VOLUME VI November-December 1980 Number 6

PRESIDENT'S MESSAGE

As I came to my desk to write this month's letter, I noted an article in the current Journal of the American Medical Association. The author has given us permission to circulate it with our newsletter and you will find a copy enclosed. At the end of the article is an Editorial from the same Journal. It seemed that it was appropriate for members of this association to be aware of this need among all persons and with special attention to the handicapped.

Last month communication was defined as the interchange of ideas. It is important to consider how a communication is "received" and the effect that will have on the value of the communication. The enclosed articles caught my eye because I was tuned-in to the problems of the Prader-Willi persons' "handicaps." As a physician, I am presented with the question, "Why me?" almost daily. As a parent, our family time is filled with an awareness of my daughter's constant battle with diets, special education, physical limitations, and the effects on the family. If I am tired from the challenge of these problems and read this article in that frame of mind, the effect could be very pessimistic. I could read the litany of problems described and focus on the terrible outcome. Were I to relate to this anecdote in this manner, it would be difficult to derive benefit from the author's message of the rarity of such severe failure in the face of the relative frequency of the problem; of the need to respect the complexity of the handicapped person's problem; and of the necessity to present the opportunity to succeed. Nothing succeeds like success. We all need some.

Last week, I flew to Fresno to testify at a State Senate Subcommittee on The Disabled. The purpose of the meeting was to identify where the system was failing to support the handicapped. The panel consisted of two State Senators and seven department directors including: Health and Welfare, Developmental Services, Employment Development, Preventive Health Services, Mental Health, Rehabilitation and Social Services. It was my purpose to make the members of the panel aware of Prader-Willi syndrome and of Prader-Willi people's needs with special attention to residential facilities. I knew that I was going to the correct place when the agent handed me my boarding pass. Written in large, round letters was the airline symbol for my destination, Fresno Air Terminal; F A T.

NATIONAL CONFERENCE

Next year's meeting of our Association will be held in Boca Raton, Florida, June 25-27, 1981. Meetings will be held in facilities of the Florida Atlantic University and Dr. Mary Lou Caldwell will be our hostess. The format of the meeting will be based on round table discussions between parents with a professional to guide each group. Both the location and format have been selected on the basis of information supplied by the members of the Association in response to questionnaires. More specific information will be provided as it is developed. If you have specific suggestions regarding things that you want to see happen at the meeting, please contact me. Make your plans now to join us, and make this a successful meeting.

Delfin J. Beltran, M.D.

GROUP HOME NEWS

The California group has received state approval to establish a PWS home. They are now looking for a site. There is also a possibility of a home on the East coast. As was mentioned in the last GV issue, there is a nurse in NJ who operates a group home that provides care for retarded and two PW people. She would be willing to start a home just for PW people if there is a need. She is awaiting to hear from parents who need placement for a child. She indicated that she will be able to accept out-of-state residents. Inquiries should be sent to me at the address listed below. Needless to say, that home is only in the planning stage so it would not be operational for some time.

The Minnesota group is working diligently on Oakwood Residence. Most of the required applications are in and we now must wait for the approvals, licenses and permits. We are hopeful that we will have the home ready for occupancy in early winter. We have received so many inquiries - young people in other care facilities that are not suitable, others still at home with parents. At this time, it appears that our capacity of 15 will be filled by Minnesota people. We have received several requests from out-of-state people and it appears that we may not be able to accept their applications. We urge people to try to get their own state to establish a home that could serve PW people, even if they would be included in a home for people with the same mental abilities that also have similar diet needs, but not PWS.

When I hear from the California group, as to their progress, we will keep all of our members informed.

Mrs. Lois Olson, Residential Facilities Chairman
3771 Dennis Lane
White Bear Lake, MN 55110

EDUCATIONAL PROGRAMS

We have received brochures from a specialist publisher named "Melton Peninsula, Inc." , 1949 Stemmons Freeway, Suite 690, Dallas, TX 75207. Two programs that they mentioned were:

The RADEA Program for $495.00 - A Developmental Program for the Moderately and Severely/Profoundly Handicapped.

