

PRESIDENT'S MESSAGE

One day, when my daughter Sarah was about two and a half, I found myself in a conference room of the University of California at San Francisco Medical School. There Dr. Bryan Hall and his associates were seeing those characteristic signs of the Prader-Willi syndrome demonstrated in the person of my daughter. I had never heard of Prader-Willi syndrome in twenty-five years of medical practice, and to this day most physicians as well as others have little or no knowledge about PWS. Those of us who read *The Gathered View* are indeed a select group. We know more about PWS than almost anyone you might care to name, including most medical people, most educators, most politicians, most relatives, and most just about anyone. Just the other day, a fellow physician who supervises the care of a large group of handicapped children sent a patient case description, complete with photos, for my evaluation as the "local expert" on PWS. This particular case was not PW, but I did spot a PWS person in the halls of the hospital the next day.

Yes, like you, the past six years have taught me a great deal about Prader-Willi syndrome. At times I think about the patients that I normally take care of, the patient that is scheduled to have heart surgery. By the time that I finish my pre-anesthetic visit they have seen so many doctors and other medical people that they know "more than they ever wanted to know" about their problem. But like that patient, I can usually come up with one more thing that I would like explained, one little question that nags at the back of my mind, one more problem that needs comforting. The amount of information available to the "new" Prader-Willi family is not only greater but it is more readily available. More and more people are hearing about the problem and more and more people are doing something about it.

When I first tried to look up information about PWS, I was able to turn up a half dozen references and copies of three of them. Right now there are five new medical journal articles on my desk about the syndrome, each one with a different focus, each journal with a differing primary area of concern. We are learning more about PWS, more people are interested in our concerns and more of our concerns are becoming more clearly defined. The genetic aspects are undergoing new scrutiny as the result of studies reporting abnormal chromosomes in PWS persons. Another article introduces the concept that certain types of genetically disordered mice that overeat and become obese could be treated with a medication that changed this abnormal eating.

PRESIDENT'S MESSAGE, Continued

Whether any of this "new research" produces benefits to the PWS person is probably years down the road, but the immediate benefit is that the light at the end of the tunnel becomes a little brighter. Most of us ignore those problems that do not directly touch our lives and very few people are touched by PWS. This letter will reach you with only about two months to plan your trip to our meeting in Boca Raton. It will be a good meeting. There will be light added to the tunnel and there will be others touched as you have been. JOIN US.

Delfin J. Beltran, M.D.
President

UNDESCENDED TESTES

The last issue of the GV contained a comment regarding treatment of undescended testes, and has caused a little misunderstanding among our members. As with other syndromes and medical problems, not all doctors agree on some aspects of treating PW patients. We have included information from three sources in order to clarify this handbook correction and present various opinions.

In our handbook, page 36, Shirley Neason, author, wrote, "If the male has undescended testes, surgery is usually performed before puberty to bring them down as a precaution in the event of malignancy."

Dr. Vanja Holm states, "In the case of a child with PWS, it is not necessary to get the surgery done before puberty. The usual reason for performing the surgery before puberty is to prevent sterility. Since people with PWS are sterile anyway, this precaution need not be taken."

Dr. Stacy Roback, Minneapolis: "The testis that fails to descend spontaneously, or that cannot be manipulated into the scrotum, after one year of age should be considered cryptorchid, and as such, requires treatment. There is considerable disagreement regarding what constitutes optimal therapy and at what age it should be provided. The previously accepted program of watchful waiting until puberty to see if spontaneous descent might occur is no longer recommended. There is no clear histological evidence indicating that when a cryptorchid testis fails to descend into the scrotum before 2 to 4 years of age, normal maturation will not occur. Although exogenously administered human chorionic gonadotropin (HCG) has been shown to stimulate descent in occasional patients with bilateral cryptorchidism, no clear evidence exists that it affects descent in the usual undescended testicle with a short spermatic.."

Dr. Hans Bode, Boston: "I would like to point out that cryptorchidism of patients with PWS will respond very well to medical therapy in most patients. ...no patient should be treated surgically unless medical therapy has failed. It is generally believed and fairly well documented that cryptorchidism of patients with PWS is a consequence of deficient gonadotropin secretion during the latter half of pregnancy and not due to a mechanical obstruction..."

(Our request for documentation from Dr. Bode was not answered.)

