



The *Gathered View*

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

PWSA's Doors Are Open in Florida!



PWSA is quickly settling into its new home on Siesta Key (Sarasota), under the capable leadership of Executive Director Janalee Heinemann, aided by Office Manager David Penn, who moved with the office from St. Louis. Janalee is already hard at work recruiting volunteers and donations from the local community, planning a December open house and the January board meeting (to be held in Sarasota), planning improvements in services, and, of course, handling the daily flood of calls. Here is a photo introduction to your new Association.



Clockwise from top: PWSA's new storefront office a block from the beach has high visibility and easy access for volunteers. Our open door and colorful logo beckon potential volunteers and donors. After packing, trucking, and unpacking, office manager David Penn tackles technical problems of the transition. Book-keeper Cathy Duquette works two half-days a week to track our finances. Janalee looks from her new office window (installed by husband, Al, and son, Tad) to a comfortable lobby seating area, which she and Al furnished.



inside

Organization News.....	3
Research: Recent Journal Articles	5
Growth Hormone: Early Results from the Madison-St. Louis Study	6

Education (IDEA), SSI News	8
The Sibling View: We Count, Too!	9
Snack Strategies.....	10
Parent Questions & Advice	11

The Prader-Willi Syndrome Association (USA)

5700 Midnight Pass Road, Suite 6
Sarasota, Florida 34242

1-800-926-4797

Local: (941) 312-0400, Fax: (941) 312-0142
9:00 a.m.-7:00 p.m. Eastern Time

PWSA (USA) on the Internet:

www.athenet.net/~pwsa_usa/index.html

E-mail: PWSAUSA@aol.com

National Office Staff

Executive Director - Janalee Heinemann

Office Manager - David Penn

Officers

President - Jerry Park, Oklahoma City, OK

Vice-President - Don Goranson, Bristol, CT

Secretary - James Gardner (*Acting*)

Treasurer - James Gardner, White Bear Lake, MN

Board of Directors

Chair - Jim Kane, Towson, MD

Paul Alterman, Marietta, GA

James Gardner, White Bear Lake, MN

Don Goranson, Bristol, CT

Wauneta Lehman, Merritt Island, FL

Daphne Mosely, Oklahoma City, OK

Fran Moss, Camarillo, CA

Gail Overton-Thune, Albuquerque, NM

Pauline Parent, Manchester, NH

Leo Schertz, Columbus, IN

Ken Smith, Pittsburgh, PA

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

Scientific Advisory Board

Chair - Suzanne B. Cassidy, M.D.,
Case Western Reserve University

Chair Emeritus - Vanja Holm, M.D.,
University of Washington

Merlin G. Butler, M.D., Ph.D.,
Vanderbilt University

Elisabeth M. Dykens, Ph.D., University of
California, Los Angeles

Jeanne Hanchett, M.D., Rehabilitation
Institute of Pittsburgh

David Ledbetter, Ph.D.,
University of Chicago

Phillip D.K. Lee, M.D.,
Texas Children's Hospital

Robert Nicholls, D.Phil.,
Case Western Reserve University

Stephen Sulzbacher, Ph.D.,
University of Washington

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

Out of the New Office

Janalee Heinemann
Executive Director



“Welcome to sunny Siesta Key (Sarasota), Florida,” states the answering machine at our home. Little did I know four months ago when I co-chaired the national conference and welcomed many of you to Orlando, that I would now be welcoming the entire world of PWSA-USA to their new “home” in Siesta Key!

It never ceases to amaze me how life can take so many turns down unexpected paths. It may not be the path that we thought we were going to take but is one that leads us to the place we should be at that point in time.

When Al and I moved to Siesta Key after so many years of being enmeshed with the syndrome on a personal, state, and national level, I think we were secretly relieved to “get away from it all.” The national conference got us re-involved in spite of ourselves, and getting to know the Florida chapter members, we began to realize that whether it’s St. Louis, Florida, or another country, this is “family.” Within our world of Prader-Willi syndrome, there is a deep bond and love that goes beyond a typical relationship. A good example is the conference. As much as we Florida state chapter members thought we would be more than glad not to have to communicate with each other so much after the national conference, after about a week of the phones being silent, we all simultaneously found our hands reaching for that umbilical cord and calling to say, “Believe it or not, I miss talking to you!”