The SAIL Program for $295.00 - A practical new approach to teaching community living skills to developmentally delayed adolescents and adults.

If you would like additional information on these teaching programs, you may write to them at the above addresss, or call their toll free number 800-527-7830.

WELCOME TO NEW BOARD MEMBER - LOTA MITCHELL

After graduating from Muskingum College in Ohio, Lota married, then went to work as an administrative assistant for the federal government at a classified site. She retired for several years to have three children before going back part-time as Co-ordinator of Christian Education for a Presbyterian church in Pittsburgh. She will receive her Master's degree in Social Work in December 1980, and would possibly like to work with parents of mentally retarded and developmentally delayed children. The part-time job market will determine that.

Husband, Dave, is President of Financial Accounting Systems, Inc., a data processing company serving Savings & Loan Associations. Julie, just turning 11, is their PW child. She is not a classical PW, but has the history and most of the symptoms and the problems. Her weight is controlled, thanks to constant surveillance and locks on the kitchen doors. Big brother, Dave is a junior at the University of South Carolina, and Doug is a freshman at the University of Tennessee.

VOCATIONAL TRAINING & PLACEMENT COMMITTEE

The following persons have volunteered to be members of the committee:

Bob Allia, Massachusetts
Mrs. Gwen Keddie, Massachusetts
Stewart & Bronnie Maurer, Georgia
Dave & Loujean Burleigh, Maryland
Pauline & Jacques Parent, New Hampshire
Ms. Elain Unger, New York
Mrs. Lenore Gordon, Pennsylvania
Ms. Marjorie Henderson, Ohio

At a brief meeting at the conference in Cape Cod, we set two initial goals toward which we would each work in our own individual states:

(1) Identifying sheltered workshops and agencies providing vocational training and placement for the mentally retarded, and

(2) Contacting those workshops and agencies to:

a. Determine if any had PW persons and how they were handling any problems with that person, and

b. Provide education and awareness of the PW syndrome by sending them a synopsis of the syndrome.

We would like to recruit some members who live west of Ohio, since everyone now is from the east and south. If anyone is willing to join us, please write to me and I will send you a packet of materials.

Also we would like to request anyone who knows of an organization that is employing or training a PW person, to please send me the name and address of that workshop or agency. At present, I know of only two — Ruby Stevens' Seven Valley Shepherds, Inc. in New York, and Washington Industrial Center in Pennsylvania, sponsored by ARC.

Lota Mitchell, Chairman
844 Foxland Drive
Pittsburgh, PA 15243

NEW YORK AREA CHAPTER FORMED

Several parents met on October 14, 1980, in Queens, New York City, to form a New York area chapter of Prader-Willi Association.

Immediate goals discussed were:

(1) Creation of group homes
(2) Development of community services (medical, vocational, educational, etc.)
(3) Information exchange

Parents in nearby areas are encouraged to join with us in building a strong chapter. Our next meeting will be in Queens, New York City, on November 20, 1980.

For information call: Elain Unger (212) 347-6766
Helene Zimmerman (212) 459-8787

If you cannot attend the meeting, please write to Elain Unger, 254-08 84th Road, Floral Park, NY 11001, and you will be placed on our mailing list.

REPORT FROM PENNSYLVANIA AREA

A parent group for those interested in Prader-Willi was formed covering the Middle Atlantic States of New York, Pennsylvania, New Jersey, Delaware and Maryland. The first meeting was held September 27th in Lancaster, PA. Isa Breneisen and Doris Jane Miller organized the group. The response was good and some people traveled more than 300 miles just for this meeting. Many had their first opportunity to meet another Prader-Willi family. More than 60 people attended, fifteen of them Prader-Willi young people.

Dr. Richard Young from Hershey Medical Center spoke on difficulties in diagnosis and an attorney advised how to make provisions for our special children. Membership in the Prader-Willi Syndrome Association was encouraged as a means of keeping in touch. A meeting is being planned for April 4, 1981 in the Philadelphia area. Let's aim for 100 people next time. Put it on your calendar now.

