trade-off – they are capable of doing and holding the job but the place of employment is not capable of keeping food out of their reach. Many very sad letters are received from parents

whose children like their jobs very much but if they are allowed to continue they will die from complications of obesity. Other letters are received, my son has gone from one placement to another, how many more failures is he expected to endure before someone wakes up and does it right. When a competitive placement is done right -- job coach for a reasonable time, not just a day or two; the workplace has been informed exactly what the situation is, not just told this person is retarded but a good worker; sufficient follow-through methods have been put in place, particular-

ly with a weight out and in, then, and only then, can we read success stories such as Mollie's.



SSI in the News

The US Supreme Court ruled on February 20, 1990, that Social Security's policy of not considering the functional limitations of children when determining their eligibility for SSI benefits was inconsistent with law. This decision means that more children will be eligible for benefits. This decision will not change denial based on family income, it applies to those denied because they said they were not disabled.

If you were denied in the past, it is recommended that you reapply. In the case of PWS, we have not heard of anyone who could not qualify, although it may take more than one try to obtain it.

New Group Home has Openings

One home in existence and one new facility in the State of California are reporting openings at this time. If you are looking for an under 18-year-old placement, please contact Dan McHargue at (714)795-8486 or (714)795-7404.

Drugs & Prader-Willi Syndrome

One mother wrote: "I am writing about the use of drugs on PWS -- and warning people about using them. My son was given (at a Developmental Center) 23 electroshock treatments plus a plethora of psychoactive drugs. These included *Mellaril*, *Stelazine*, *Ouide*, *Dalmane*, *Valium*, *Haldol*, *Thorazine*, and *Laxitane*. This was since 1962. None of them ever changed his behavior one bit, except to make him dopey, sleepy, drooling. Now I find he has lost 20 I.Q. points and his classification has been changed from mild to moderate (going from 66 to under 50).

These types of drugs tend to exacerbate the weaknesses of PW people if they get the side effects. They affect coordination, the eyes, the speech. Even short term use can cause permanent damage. Perhaps very short term use for a major upset or if there is a dual diagnosis of an overlaying mental illness such as schizophrenia, would be understandable.

Parents must be very wary of a diagnosis of a mental illness to justify drug use. Now at age 43 my son was recently diagnosed as "Borderline Personality Disorder" and I am fighting that.

His personality or behavior has never changed. He suffered excruciating withdrawal dyskinesia (difficulty in performing voluntary movements) on the abrupt cessation of *Haldol*. Now when he gets upset he is much more violent and self destructive than he ever was in the past, necessitating restraints. He also developed pica (eating non-food items) during the withdrawal dyskinesia and had three major surgeries during 13 months time. During this period his weight went from 160 lbs. to 109 lbs. in 6 weeks. Calories had to be increased to 4-5000 a day during the weight loss, and now that his weight is back up to 155 they again have to go back to a lower level and control." This mother also included a copy of a paper titled *Suggestion*

for *Clinical Research, Compulsive Eating, Medication, and PWS*, by Russell M. Steinberg, Ph.D., M.D., Fairview Developmental Center.

The doctor presents a couple of case histories of two people with PWS, states they are confined to an institution because of their inability to control their eating and their maladaptive behavior. He concludes with if a medication were to be found which is effective in controlling the eating behavior of people afflicted with PWS, the discovery would be one with major humane and economic benefits. He suggests a study with the two drugs *Prozac* (Fluoxetine) and *Anafranil* (Clomipramine).

Prozac is a member of the phenylpropylamide group of non-tricyclic antidepressants. Side effects of nausea, nervousness, headache and insomnia occur more frequently than with some other drugs but other types of side effects are less. A 30-day supply is said to cost about \$40. *Anafranil* is a tricyclic antidepressant which has been available for treatment of depression in countries other than the U.S. for many years. There are more side effects noted with this than with *Prozac*, but Ciba-Geigy is offering this drug

free of cost to psychiatrists for investigational use in treatment of Obsessive-Compulsive Disorders, patients between the ages of 18 and 70.

It is true, a medication to effect control for PWS would be a major breakthrough for our children. However, at the moment there are so many unknowns. Do we have professional members who have extensive experience in working with these problems? The national office would certainly like to hear from anyone so this could be shared with our membership. PWSA has funded an investigational study of drug treatment in PWS. We look forward to being able to share the results of this study when data is available.