Now when I pick up the phone and hear the voice of a new parent looking for someone to hear their grief and give them support, or an “old” parent looking for an understanding heart, I know I have come home.

So, in spite of how busy it is trying to get the new office arranged and organized, please know that our phone lines and our door are always open to you. We are always glad to hear from you and will try to help you in any way we can.

❖ **New Office Hours: 9:00 a.m. to 7:00 p.m., Eastern Time** ❖

The Gathered View (ISSN 1077-9965) is published bimonthly by the Prader-Willi Syndrome Association (USA) as a benefit of membership. Annual membership dues are: \$30 Individual, \$35 Family, and \$40 Agencies/Professionals for U.S. members and \$40, \$45, and \$50 (US Funds), respectively, for members outside the United States.

Opinions expressed in *The Gathered View* are those of the authors or editors and do not necessarily reflect the views of the officers and board of directors of PWSA (USA). *The Gathered View* welcomes articles, letters, personal stories and photographs, and news of interest to those concerned with Prader-Willi syndrome.

Editor: Linda Keder, Silver Spring, Md. (e-mail: keder@erols.com)

Associate Editor: Lota Mitchell, M.S.W., Pittsburgh, Pa.

Communications regarding *The Gathered View* or PWSA membership and services should be directed to the national office of PWSA (USA), 5700 Midnight Pass Road, Suite 6, Sarasota, FL 34242. Telephone 1-800-926-4797, or, in the Sarasota area (941) 312-0400. Fax (941) 312-0142. E-mail: PWSAUSA@aol.com.

The Ultimate Consumer

What kind of consumer are you? In your recent purchases for food, clothing, or the basic necessities, what did you expect? In every situation as a consumer, when you go to a place of business you expect purchases to always be available. It never crosses your mind that there will not be milk, gas, batteries, or dog food at your disposal. We all have Walmart expectations that there is plenty at "rock bottom prices." Though my concerns for excess in America or starvation and deprivation in the world are real, I would like to emphasize what all of us have come to expect.

Our organization is going through some very positive changes. Throughout the move to Florida, as the board and staff analyzed every function, it was very impressive to see what the organization accomplished. Within our small budget we provide a tremendous assortment of basic necessities and a plethora of information and support services on a daily basis to our members. As our new executive director, Janalee will have as much available as is financially possible, but when a crisis arises the organization will respond with all of our resources. When the chapter, with an even smaller financial base, asks for material and support we will UPS quickly and be there personally as needed.

The move was efficient and—thanks to Janalee and David and all the volunteers (primarily Janalee's family)—we moved without missing a beat. Our membership should be proud that such a major task was made easier by the strength of our organization known as PWSA-USA. Our Prader-Willi ultimate consumer can now shop 'till they drop.

Organization News

Lehman Replaces Koerber on the Board

Shortly after his election to the PWSA board of directors in July, Jim Koerber's job became a full-time overseas assignment, leading him to resign his PWSA post. Wauneta Lehman, who was runner-up in the last board election, has been appointed to serve the first year of Jim's original term. In July 1998, the membership will elect a replacement for the remaining two years of this board position, as part of the annual board elections.

Wauneta and her husband, Bob, reside in Merritt Island, Florida—the "Space Capital"! They are the parents of four grown daughters including Barbara, now 36, who has Prader-Willi syndrome. For the past eight years, Barbara has resided and worked in Wisconsin at the Oconomowoc Developmental Training Center.

Bob and Wauneta have been members of PWSA (USA) since 1975 and are charter members of the Prader-Willi Florida Association, which was formed in 1991. Wauneta has served as chapter treasurer, president, and state board member. Dur-

ing her tenure as president, the chapter was successful in obtaining legislative change so that Florida residents with PWS are recognized as developmentally disabled and can receive services regardless of I.Q. Wauneta was actively involved in planning and operating our national conference in Orlando last summer and now serves on the national PWSA conference committee.