TV SPECIAL

NBC will be airing a special documentary on December 7, 1980 entitled, "Our Largest Minority: The Disabled."

They are campaigning to have the showing the same date across the nation but you may want to contact your local station to make sure it will be aired on that date. Although this will not contain any material directly related to PWS, I'm sure it will be interesting.

NY RADIO PROGRAM

Rae Weiss and husband, Karl, psychiatrist, have a radio program in New York City, 89 U FM, "Coping thru Day." This program is broadcast on Friday evenings from 9:00 to 10:00 P.M. This is an opportunity to call in and mention the Prader-Willi syndrome. Any type of good publicity can be very helpful to all of our children.

THANKS FOR THE SUPPORT

In August, the VFW Arthur W. Jones Post 7564 of West Fargo, ND donated $500 to the Committee on Clinic Services to be used for the purpose of assuring the work on the Directory of Clinic Services would be completed. This donation was made possible through the efforts of Mrs. Evelyn Krebsbach, a member of our organization and a resident of West Fargo, ND. Evelyn is on the Committee of Clinic Services.

Betty R. Schultze, Ed.D., Chairperson
Committee on Clinic Services

INFORMATION ON PRADER-WILLI SYNDROME PRESENTED

On November 24, 1980, Dr. Betty R. Schultze will make a presentation entitled "Prader-Willi Syndrome: Evaluation and Treatment of Associated Communication Disorders," at the American-Speech-Language-Hearing Association Annual Meeting held in Detroit.

STATUS OF THE POSSIBLE CHROMOSOME ETIOLOGY OF THE PRADER-WILLI SYNDROME

Papers were presented at the San Diego Birth Defects Meeting (September 4-6, 1980), and the American Society of Human Genetics Meeting (September 24-27, 1980) in New York City which reported a missing band on the proximal portion of the long arm of chromosome number 15 in Prader-Willi individuals. Approximately 27 Prader-Willi individuals had their chromosomes studied with high resolution prophase banding and 18 were found to be missing the q11 to the q13 band of the number 15 chromosome. None of the children's parents were found to have abnormalities of their chromosomes, specifically of their number 15 chromosome.

The above findings are exciting and are of great potential importance, however, the final word is not in yet. We do not know the incidence of this deletion in the "normal" population. We are not sure that this segment is genetically active. We cannot explain why 9 of the Prader-Willi individuals did not show the deleted q11-q13 band on chromosome 15. We are surprised that no parents were carriers on their chromosome testing, as we would have expected a significant number of siblings to be affected with the Prader-Willi syndrome, a situation which does not exist at the present time. It may be that the classical Prader-Willi syndrome is non-chromosomal in etiology and the Prader-Willi "copy" syndrome is due to the q11-q13 bands being deleted from chromosome number 15. However, both situations could be due to the 15 chromosome deletion (15q11-13).

It will take quite a while to accumulate enough cases and controls to make statistical sense out of the above dilemma. Prophase banding is not a standard banding technique in most labs, so it will be very time consuming and expensive. The value of this technique is that many more bands can be observed than with the standard banding techniques utilized in most labs.

NUTRITION BY JEAN MAYER AND JOHANNA DWYER

The role of essential fatty acids in human nutrition is by no means understood. However, what is known is that they perform a number of important functions. They regulate the way the body handles cholesterol.

They also are the raw materials the body uses to make prostaglandins, a group of hormone-like compounds that stimulate the contraction of smooth muscles, regulate blood pressure, promote conception, regulate the transmission of nerve impulses and inhibit the flow of gastric juices.

The amount of essential fatty acids we need on a daily basis is really quite small, in fact, the most recent revision of the Recommended Dietary Allowances estimates that enough essential fatty acids can be provided in the adult when 1 percent to 2 percent of the calories come from linoleic acid, the essential fatty acid of primary importance to man. The American Academy of Pediatrics recommends that essential fatty acids provide 3 percent of the calories in infant formulas.