EXCERPTS FROM A LETTER FROM A FOSTER PARENT

R came to our home weighing 138 lbs. and 56" tall. I read everything I could get my hands on about PWS, which as you know was very little. So I began watching what he ate and trying to note the results of his weight and emotions. It didn't take long to realize that he would eat almost anything. He had not been on a diet before so was started on a 1200 calorie diet. He began losing a little weight almost every week. Salt was limited, as were other spices. Needless to say desserts and sweets became a thing of the past except on very special occasions.

As time went on I noticed that when R had sweets in an average amount he was prone to tantrums. The diet pops, artificial sweeteners and other food additives also have an adverse affect on R but not nearly as much as the sweets. In order for him to have adequate nutrition on 1200 calories there can be literally no empty calories. Nutrition is of equal importance as calories.

R swims very well, rides a bike (it took him longer to learn than most) and also walks a mile to school and back if weather permits. He takes great pride in his weight loss. His emotional control has expanded so much that one month he had no tantrums.

When R had been here several months he visited his previous foster home. He returned and tantrumed several times a week for several weeks. He had been in the same foster home for a number of years and when he tantrumed he was given his own way or even bribed with sweets if he would stop.

It was a very difficult job to keep him on his diet, properly nourished and reasonably content. The emotions of these children are definitely different. If we laugh at TV, R makes a big production out of it. Long after we have stopped laughing he is still guffawing. Also his muscle tone is flabby.

I know now that R needs a firm hand, encouragement, praise and love much more than he needs candy, cake, soda and his own way. With God's help we will make it and hopefully by the time he is older he will be able to go into a group home and function well.

R is a big help also. He makes his own bed, helps with dishes, clean about his person and also looks well groomed.

After writing this lengthy letter, if nothing else I feel less guilty about my feelings toward R at times. Just telling myself he can't help it doesn't always help what I feel inside, but knowing others have the same things to contend with helps.

MIDDLE ATLANTIC GROUP MEETING

The middle Atlantic group of the PWSA is planning its second get-together on April 4th, from 10:00 A.M. until 4:30 P.M.

The meeting will be held at Williamson's Restaurant, Horsham, PA.

The program will consist of speakers Dr. David L. Margules, Ph.D., of Temple University on the topic of obesity and Dr. Philip Kruper, psychologist at Norristown State Hospital who will speak on behavior modification.

Reservations will be necessary. Please contact Doris Jane H. Miller, 238 East Court Street, Doylestown, PA 18901, if you are interested and did not receive a notice of this meeting.

COMMENTS ON PROBLEMS

Excerpts from a letter of a PW sibling:

"You are an answer to prayer! Quite by accident, a friend of mine found an article in our paper about the PW victims and cut it out for me. My little sister is a PW child and I am glad to discover there is an organization to help them and their families. I am interested in receiving the newsletter your association distributes and also I would appreciate your sending me information concerning special schools and homes for PW people.

I cannot tell you how glad I am to hear of your organization and that there are homes for PW people. My mother has cared for R for nearly 13 years, and quite frankly, is tired of the never ending battle. Mom is finding it very difficult to put R in a home because she is afraid the people there will not understand her problems.

R goes to a public school class with mentally retarded and brain-damaged children. She has had several 'run-ins' with her teachers over food. It is obvious to us that these teachers simply do not understand her problem no matter how much we explain. It is very frustrating for my mother when they tell her to put R on a diet and in the same breath ask her if she can make cupcakes for the class Valentine's Day party. One teacher even put a piece of cake in R's lunchbox (so she wouldn't forget to take it with her to the cafeteria). When they got to the cafeteria, the teacher wanted her cake back and you can imagine what happened. The teacher said she got so exasperated with R that she threw the cake on the floor and left the room. When she returned, she was horrified to find that R had eaten the cake.

I'm not relating these stories to see how gruesome I can get--I suppose I'm just getting some of it off my chest. I'm truly relieved to know that there is someone who knows what we have been going through. Other people just do not understand. I am happy to see articles in the newspaper about the syndrome because it proves to people (like R's teacher) that we are telling the truth and not making up stories.

Thank you for organizing the association. It is moral support!"

Ms. M's letter is typical of problems faced by many families with PW young people. As the PW grows (age of onset varies), behavior problems and food control have become severe to the point that institutionalizing is the only means of survival. Alternative placements should be available, but in many cases are not.