PWSA Secretary Needed

Our thanks to Bronnie Maurer from South Carolina for her year of service as Association secretary. Bronnie resigned the post following completion of her one-year term in July. Treasurer Jim Gardner has been appointed by the board to serve as acting secretary, and Past Secretary Viki Turner has volunteered to take the minutes at the January board meeting in Sarasota. A new volunteer is needed to serve a one-year term as secretary beginning in July 1998.

The Duties of the Secretary

According to the PWSA bylaws, "The secretary shall have the responsibility to act as a recording secretary for

Thanks...

to the Many Volunteers
Who Made Our Move Possible

In St. Louis—

Paula Kollarik
Barb Radinsky & Harriett Radinsky
Wauneta & Bob Lehman (who were visiting from Florida)

In Florida—

Tad Tomaseski
Al Heinemann
Bob & Eileen Comisky
Adele Engel Behar (SCORE)
Ann Durell
Sara Lind
Jennifer Burns
Elaine Bruen
Richard Marks
Dave Zwelling
Dan Walker
Meredith Neirenberg

Gifts in kind—

Janalee & Al Heinemann
Tad Tomaseski

Special thanks to Skip Potter of Columbus, Ohio, who owns the building in which our office is located, for the reduced rate on the lease and donation of office furniture.

the transactions and procedures of the board at all board meetings." There are currently two board of directors meetings each year—one held at the national conference and the other in January. The secretary is required to attend both meetings. Travel and other expenses of the secretary are reimbursed up to \$500 per year. The secretary is appointed for a one-year term, with no limit on the number of terms.

The position requires: PWSA membership; the ability to take accurate meeting notes and to prepare minutes of meetings in a timely manner; and the interest to be involved at the national level and to serve our board and membership.

Interested members are urged to contact Janalee at the national office.

Additional Participants Needed for Study of Behavior Change Medications

Caregivers of persons with Prader-Willi syndrome (parents or direct care staff) are needed to participate in a study of the effects of behavior change medications in the syndrome. The study is part of ongoing research on PWS by Drs. Barbara Whitman of St. Louis University School of Medicine and Louise Greenswag of the University of Iowa Hospital and Clinics. Their goal is to gather systematic data on the administration, effectiveness, and side effects of behavior change medications in at least 100 individuals with PWS.

Participating caregivers will be asked to respond by telephone to a 20-minute structured survey inquiring about these medications. In addition, for those who give permission, the individual's medical records will be requested from the prescribing physician and reviewed. Caregivers who agree to the review of medical records will be asked to sign authorization forms for the release of records. All personal information will be kept confidential.

Summary results of this drug survey will be shared with the study participants (parents and other appointed caregivers), doctors of the individual subjects, and with PWSA (USA).

To participate in the study, please contact:

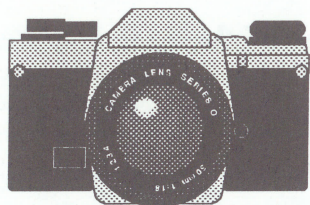
Barbara Y. Whitman, Ph.D.

St. Louis University Hospital Department of Pediatrics

1465 S. Grand

St. Louis, MO 63104

Tel: (314) 577-5609; Fax: (314) 268-6411



Call for Photos

Keep that camera handy ...

The national office of PWSA is looking for more photographs of children and adults with Prader-Willi syndrome—photos we can use in our publications, publicity, and fund-raising efforts. We especially need good close-up shots and action photos. We can use both color and black-and-white prints.

If you have photos that you are willing to share, please send them to our national office in Sarasota. Label each photo with the names of the subjects and give your name, address, and phone number. Also, please enclose a signed note giving PWSA permission to use the photo(s), and tell us if you want to be contacted before we use the photo(s).

PWSA Exhibits at International Genetics Meeting in Baltimore

PWSA (USA) hosted its first-ever exhibit booth at the annual meeting of the American Society of Human Genetics, held October 28-November 1 in Baltimore, Maryland. ASHG is the major U.S. professional association for researchers and clinicians in the field of genetics, and its annual meeting is attended by more than 1,000 members.