The best sources of linoleic acid include such vegetable oils as corn, cottonseed, soybean, safflower and sunflower oils. Walnuts, poultry fats, margarine and peanut oil also are good sources.

Incidentally, essential fatty acid deficiency has occurred in man but only under the most unusual circumstances. Poor growth and an eczema-like skin condition were first identified many years ago as symptoms of essential fatty acid deficiency in infants fed a skim-milk formula for an extended period. More recently, the deficiency has developed in children and adults maintained for a long time on a fat-free intravenous feeding.

(Authors: Mayer is a nutritionist and President of Tufts University. Dwyer is an associate professor of Tufts Medical School. They will answer questions if you write to them in care of The Minneapolis Star, 425 South Portland Avenue, Minneapolis, MN 55488, or a local paper that carries their column.)

EDUCATIONAL NEEDS

Several parents have written lately seeking help in their eduational programs.

One parent from Maryland wrote, "I am presently forced to appeal a placement recommendation made by the public school system which attempts to place my son, who evaluates at worst in the borderline intelligence region, into a mentally retarded (EMR) school."

A mother from Wisconsin wrote, "J has had a very traumatic school experience these first two weeks of Middle School. After four years of some really successful programming, I was not prepared for this transition and apparently neither was the school personnel. We had behavior problems surface that haven't been here for five years, tantrums, running away, etc. He was also switched from a Learning Disability class to an EMR class this year. The need for structure, continuity, some familiar faces, etc. are the needs of quote-unquote 'normal' kids. They have managed to have J in seven schools in seven years."

"Mainstreaming into Junior High was a disaster for our PW child," writes another parent.

Doris Jan Miller, 238 East Court Street, Doylestown, PA 18901, is the Chairman of our Education Committee. Any helpful information regarding educational problems and suggestions will be appreciated.

TREATMENT PROGRAMS

A doctor in Wisconsin is seeking a location for treatment for a 21 year old female Prader-Willi patient who is having behavioral problems. In addition to the PWS, this patient is also deaf. If anyone has any information that would be useful in this situation, it would be greatly appreciated.

INFORMATION REQUESTED

A New Jersey parent is asking members who have had experience with growth hormones to share their experiences with her. She has been given conflicting information regarding their use with Prader-Willi children and would like to hear what others have done.

CONTROVERSY ON HORMONE TREATMENT

There remains controversy as to the usefulness of hormones (testosterone, endrogen, and estrogen) in improving the ultimate adult height in Prader-Willi individuals. One doctor's feelings are that if it improves height at all (no adequate controlled study has been done), it is a minimal improvement. This opinion is held by the majority of people working with Prader-Willi individuals; however, some people are very positive about the increased height attained by individuals with the Prader-Willi syndrome who have been on hormones. Other uses for hormones may be beneficial, such as testosterone in males for improving testicular and penis size as well as hair growth (axillary, pubic and facial), and estrogen in females for breast enlargement and potential improvement in menstrual cycles.

OTHER SYNDROMES CONFUSED WITH PWS

In reply to a reader's question, this list includes some of the other syndromes that have some similar features to the PWS and are sometimes confused with PWS:

Laurence-Moon-Biedle Syndrome
Vasques-Hurst-Sotos Syndrome
Congenital Myotonic Dystrophy
Cerebro-Hepato-Renal Syndrome
Pseudohypopara-thyroidism
Cohen Syndrome
Urban-Rogers-Meyer Syndrome
Werdnig-Hoffman Disease
Down Syndrome (Mongolism)

"Y" PROGRAMS

A Wisconsin parent writes, "The 'Y' program has been and continues to be the best short time break, and also offers the exercise so necessary for PW kids. If your 'Y' is not yet offering a 'Special Needs Program,' I will be happy to let you know how this one got off the ground and continues to be so effective. I've gotten them interested in job training now, too."

A Minnesota mother announced her daughter had gained ten pounds last year in her group home placement. This gain was made during the summer when the group home discontinued the "Y" swimming program.