Studies have found the PW patients to be more verbally aggressive, self-assaultive and regressive than other retarded patients. Yet, we haven't managed to convince many physicians, other medical professionals and educators that PW children do have unique problems. Many parents write that their children's unpredictable temper tantrums are a major problem, and a great deal of these tantrums are food related.

A recent article appearing in "Behavior Therapy" written by Travis Thompson, a University psychologist and Steven Kodluboy, a social worker, gives details of a study done on behavioral treatment of obesity and concludes a one-to-one basis of control is the only one that works over a long period of time. Other studies have been done, some by institutions, showing success in a situation not so closely monitored.

"Prader-Willi Syndrome", a book soon to be released by Dr. Holm, Dr. Sulzbacher and Ms. Pipes, contains a chapter by Stevan Nielsen and Dr. Sulzbacher on their studies with relaxation training for PW patients. Other papers include studies in other behavior areas.

Dr. J. Herrmann, Medical College of Wisconsin, at our 1979 National Conference, stated the two principal problems of obesity and behavior have a very significant impact on the family. His suggestions include education of the public and professionals which has been a definite problem of many parents. Many parents state they cannot do this alone.

PROBLEMS (Continued)

We then have the age-old problem--Yes, we know what the problem is, what are some answers?

The PWSA's purpose is to make more material available so parents, who are not located near large centers with experienced PWS professionals, can have information to pass on to the people working with their child. Until more information does become available, we need parents willing to share their experiences with others. This has taken place at the conferences but unfortunately everyone can't attend the conferences. Local chapters are also filling this need in some areas but again, not everyone has enough people located close enough to warrant a local chapter.

I feel the "Overview" by Lota Mitchell, a social worker, published recently by PWSA helps tremendously when sharing knowledge of the syndrome with professionals. Only through their understanding will we be able to reach these people, and have them accept the uniqueness of our children. Also the coming publication of the PWS book will be a tremendous tool.

All too frequently most articles regarding the PWS refer to the major difficulties of the older child. As knowledge of the syndrome is being expanded, more children are being diagnosed at an earlier age. The parents are faced with a hypotonic (floppy) baby, and the "failure to thrive" stage, lack of sucking ability, "model" babies that only sleep and never cry. As the infant progresses and a diagnosis can be made, the parents began to read the reports of the major difficulties of compulsive eating and temper tantrums and say, "this can't be my child." And all too frequently the child is diagnosed by a doctor that doesn't have very much information to offer the parents because of his lack of information.

This is an area where we need parents to share their experiences with others. And we need to reinforce the idea not every child will develop these problems to the same degree. I personally feel it is far better to face the reality of the future even though it is far from a "rosy" picture.

In the meantime, I feel our organization is here to share whatever information that we can obtain. How many of us take the time to write to the GV when we do obtain some success along these lines?

In response to request from parents, I wrote the following comments to share with you:

1) "Prader-Willis are all alike" Specialists have found young people with this syndrome have a very definite pattern of signs and symptoms, particularly in relation to food and behavior. Naturally every person is an individual, and no one can be "lumped" under a label of any type and expect a perfect fitted mold, but I have found the similarities of the people with this syndrome are remarkable.

2) "I have had other retarded people with the same I.Q. as your child, and they have succeeded in this program" PW people do not function at their I.Q. level. Expecting their functioning to follow a typical M.R. pattern will only lead to failure in a program.

These people have a very selective learning ability and require a specifically designed program for them. Their program needs to be very structured as they resist change. A greater percentage of the young people need constant supervision. (This is not as true for younger children nearly as much as it is for the teens and older young people.)

PW people do respond to praise. You can get a lot of mileage out of a smile or a hug.

(I might note not all PW people are retarded but a greater percentage of them do fall in this I.Q. range.)

PROBLEMS (Continued)

3) "This child is retarded but certainly can learn that this is not acceptable behavior" I do not believe a PW can learn "acceptable behavior". Certainly, we cannot excuse them from following behavioral guidelines for acceptable behavior but if we expect them to be able to learn like 'normal' people, I am afraid in a greater percentage of the cases, we are expecting the impossible. I feel their compulsions relate to their brain defect and are not controllable in the usual manner. These young people have very concrete (fixed) ideas and cannot be threatened with punishment, denials or that type of regulation because it just doesn't work. Their stubbornness and manipulative behavior is as much a part of their make-up as breathing.