It was a timely arrival for PWSA—there were at least 20 different posters and presentations involving Prader-Willi syndrome at this year's meeting, including a morning symposium on imprinting mechanisms, facilitated by Drs. Dan Driscoll (University of Florida) and Rob Nicholls (Case Western Reserve) and a slide session by Dr. Suzanne Cassidy (Case Western) on the phenotypic spectrum of PWS as revealed through diagnostic testing.

The week's presentations generated a great deal of interest in our booth over the three-day exhibit period. Geneticists and genetic counselors from the United States and many other countries stopped by for free literature and to view our display on the faces and characteristics of PWS and the Association's services.

PWSA's exhibit was designed and staffed by members of our Maryland and Metropolitan Washington D.C. chapter. Thanks to Linda Keder, Bob Baker, Bob Gootzit, Jim and Kate Kane, and Nell Elder for taking on this public relations and awareness project.



IPWSO 1998 Conference Packets Available

PWSA has mailed to researchers the registration packet and call for papers for the Third International Conference on Prader-Willi Syndrome, to be held May 21-24, 1998, near Venice, Italy. Members who are interested in attending this conference, but who did not receive a packet, should contact the national PWSA office. The international conference is for parents as well as for scientists and professionals.

Recent Journal Articles on Prader-Willi Syndrome

These articles on medical and behavioral issues were indexed since the February 1997 issue of *The Gathered View*. A full list of 1996 articles and 1997 articles to date, including those on the genetics of PWS, is available on PWSA's web page (http://www.athenet.net/~pwsa_usa/index.html) or by mail or fax from the national PWSA office (1-800-926-4797).

MEDICAL

- Banks, P.A., Bradley, J.C. & Smith, A. (1996) Prader-Willi syndrome—a case report of the multidisciplinary management of the orofacial problems. *British Journal of Orthodontics*, 23(4), 299-304.
- Bassali, R., Hoffman, W.H., Chen, H. & Tuck-Muller, C.M. (1997) Hyperlipidemia, insulin-dependent diabetes mellitus, and rapidly progressive diabetic retinopathy and nephropathy in Prader-Willi syndrome with deletion. *American Journal of Medical Genetics*, 71(3), 267-70.
- Beccaria, L., Benzi, F., Sanzari, A., Bosio, L., Brambilla, P. & Chiumello, G. (1996) Impairment of growth hormone responsiveness to growth hormone releasing hormone and pyridostigmine in patients affected by Prader-Labhardt-Willi syndrome. *Journal of Endocrinological Investigation*, 19(10), 687-92.
- Beccaria, L., Bosio, L., Sanzari, A., Aimaretti, G., Ghigo, E. & Chiumello, G. (1996) GH secretion in Prader-Labhardt-Willi syndrome: somatotrope responsiveness to GHRH is enhanced by arginine but not by pyridostigmine. *Journal of Pediatric Endocrinology and Metabolism*, 9(6), 577-83.
- Brambilla, P., Bosio, L., Manzoni, P., Pirotelli, A., Beccaria, L. & Chiumello, G. (1997) Peculiar body composition in patients with Prader-Labhardt-Willi syndrome. *American Journal of Clinical Nutrition*, 65(5), 1369-74.
- Cento, R.M., Alberti, A., Brancati, A., Raqusa, L., Pettinato, R., Rossodivita, A., Colabucci, F. & Lanzzone, A. (1997) Bilateral non-communicating paraurethral meatus in Prader-Willi syndrome. *Gynecological Endocrinology*, 11(2), 135-7.
- Ebert, M.H., Schmidt, D.E., Thompson, T. & Butler, M.G. (1997) Elevated plasma gamma-aminobutyric acid (GABA) levels in individuals with either Prader-Willi syndrome or Angelman syndrome. *Journal of Neuropsychiatry and Clinical Neuroscience*, 9(1), 75-80.
- Forbes, G.B. (1997) A distinctive obesity: body composition provides the clue [editorial; comment]. *American Journal of Clinical Nutrition*, 65(5), 1540-1.
- Inui, A., Uemoto, M., Takamiya, S., Shibuya, Y., Baba, S. & Kasuga, M. (1997) A case of Prader-Willi syndrome with long-term mazindol treatment [letter]. *Archives of Internal Medicine*, 157(4), 464.