"It's long overdue but I'd like to express this parent's great appreciation for our Association and its newsletter. Although there are many reasons, I'd like to share thoughts on a couple articles:

First, Janalee's *Have We Gone Too Far* (November-December 1989): That kind of sharing, especially from a "professional" type mom, provides great help for many of us who experience terrible guilt when there's regression in a PW problem area we thought we'd found solution for or when unexpected behaviors occur. I usually feel at such thankfully rare times that something was wrong with what I did or said, or didn't do or say rather than realizing that the cause of the incident might be beyond anyone's guidance skills. The guilt or resolution needed feelings should be saved for other occurrences, i.e. the ones where rethinking the incident shows that better judgment could have been used or the level of patience wasn't where it has to be. Please let Janalee know that the time she took to share with us is appreciated for it helps to recall at black moments that we're not the only one in the world with an unexpected, impossible problem. Secondly, I'd like to thank the parent who submitted the *Analogy* article (May-June 1990) and the Editor's response! I can't provide "...medical expert comment"...to the parent but would like to share my experience of raising a 32-year-old daughter with whom I dealt daily until she was 28. Holly told me frequently during her school years and later work years that within limits there were things more important to her than food!

On an issue of feelings, on occasions when persons directly afflicted with PW are able to clearly express their opinions I think the opinions should be credited as "expert" answers. Thru the years of trial and error with various rewards to help her weight control, which had gone as high as 170, what she taught me was that first school and then work were more important to her than food. Thus, they at times became rewards and on occasions when I had to follow through and deny her a day at school or work, along with the "I'll never do it again," she'd plead with me to take away TV, weekly social event and not give her any

system was informed on PWS because of a state-wide mailing of information to the school systems, by a working group from that state.)

Another Parent Shares: "I am writing in response to the letter in the May-June issue in which a parent gave the analogy of PWS's being like starving. I remember a speech at a PW Conference a few years ago in which a professional made this same analogy, submitting it, I believe, as a theory based on observation of metabolic and food-seeking aspects of the syndrome. The important aspect, I believe, of the parent's response to this analogy, is for the parent to always remember one thing. Even if it were true that the hypothalamus is giving a false signal that the individual with PWS is starving, allowing him/her to overeat will not satisfy the hunger. Parents must act on the situation as it really is,

Two Comments on Past Issues of *The Gathered View*

dinner rather than miss school or work. ...Had the need for food been at a starving level my expert could not have made such choices....This aspect of Holly and the newsletter were recently shared with a local school staff person who called me for any help with a 300 lb. PWS mainstreamed 9th grader whose parents weren't doing food control strategies at home. Here Mom may have the hopeless-starving-attitude about her child which allowed her weight to become so intolerable. I'm hoping the analogy and your excellent response may help that Mom rethink what makes her daughter tick and seek out the kinds of help that may save the child's life." (This mother continued in sharing that the school

not as they think their offspring might feel. Abiding by the diet and not by the requests for food is the life-saving action to take. I can remember that my own son frequently would say to me, "I don't feel hungry, I just like to eat." My analysis of his comment was that he felt the same way all the time. If one never knows what it feels like to be satisfied how can he know when he is feeling hunger?

In the final analysis, it does not matter whether the behavior of food-seeking is compulsive in origin or stimulated by a false brain signal. We as parents and/or care providers must choose to respond in the way that is best for the person in our charge."

We thank these parents for taking the time to share.

Some Food for Thought

Italian Salad

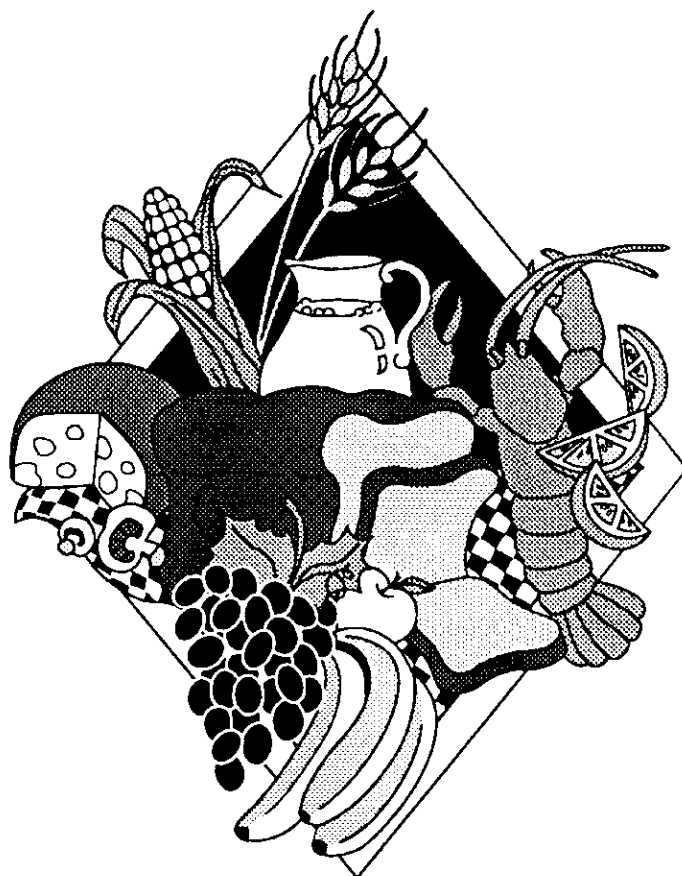
2 cups red leaf lettuce, 4 cups fresh spinach, 1 green pepper or sweet yellow pepper cut into strips, 1 small red onion, sliced, 2/3 cup sliced radishes, 2 Tbsp. red wine vinegar, 1 Tbsp. water, 4 tsp. olive oil, 1/2 tsp. each garlic salt & dried oregano, 2 Tbsp. grated Parmesan cheese. In large salad bowl toss together lettuce, spinach, pepper, radish and onion. To make dressing, combine remaining ingredients, except Parmesan cheese, in a small jar with lid. Cover jar and shake well. - Pour dressing over salad, toss, and sprinkle on Parmesan cheese. Makes 4 servings. 83 calories -- low cholesterol. Preparation time: 25 minutes.