NUTRITIONAL AND EXCHANGE VALUE FOR FAST FOODS

NUTRITIONAL VALUES THE EXCHANGE SYSTEM

Fast Food Chain	Total Calories	Carbohydrates (grams)	Protein (grams)	Fat (grams)	Bread	Meat	Fat
Arthur Treacher's							
3 pc. dinner (fish, chips, slaw)	1100	91	38	65	6	4	9
2 pc. dinner	905	83	28	51	3 1/2	2 1/2	8
Burger King							
Hamburger	230	21	14	10	1 1/2	1 1/2	1/2
Double Hamburger	325	24	24	15	2	3	1
Whopper	630	50	29	35	3	3	4
Whopper, Jr.	285	22	16	15	1 1/2	2	1
French Fries	220	26	2	12	2	-	2 1/2
Choc. Milk Shake	365	66	8	8	4 1/2	-	1 1/2
Kentucky Fried Chicken							
3 pc. dinner-original	830	61	50	43	4	6	2 1/2
3 pc. dinner-crispy (chicken, potato, slaw & roll)	1070	74	54	62	5	6	6 1/2
2 pc. dinner-original	595	51	35	28	3 1/2	2	1 1/2
2 pc. dinner-crispy	665	40	37	40	3	4 1/2	3 1/2
Pizza Hut							
1/2 of 13"-thick crust	900	113	65	21	7 1/2	7	-
thin crust	850	103	50	26	7	5	-
1/2 of 15"-thick crust	1200	148	83	31	10	9	-
thin crust (cheese pizza)	1150	144	66	35	9 1/2	7	-

My son eats catsup on "everything" and I understand he is not the only PW that has similar tastes. This catsup has 12 calories per tablespoon which is supposed to be lower than regular bottled catsup.

JIFFY CATSUP

1 teaspoon Sweet n' Low
3 Tablespoons cider vinegar
3/4 cup tomato paste (6 oz.)
1 teaspoon salt
1/4 teaspoon celery salt
1/2 teaspoon paprika
1/8 teaspoon ground cloves
1/4 teaspoon dry mustard
1/8 teaspoon nutmeg
1/4 teaspoon garlic powder

Dissolve Sweet n' Low in vinegar. Stir in other ingredients, stirring well. Cover, store in refrigerator. Makes 1 cup.

A VIEW OF WHO'S WHO

Michel is age 3 now and was tentatively diagnosed as PW at 6 weeks and final at 1-1/2 years. In that spanse of time I took aggressive action on my own to keep him healthy and gain weight and later to control weight gain.

In general, Michel's diet has consisted of natural foods (no colorings, preservatives, etc.) with emphasis on low carbohydrate and high protein low fat. I have been somewhat concerned with caloric intake but mainly carbohydrate restriction.

Michel takes numerous vitamins, minerals, and organ extracts as well as thyroid. He is very content with his diet and respects the statement "No More." However, his interests truly lie in food. Michel weighs between 29 and 30 pounds and has held that weight since October 1979.

He has been under chiropractic care since six weeks of age. His neurological development has made steady and excellent progress. From an 18 month old level to 36 month old level in 9 months.

Michel has attended a school for preschool handicapped since age 1-1/2. The past year and this summer, he attended 5 days a week for 4 hours a day. His IEP program has always included speech and physical therapy.