- Kodish, E. & Cuttler, L. (1996) Ethical issues in emerging new treatments such as growth hormone therapy for children with Down syndrome and Prader-Willi syndrome. *Current Opinion in Pediatrics*, 8(4), 401-5.
- Lennox, N., Cook, A. & Slater, H. (1997) Prader-Willi syndrome. *Australian Family Physician*, 26(5), 560-3.
- Robson, W.L.M., Shashi, V., Nagaraj, S. & Norgaard, J. P. (1997) Water intoxication in patient with Prader-Willi syndrome treated with desmopressin for nocturnal enuresis. *The Journal of Urology*, 157(2), 646-7.
- Schluter, B., Buschatz, D., Trowitzsch, E., Aksu, F. & Andler, W. (1997) Respiratory control in children with Prader-Willi syndrome. *European Journal of Pediatrics*, 156(1), 65-8.
- Silvestri, J.M. & Weese-Mayer, D.E. (1996) Respiratory control disorders in infancy and childhood. *Current Opinion in Pediatrics*, 8(3), 216-220.
- Vgontzas, A.N., Bixler, E.O., Kales, A., Cretione, A., Rogan, P.K., Mascari, M. & Vela-Bueno, A. (1996) Daytime sleepiness and REM abnormalities in Prader-Willi syndrome: evidence of generalized hypoparousal. *International Journal of Neuroscience*, 87(3-4), 127-39.
- Weigle, D.S., Ganter, S.L., Kuijper, J.L., Leonetti, D.L., Boyko, E.J. & Fujimoto, W.Y. (1997) Effect of regional fat distribution and Prader-Willi syndrome on plasma leptin levels. *Journal of Clinical Endocrinology and Metabolism*, 82(2), 566-70.
- Wharton, R.H. & Loechner, K.J. (1996) Genetic and clinical advances in Prader-Willi syndrome. *Current Opinion in Pediatrics*, 8(6), 618-24.

PSYCHOLOGY & BEHAVIOR

- Akefeldt, A. (1997) Voice, speech and language characteristics of children with Prader-Willi syndrome. *Journal of Intellectual Disability Research*, 41(Pt. 4), 302-11.
- Dykens, E.M., Goff, B.J., Hodapp, R.M., Davis, L., Devanzo, P., Moss, F., Halliday, J., Shah, B., State, M. & King, B. (1997) Eating themselves to death: Have "personal rights" gone too far in treating people with Prader-Willi syndrome? *Mental Retardation*, 35(4), 312-4. [Note: This article is available from PWSA.]

- Glover, D., Maltzman, I. & Williams, C. (1996) Food preferences among individuals with and without Prader-Willi syndrome. *American Journal of Mental Retardation*, 101(2), 195-205.
- Hodapp, R.M., Dykens, E.M. & Masino, L.L. (1977) Families of children with Prader-Willi syndrome: stress-support and relations to child characteristics. *Journal of Autism and Developmental Disorders*, 7(1), 11-24.
- Joseph, B., Overmier, J.B. & Thompson, T. (1997) Food- and nonfood-related differential outcomes in equivalence learning by adults with Prader-Willi syndrome. *American Journal of Mental Retardation*, 101(4), 374-86.
- Sarimski, K. (1997) Behavioural phenotypes and family stress in three mental retardation syndromes. *European Child & Adolescent Psychiatry*, 6(1), 26-31.
- Watanabe H., Ohmori, O. & Abe, K. (1997) Recurrent brief depression in Prader-Willi syndrome: a case report. *Psychiatric Genetics*, 7(1), 41-4.



Sources: MEDLINE and PubMed databases, National Library of Medicine, and UnCover database, The UnCover Company. Each cite shows: Authors, Title, *Journal*, *Volume* (Issue), Pages. Cites from UnCover show only the first page number of an article.