4) "All people, handicapped or not, should achieve some independence and be allowed to 'grow up' and develop responsibilities and social relationships" It is very difficult for people working with other handicapped people to accept the uniqueness of the PW person. PW people cannot achieve independence because they cannot control these compulsive behaviors. Educators are leaning toward 'mainstreaming', creating a more 'normal' type of education for handicapped people. I feel this is great, but 'mainstreaming' is not the answer for everyone and I definitely believe very few PW young people can function under this system.

The food drive is not susceptible to self-control or self-discipline, and attempts to teach control of this drive with a reward and punishment system has rarely been achieved.

5) "Why is food control so essential" A great emphasis is placed on food control and I feel this is essential. Obesity control is a life-long problem. The fact that PW people gain weight on less calories, develop complications when overweight, and reach life-threatening situations at a lower weight level than other obese people, I feel justify these controls.

6) "We will have no difficulty placing your child in a proper group home" I do not feel the usual PW person can achieve success in a group home as most are presently being operated.

Associations for retarded citizens are advocating group homes of 6-8 people with as near "homelike" atmosphere as possible. These PW young people already have a "home" atmosphere in their own homes and it is not working. PW people cannot be molded to fit into existing group homes. The group home program must be designed to fit their needs.

I also feel a larger home with more staff has a much more likely survival rate. "Burned out" staff can be replaced one at a time without disrupting the structure and continuity of the program.

7) "What are some suggestions?" A successful school placement and living arrangement are achieved with programs designed with the uniqueness of the PW in mind. A placement with complete food control and a behavior modification program can only be achieved if the PW's needs are understood. Many of these people are placed in school programs, group homes, vocations, job situations, and they do not succeed. Only in the past few years has there been much success in many of these programs as more material becomes available to the personnel working with these young people. Many doctors have begun to talk of the frustration of these individuals and the removal of this frustration, by a properly designed placement, has made the world of difference in attempts to work with them.

The stress on the family has been heavy and constant, particularly in families where a diagnosis has not been obtained early. The outlook has been somewhat more hopeful in the past few years.

I strongly advocate a structured school placement and a completely controlled living situation. This is where parents have run into a great amount of difficulty with their children. The only existing controlled living situations are for more severely retarded people. We cannot justify "locking these children up" because they cannot function in a group home designed for people in their I.Q. range. Several states are now in the process of developing group homes designed for PW people. It takes a great deal of time and effort to establish such programs, and it will be a considerable time before they will be available in many states.

PROBLEMS (Continued)

The above statements are my own opinion and solutions that I have concluded through my personal experiences with my own child, as well as the associations I have experienced over the past few years in my working with the PWSA and its members. I have also drawn from my experiences of working with other handicapped young people. Being married to a medical doctor, and raising six "normal" children also weighs on my experiences. If you disagree with some of the statements and assumptions I have made, please send your comments and we will be happy to share them with others in the GV.

I call the GV our "sharing and caring" newsletter. Are you a contributing member?

Marge A. Wett

RESEARCH COMMITTEE

Approximately 200 questionnaires have been sent to doctors and institutions throughout the United States and Canada. The results of these will give us an idea of who is currently doing research and who is interested in conducting new projects and of what type.

I am also writing letters of inquiry to get copies of information already compiled so that we might have them filed in one central location.

Toni Parker, R.N.
Chairman of Research Committee

NEW PUBLICATION

"A Difference In The Family" (Life with a Disabled Child) by Helen Featherstone

Some five million families in this country live with a seriously disabled child. How are their lives affected by this "difference" in their midst? How do they cope with the constant fear, the bitter anger, the sense of guilt and personal inadequacy, and above all the terrible loneliness that makes it so hard to communicate even with well-meaning friends and relatives? What happens to marriages and to "normal" brothers and sisters? And why is it so hard for professionals -- the doctors, therapists, teachers -- to provide the desperately needed support and understanding.

Helen Featherstone, a professional educator (formerly an Assistant Professor of Education at Wellesley College), and the mother of a seriously disabled child, draws on interviews with parents and professionals as well as her own experience in the publication of this book.

Orders may be placed with Basic Books, Inc., Attn: Mail Order Department, 10 East 53rd Street, New York, NY 10022. Include \$13.95 and they will pay postage and handling. NY state residents, please add sales tax.