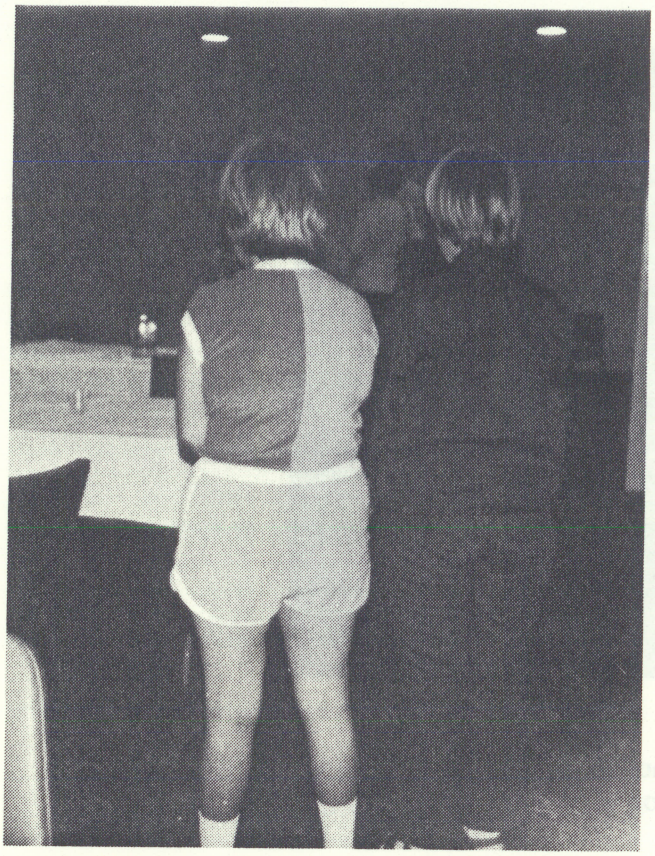
Michel is a very happy, affectionate child with numerous friends. He has a fantastic energy level—going from 6 A.M. to 8:30 P.M. with rarely a nap in between. He loves school with a passion and is one of the United Way poster children for this year.

He has enjoyed excellent health and we have not as yet had difficulties with skin rashes or picking. His scrapes and bites heal rapidly in 2 or3 days. He has excellent teeth so far and we have avoided too many difficult temper tantrums. I am thankful for an early start.

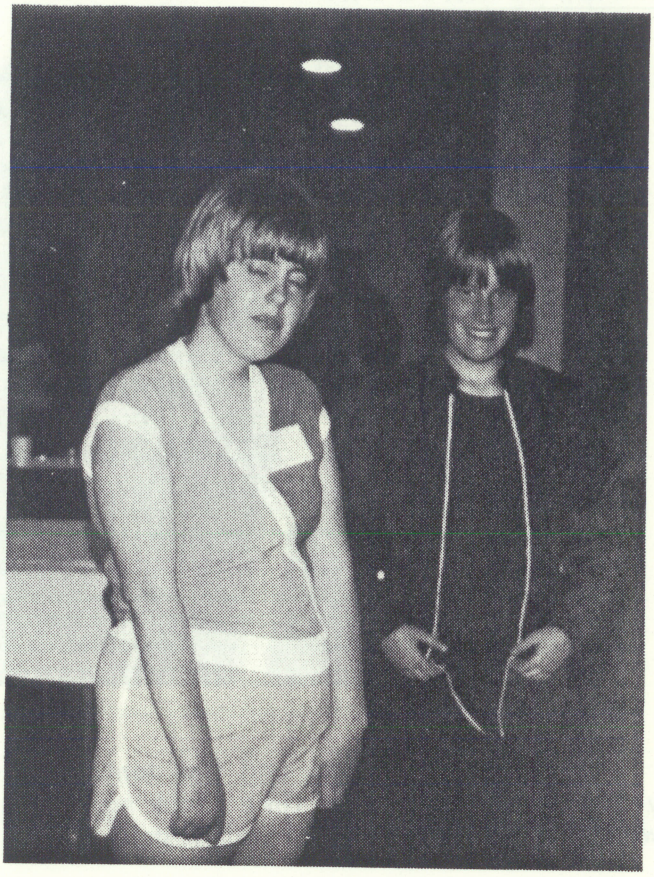
Michel attended the convention also and enjoyed the playroom and socializing with everyone.

Washington Parent

A VIEW OF WHO'S WHO, continued



Margaret & Andrea



Margaret & Andrea

I attended the Prader-Willi conference in June and took these pictures which I think are interesting. I went up to Andrea several times to speak to my daughter only to find that it was not Margaret. The resemblance was remarkable, enough alike to be twins. Since the information about Prader-Willi says they look like family, I thought I would like to point out how much.