How to Get Copies of Articles

U.S. copyright law generally prevents PWSA from freely distributing copies of published journal articles. Abstracts (summaries) of many articles are available free of charge on the Internet (through MEDLINE or PubMed—<http://www.nlm.nih.gov/databases/free.medl.html> — Just type **Prader Willi** in the search terms box and click on an article title that is of interest.). Copies of some articles can be ordered via the Internet, provided you register for credit card payment. There is typically a charge of \$10 to \$25 per article to order a photocopy for personal use. Most of the journals can be found in your nearest university medical library. Call 1-800-338-7657 to locate a regional medical library near you.

Growth Hormone Therapy in Children with Prader-Willi Syndrome—A Preliminary Report

Aaron Carrel, M.D., Fellow in Pediatric Endocrinology, University of Wisconsin Children's Hospital, Madison

Susan Myers, M.D., Assistant Professor, Cardinal Glennon Children's Hospital, St. Louis

David B. Allen, M.D., Professor of Pediatrics, Director of Pediatric Endocrinology, University of Wisconsin Children's Hospital, Madison

During the past two years, we have studied the effects of growth hormone (GH) on growth, body composition, and energy expenditure in children with Prader-Willi syndrome (PWS). While anecdotal reports show growth acceleration, improvement in body composition (decreased fat mass), and greater energy expenditure (which allows for additional caloric intake), no controlled studies (i.e., compared with a group of PWS children not receiving GH therapy) have been reported. With the help of a very motivated group of children with PWS, and their families, we have been able to follow 31 children at the University of Wisconsin Children's Hospital in Madison, Wisconsin, and an additional 21 children at Cardinal Glennon Children's Hospital in St. Louis, Missouri. This comprises the largest population of children with PWS studied in a controlled fashion. This study

is still in progress, and we would like to share some preliminary data from our research.

Growth Rate

At the start of the study, all children underwent GH testing and monitoring for baseline growth rate prior to therapy with GH. At baseline, all children had low stimulated GH levels and an average growth rate of 4.3 cm/year (normal growth rate is 6 cm/year). During the first year of the study 60 percent of the children were randomly assigned to begin GH therapy, and the remaining 40 percent did not receive GH. After 12 months of GH therapy, the average growth rate increased to 10.5 cm/year, while the children not on treatment had no significant changes in their growth rate (shown in Figure 1).

Body Composition

Growth hormone has metabolic effects other than increasing linear growth. One effect of potential clinical value in children with PWS is the lipolytic (fat breakdown) effects of GH. During this study children had careful measurements of body composition obtained using DEXA scans. This machine provides a quick and accurate measurement of percent body fat, bone mineral density, and percent lean (muscle) mass. Baseline measurements prior to GH therapy showed elevated body fat in all children, even in those who were not overweight. Prior to GH therapy, the average body fat was 44 percent, compared with 16 percent in age-matched, healthy non-PWS children. One year of GH therapy decreased body fat by 8 percent overall, without significant change in untreated children (Figures 2 and 3).

Resting Energy Expenditure

Using indirect calorimetry to assess resting metabolic rates, PWS children showed markedly reduced energy expenditure at rest compared with non-PWS children matched for body surface area—24.7 vs. 43.1 kcal/m²/hour. There is a trend toward increasing energy expenditure with GH treatment, but this is currently not statistically significant.

Summary

Baseline data from our investigation of children with PWS demonstrates: low GH secretion, markedly decreased energy expenditure, abnormal body composition with percent body fat twice that of normal controls, and normal bone mineral density. Significant improvements in body composition, as well as a trend toward increased energy expenditure, occurred