Stuffed Tomatoes

4 tomatoes, 2 cups frozen mixed vegetables (zucchini, carrots, lima beans, Italian beans), 2 Tbsp. grated Romano cheese, pepper. Cut tomatoes in half and spoon out pulp. Chop tomato pulp and discard seeds. Cook vegetables according to directions on package, drain, and stir in tomato pulp. Spoon the mixture into each tomato shell and place in a 10x16x2 inch baking dish. Sprinkle with cheese and pepper. Bake at 350 degrees for 15 to 20 minutes. Makes 4 servings. 53 calories -- low cholesterol. Preparation time: 15 minutes.

Pasta with Ham and Vegetables

4 oz. (1-1/2 cups) pasta (macaroni, fettucini, or mostaccioli), 2 cups sliced zucchini, 1/2 cup sliced green onion, 1/4 cup water, 4 tsp. cornstarch, 1/4 tsp. each crushed dried basil and crushed dried marjoram, 1 can (13 oz.) evaporated skim milk, 5 oz. (1 cup) cooked ham, cut into strips. Cook pasta, drain and set aside. In saucepan combine zucchini, onion and water. Bring to boil, reduce heat and simmer, covered, for 4 to 5 minutes until vegetables are crisp-tender, then drain and return to pan. In a small pan combine starch, basil, marjoram, 2 Tbsp. of the milk and a dash of pepper. Stir in remaining milk. Cook and stir until thick and bubbly. Stir sauce, ham and pasta into vegetables and heat through. Makes 4 servings. 291 calories -- low cholesterol. Preparation time: 35-40 minutes.



Our Supporters

From mid-April to mid-June, contributions added the following amounts to our various funds:

Research: Thanks to VanZomeran (5), XI NU CHI Chapter Beta Si, Shoemaker, Boyd (2), Shuell, Weeks, Brunstad, Schneider, Alterman, Sunlighters, Neason, Hogue, Braun, and the following contributors to the Gordon Memorial Fund: Feldman, Bram, Ford, Gordon (2), Paul, Morris, Spiller, Davis, Wintner, Pieri, Maw, Beshunsky Co., Wright, Gross, Porat, Brenner, Baral, Goldblatt, Feldman, Norristown Assoc. & Goldberger, others: Needel for Weiner and 1st Congregational Church for Ingalls, Portalanza (Maranon) Memorial Fund: Davis, Bresani, Maranon & Gomis, we added \$639.63 to this fund.

CIT: Thanks to Beltran, MacLeod, Zacher, Olson (2), and Parent, we added \$200 to this fund.

Operating Fund: Thanks to the above Research Donors (50% of the donation) and direct donors, Battoglia, Zacher, Abramek, Calo, Brunstad, Schneider, Alterman, Scanlan, Neason, Sunlighters, Beltran, Hogue & Englert, we added \$772.87 to this fund.

We also thank members for *Contributing Dues:* Koerber, Braunreiter, Horrocks, Marcinko, Tsai, Noll, Kowtna, Strieby, Mackay, Ingalls, Greenall, Totten, Carton, Bennett, Sidlo, Krueger, Kraft, Stone, Ragland, Tarica, Kania, Jackson, Schultz, Morgan, Stickle, Bayless, Underwood, Bunker, Maloney, Nichols, Post & Trachtenburg. *Patron Dues:* Weiner, Eisen, Bell, Basker, Latt, Neason, Vektor, Waldon & McCall.

Statistics from England

In response to a question of has your daughter menstruated, the following numbers were reported:

Age:	Periods Not Started	Periods Started
under 10	29	0
10.1-13	6	1
13.1-16	12	1
16.1-19	4	4
19.1-22	6	5
22.1-25	2	1
over 25	8	9
Totals:	67	21

Many of the girls who had periods did not have them regularly -- some have only experienced one or two. The theory could be put forward that at least half of all girls with PWS will experience some form of menstrual cycle, albeit in many case very irregular.