GATHERED VIEW

My wife and I would like to thank you for all the information you've provided us with for the last 6 months. That information helped us to understand much better our child's needs and we were able to help ourselves and our dear Prader-Willi son.

Our son, Shachar, died on June 27 of this year, one month exactly before his third birthday. He had an infection in his blood that caused him his death because he couldn't fight it.

We'll always remember his kindness, love and special smile he gave to us. He was like something from heaven. That God gave him and probably wanted him as His property back after such a short time.

We want to thank you all for everything you're doing and we want to tell all the Prader-Willi parents: "All the children are a gift that God gave to us because He wanted us to feel a little bit of heaven. The wonderful love and specialty of something we only will be able to have in the next world."

Ziva & Uri Tofach

ORDER FORM - 1980 CONFERENCE MATERIAL

			Cost
#1	OPENING REMARKS by Gene Deterling	3 Pages	$2.00
#2	THE COMPREHENSIVE LONGITUDINAL CARE OF THE PRADER-WILLI SYNDROME by Andree Walczak, M.D., Michael Rees Hospital, Chicago, IL	8 Pages	$2.00
#3	SUCROSE INDUCED BEHAVIOR CHANGES IN CHILDREN WITH PRADER-WILLI SYNDROME (ABSTRACT) by Peggy L. Otto, University of Washington (Presented at conference by Stephen Sulzbacher, Ph.D.)	1 Pages	$1.00
#4	PROCEDURAL DUE PROCESS: BACKGROUND AND INTENT FOR SPECIAL EDUCATION by Betty R. Schultze, Ed.D., Judevine Center, St. Louis, MO	7 Pages	$2.00
#5	HOW TO CONTRACT FOR BEHAVIOR CHANGE (Handout at conference)	4 Pages	$2.00
#6	LESS FREQUENT? FEATURES OF THE PRADER-WILLI SYNDROME by Bryan D. Hall, M.D., University of California	3 Pages	$2.00
#7	SURGICAL CORRECTION OF UNDESCENDED TESTES by Stacy A. Roback, M.D., Children's Hospital, Minneapolis, MN	2 Pages	$1.00
#8	QUESTION SESSION - CONFERENCE CLOSE Moderated by Drs. Beltran and Wett	7 Pages	$2.00
#9	COMPLETE SET OF ABOVE PAPERS	35 Pages	$10.00

CIRCLE PAPER NUMBERS WANTED

TOTAL COST ENCLOSED _____

NAME _____

ADDRESS _____

_____

PLEASE SEND ORDERS TO:

PRADER-WILLI SYNDROME ASSOCIATION
5515 MALIBU DRIVE
EDINA, MN 55436

As you may note the above order form has changed from the last issue of the GV. In order to place this order form two months ago, it was necessary to estimate costs of preparing and printing. We were able to obtain better prices than was expected, therefore, we have lowered the cost to you.

ADDITIONAL MATERIAL AVAILABLE

The PWSA has published another paper that was made available to us by one of our members, Lota Mitchell, a social worker, from Pennsylvania. We feel this is an excellent overview of the syndrome and highly recommend it to our members. We particularly feel this would be an excellent paper to furnish to professionals working with your child.

"AN OVERVIEW OF THE PRADER-WILLI SYNDROME" by Lota Mitchell, $2.50 per copy

The Prader-Willi California Foundation held their annual meeting November 2. Included in the agenda were speakers on the study of chromosomes, the study of present residences for PW people and available programs, camp prospectives report, fund raising and the progress of the San Diego group on their PW residence.

SHARING INFORMATION

Information on homes and treatment published in the newsletter are not recommendations, but a sharing of information. It is assumed that each individual will check thoroughly before making any decisions.

We welcome any information members would like to contribute to the newsletter. The Gathered View is the members vehicle of communication with one another. We would like to have your response to any letters and information published in The Gathered View. We are limited in the amount of information we can publish; however, an attempt will be made to publish as much as possible. Send correspondence to the editor for publication in a future newsletter.

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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.

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