Growth Velocity Standard Deviation Score in PWS

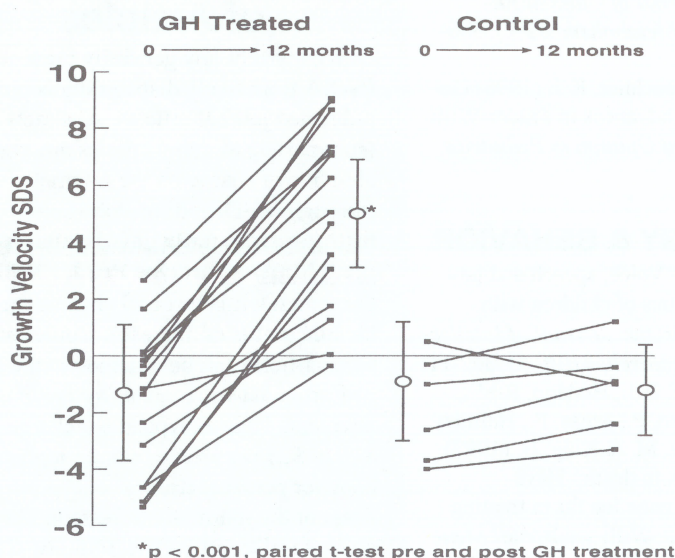


Figure 1

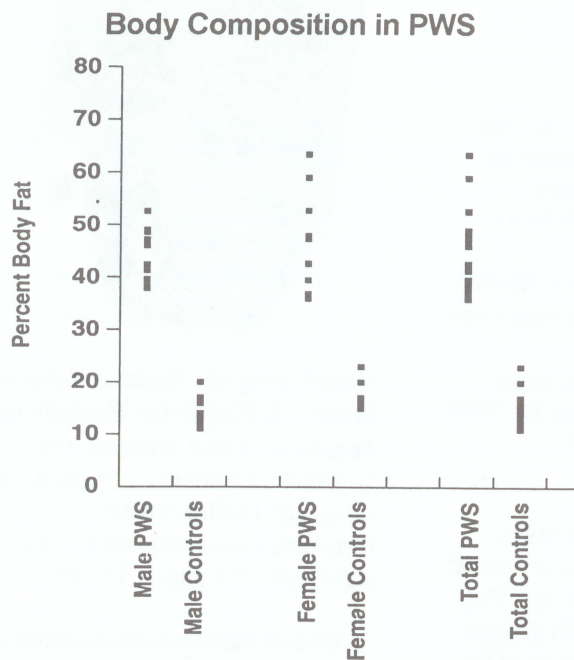


Figure 2

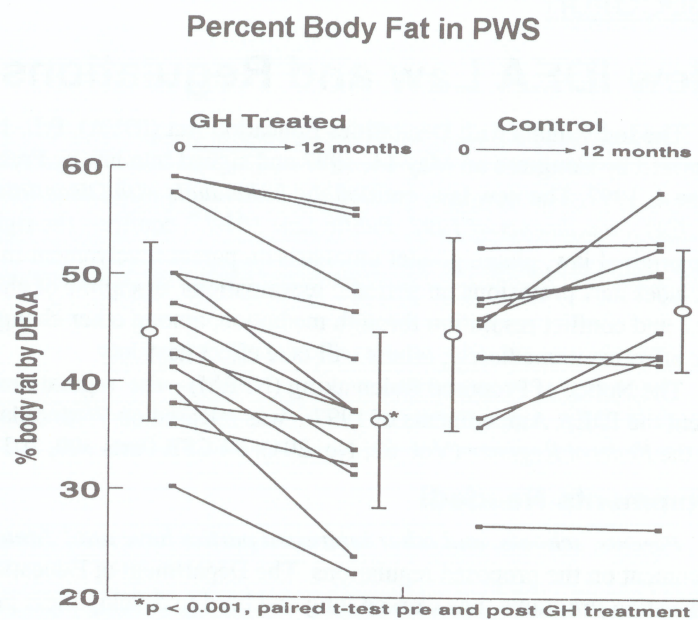


Figure 3

following 12 months of GH treatment. These data suggest that subnormal growth rate, "GH deficient-like" body composition (i.e., markedly increased body fat, reduced lean body mass), and reduced energy utilization may be related to low GH and that children with PWS may benefit from GH therapy.

For More Information on GH Use in PWS—

"Growth Hormone and Prader-Willi Syndrome," by Phillip D.K. Lee, M.D., *The Gathered View*, October 1995.

PWSA Policy Statement on "Growth Hormone Treatment and Prader-Willi Syndrome," approved 7/21/96.