In response to a question of male treatment, it was reported that 51% had some treatment of "normalizing" sex organs, mostly operations to bring down undescended testes, some of which were successful, some less so. Two boys had had plastic implants, purely for cosmetic or psychological reasons, and three teenage boys are on a course of treatment to aid pubertal development.

Association Study

The American Society of Association Executives recently conducted a study at the Hudson Institute in Indianapolis on the activities and value of associations. The study, titled "The Value of Associations to American Society," obtained data from results of 5,500 questionnaires sent to national associations, by reviewing association literature, and by discussing association activities with executives. The study was conducted to raise the awareness of the public and of policymakers about associations and their activities. Some of the major findings include the following:

- Associations employ more than 500,000 people, this is more than the computer, steel, or airline industries.
- Associations contribute \$48 billion directly to the economy.
- Associations spend \$14.5 billion on setting and achieving standards that protect consumers' health and ensure the quality of products and services. That's an enormous amount -- more than 400 times greater than the \$34 million the government spends to set and enforce standards.
- Associations devote \$8.5 billion to continuing education (usually of a very specialized nature) of their members.
- Two-thirds of all associations engage in research or gathering statistical data.
- While some folks believe lobbying is the major activity for associations, about two-thirds don't lobby at all. Those that do spend an average of only five percent of their budgets, most of which is devoted to explaining the government's actions and regulations to their members.
- President Bush was given a booklet which highlighted the results of the study. He emphasized the value of associations and encouraged all associations to become more actively involved in community service. A major aspect of associations is the education they provide to members and the public. Associations attempt to gather information and make it available to people in a way that will be most helpful to them. In some cases, associations are the best or only source of new information and continuing education. Members of associations spend over \$5 billion a year on educational offerings of their associations. Associations play an important role in the education of the public, and with rapid changes in technology, continuing education is vital.

Message From the President (continued)

Recently I had an exciting and wonderful week with my daughter, Sarah, at her new school. I had the privilege of attending her classrooms and activities at the Institute of Logopedics in Wichita, KS. The changes that they have wrought were a marvel to me. Her feelings of happiness and self confidence and success were apparent throughout the week. This school recognizes the language deficits of the Prader-Willi syndrome and with their great experience in supporting language deficits in other multi-handicapped persons have been able to give my daughter new avenues of education, socialization and hope for vocational development.

Earlier last month I was fortunate to be one of the first participants in the PWSA course on PWS residential management. The people at our home office and the Minneapolis PWS residence put on a superb three day tutorial. The material presented will be used to expand the "Yellow Manual" that Marge Wett and Dorothy Thompson have compiled as a guide to residential management. This program will be held

again in the future and should be put on your "don't miss" list.

Not long ago I had a discussion with my fifteen year old daughter on what really constitutes happiness. We have been blessed with my financial success as a physician; we live in one of the most desirable parts of the country (if earthquakes don't bother you); and we have enjoyed good health. My daughter is successful in her interests in equitation and school. We talked about all of these things, but in the end we agreed that the type of thing that produced a real sense of happiness was being involved with others and giving of yourself in support of their needs, enjoying the privileges of friendship. These are the things that I have had the privilege of doing that have added to my own personal happiness in the past month. The national PWSA conference is nearly here. Come and join us with a part of your life and you, too, can have the privilege of a little real happiness.

Sound Familiar?

George Popper, Ph.D., writes that some special children insist on having things their way and have little tolerance for disruption of their own routine. Truly compulsive individuals not only engage in compulsive behaviors, they are often seen as emotionally restricted and preoccupied with trivial rules to the point where they no longer are able to function effectively.

For a behavior to be truly compulsive it must be something a person doesn't really want to do yet feels powerless to stop. Tantruming is an example of compulsive behavior. For the special child, the compulsive behavior seems to provide a kind of comfort and security. Understanding the child's need for security is important when trying to modify compulsive behaviors. Punishment is usually ineffective. A more productive approach is to substitute a more desirable behavior for the one you wish to extinguish. Remember, they are difficult to modify, and doing so calls for patience and understanding.

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 the PWSA. Duplication of this newsletter for distribution is prohibited. Quotations may be used if credit is given to PWSA. Membership dues are \$20.00 per year Individual; \$25.00 per year Family, and \$30.00 per year for Agencies/Professionals (U.S. Funds). Send dues and change of address to: PWSA, 6490 Excelsior Blvd., E-102, St. Louis Park, MN 55426. Any questions? Call us at (612) 926-1947.

PRADER-WILLI SYNDROME ASSOCIATION
6490 Excelsior Boulevard, E-102
St. Louis Park, MN 55426

First Class Mail