AGENDAS WILL BE IN THE MAIL SOON FOR THE THIRD ANNUAL NATIONAL
CONFERENCE - JUNE 25-27 IN BOCA RATON, FLORIDA

FINANCIAL REPORT DECEMBER 31, 1980

PRADER-WILLI SYNDROME ASSOCIATION

RECEIPTS

Membership Dues	\$5,758.17	
Donations	8,174.96	
Material Sales	2,151.50	
National Conference	3,768.97	
Savings Account Interest	<u>253.41</u>	
		\$20,107.01

EXPENSES

Salaries	\$ 750.00	
National Conference	4,470.12	
National Committees	24.94	
Printing	3,006.72	
Postage	1,365.92	
Computer Service	419.80	
Office Supplies	399.41	
Secretarial Service	345.00	
Currency Devaluation	55.56	
Telephone	41.80	
Office Equipment	1,715.50	
Miscellaneous	<u>109.45</u>	
		\$12,704.22

Net Gain for year	\$ 7,402.79
Cash on hand, December 31, 1980	10,082.59
Cash on hand, December 31, 1979	2,679.80
Net Increase in Cash on hand	\$7,402.79

As you can see, without donations we would have a loss for the year instead of a gain. We do appreciate your continued support. The \$750.00 salary was paid in the beginning of the year, and for the remainder of the year, all personnel have been donating their services to your organization.

We now have a balance of cash on hand that enables us to do some spending. Since the dissemination of written material is the chief goal of the organization, we would like to use this money in purchasing services for more material. If anyone has any suggestions or comments, we would welcome them.

PRADER-WILLI CALIFORNIA FOUNDATION NEWS

The Dubnoff Center Group Home has began accepting applications of young people between the ages of 12 and 15 years of age in order to fill the four places that they will have available. This is the group home that the California group has been seeking to open for the past many months.

The Meteor Ranch Bible Conference in Upper Lake, CA has set aside a two week camping session for Prader-Willi people. Information can be obtained from the foundation, by calling John Shimmin after 6:00 p.m. at (213) 287-5332. The two week period starts July 12th.

Mrs. Henry Tarica, 18549 Ventura Boulevard, Ventura, CA 91356, (213) 981-6138, is assembling a list of respite care facilities which have previously served PW people. If you know of any California area facility that has provided this service, please send the name and address of the facility along with the name and address of attending physician to her.

OPHTHALMOLOGIST SEEKING INFORMATION

Helen M. Hittner, M.D., F.A.C.S., Pediatric Ophthalmology Associates, Doctors Center, 7000 Fannin, Suite 2250, Houston, TX 77030 has been treating a Prader-Willi patient with the following condition:

Her patient has partial albinism. Vision approximately 20/40, mild astigmatism, more in the left eye than the right. He had an esotropia, primarily of the left eye. His slit lamp examination revealed transillumination of the irides 3+. The fundus examination revealed very blond fundi with some graying of the optic nerve and slight decrease in the macular reflexes. The child also had an electroretinogram and visual evoked response which were consistent with the diagnosis of ocular albinism.

She would be most interested in seeing other patients with the PWS especially if they have nystagmus and are very blond complected so that it can be determined whether this is a chance association of partial albinism with the PWS, or if in fact, the small deletion known to be present is the cause of the ocular albinism.

She would be very happy to have anyone contact her.

NEW JERSEY GROUP HOME REPORT

Zina Levin, 902 Folsom Avenue, RD #1, Mays Landing, NJ 08330, reports that some progress is being made with gathering of material for their Certificate of Need for the group home. A property and staff have been found.

If anyone is interested in this home, or working with the Levins, please contact her.

PEN PAL REQUEST

Several PW young people have expressed a desire for a pen pal list. If you would like to add the name and address of a young PW person to this list, we will accumulate the names and then send them to any PW requesting Pen Pals.

TRANQUILIZERS

We have received letters from a few parents of PW young people who seem to have more severe behavior problems than the "average" PW. These parents would appreciate anyone having any information in this regard to write to the editor. Comments are particularly wanted in regard to the usage of tranquilizers for these young people.

As with all of our requests for information, letters to the editor will be published whenever possible. This is your newsletter!

NEW YORK GROUP

A New York group of PW parents have been meeting monthly at the Bernard Fineson Developmental Center in Howard Beach. Nine families have been attending the meetings and others have been contacted. Two guest speakers were on the agenda for February's meeting, one a genetic counselor from St. Mary's Hospital and the other a Queens coordinator for the March of Dimes.

If interested in joining the group, contact:

Mr. & Mrs. S. Levine, 234 Locust Avenue, Freeport, NY 11520

GOLD CREEK PROVIDES HAPPY CAMPING EXPERIENCE

The Prader-Willi Northwest Association sponsored a successful camp for children and youth with Prader-Willi Syndrome in August of 1980. Named Gold Creek Camp, the Seattle area facility was close enough to urban areas to provide variety in field trips, but at the same time was rural enough to be camplike in setting.

The primary stated purpose of the camp was respite care, thus making available state funds for those parents who applied for them. The primary focus for the campers was recreational, with additional emphasis on exercise and diet. A number of outings to museums, parks, beaches, fish farms, and other places provided opportunity for the campers to experience the larger community. In addition they panned for gold, swam, slept out, hiked, sang and played games.

Among the skills that were emphasized were caloric awareness, conversation skills, behavioral skills, being helpful, table setting and manners, water play, traffic safety, creativity and planning, fire safety, honesty, singing and listening.

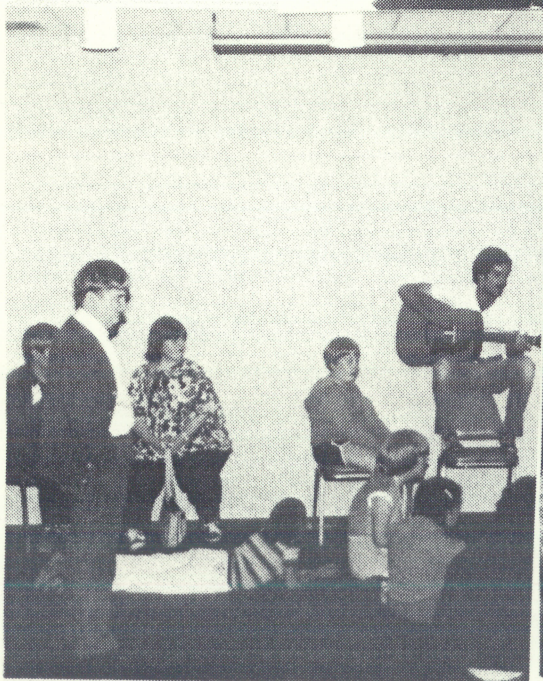
Among the responses from parents was the following: "Thanks for all your work in organizing the camp. I appreciated the daily diary, which gave us an idea of what they were doing. You did a marvelous job in planning and executing the whole program."

Another camp is being planned for the same location for the last three weeks in August this summer. Any parents interested in this camp should contact the following address:

Prader-Willi Northwest Association
14255 S.E. 40th
Bellevue, WA 98006
Phone: (206) 747-9997

Cost of this year's camp will be \$150 per week. Those planning to send their children or young people should apply early to their state social service agency for respite care funds. Camperships may also be available through local chapters of such agencies as United Way or March of Dimes.

Conference 1980



INTERNATIONAL SYMPOSIUM ON GILLES DE LA TOURETTE SYNDROME

Sponsored by the National Institute of Neurological and Communicative Disorders and Stroke, The Tourette Syndrome Association and the Gateposts Foundation: Roosevelt Hotel, New York, May 27-29, 1981. Program Chairmen: Dr. Tom Chase (NINCDS), Dr. Arnold Friedhodd, NYU School of Medicine. The following topics and their relevance to Tourette Syndrome and related disorders of the CNS will be covered: pharmacology, biochemistry, genetics, animal models, neuropsychology and clinical aspects. For registration information and program: Sheldon Novick, M.D., TSA Medical Director, Gateposts Foundation, 42-40 Bell Boulevard, Bayside, NY 11361, Phone: (212) 631-0177.

AVAILABLE SOON

In the near future, we expect to be mailing you information on obtaining the book being published in March. THE PRADER-WILLI SYNDROME by Dr. Holm, et al. A collection of papers presented at a medical conference in Washington. We have reviewed the manuscript and are very pleased and excited about sharing it with you.

THE GATHERED VIEW is the official newsletter of the Prader-Willi Syndrome Association and is sent to all members. Membership dues are \$10.00 per year for U.S. members; \$15.00 per year for Canada, Mexico and overseas members. Send dues and change of address notices to PRADER-WILLI SYNDROME ASSOCIATION, 5515 Malibu Drive, Edina, MN 55436.