"Growth Hormone Secretion and Effects of Growth Hormone Therapy on Growth Velocity and Weight Gain in Children with Prader-Willi Syndrome," by Moris Angulo et al, *Journal of Pediatric Endocrinology & Metabolism*, Vol. 9, 1996, p. 393-400.

Also see articles by Beccaria, Brambilla, Kodish, and Wharton in the listing of recent journal articles on page 4 of this issue of *The Gathered View*.

A Family's Experience with Growth Hormone

Our son Jesse, who is now 18 years old, was diagnosed as having PWS when he was about 2 or 3

years old. Jesse was around 10 years old when tests revealed that he had almost stopped growing. He was 128 cm [51.2 in.] at that time, weighing about 50 kg [110 lbs.]. His pediatrician suggested growth hormone might be an option, and after much discussion we decided it would be worth giving Jess the opportunity to reach a more normal height.

He had to undergo some extensive tests which involved being in the hospital for one day and having blood taken every half hour or so. Jesse was not at all worried by this and was watched over all the time and even had a chocolate milkshake at the end! Jesse began treatment on April 7, 1989, just before his 11th birthday. To begin with, it involved a daily injection, which was a bit daunting in the early days, but it was arranged for a nurse to visit each day. A week or so later I was able to give Jess the injection myself, and after a very short time he was able to inject himself. This at first involved having to mix the solution and use a syringe, but after a couple of years they introduced the "pen," which made both mixing the correct amount and the actual injection much easier.

Jess's height and weight were then closely monitored with regular visits to his doctor. He also had regular x-rays to check on his bone-age and to check that the correct dosage was still being used. He continued using the growth hormone for the next six years and grew at a fairly steady rate. On completion of these injections he had reached 166 cm, which certainly made him a more normal height. He had also put on a fair amount of weight, which is a fairly natural occurrence at that age, even for people without PWS. He is quite a strong boy and able to play sports—netball, cricket, etc.

To date we have never experienced any "side effects" from the use of growth hormone. I think it helped Jess feel more confident being taller, and it never bothered him at all having to give himself injections. It became a fairly routine thing as far as our family was concerned. If we were going anywhere, or Jess went away by himself, he was able to take his injections with him as part of his daily routine.

If anyone is considering the use of growth hormone, although it will not be an easy decision and there are lots of things to consider, I hope that this letter may be of interest and maybe of some help.

—Nan Adams, Australia

(Reprinted from *Wavelength*, Volume 5, Issue 2, October 1996, the newsletter for parent representatives to the International Prader-Willi Syndrome Organisation—IPWSO.)

New IDEA Law and Regulations

The Individuals with Disabilities Education Act (IDEA), P.L. 101-476, was reauthorized by Congress on May 14, 1997 and signed into law by President Clinton on June 4, 1997. The new law, entitled the *Individuals with Disabilities Education Act (IDEA) Amendments of 1997, Public Law 105-17*, confirms the rights established in the original law, places greater emphasis on parent involvement in the process, and includes new provisions on periodic reevaluations, discipline of children with disabilities, and conflict resolution through mediation, among other changes. Some provisions are effective immediately; others will take effect next July.

The Notice of Proposed Rulemaking (NPRM)—the regulations that will implement the IDEA Amendments of 1997—was released on Wednesday, October 22, 1997, in the *Federal Register* (Vol. 62, No. 204): 34 CFR Parts 300, 301, and 303.

Comments Needed!

Parents, schools, and other interested parties have until January 20, 1998, to comment on the proposed regulations. The Department of Education, Office of Special Education Programs, has worked long and hard to develop these proposed rules. They need and have asked for help. Advocates for children with disabilities should address any areas in which there are concerns and/or areas where new language is needed for clarification. We also need to let them know what is good and why.

Comments should be addressed to: Thomas Irvin, Office of Special Education and Rehabilitative Services, U.S. Department of Education, Room 3090, Mary E. Switzer Building, 330 C Street, SW, Washington, DC 20202.

Comments can also be submitted via e-mail at comment@ed.gov. Use the words "Assistance for Education" in the "Subject" line of your message.

Internet Users—

For the full text of the new law and proposed regulations, try these two Web sites:

<http://www.ed.gov/offices/OSERS/IDEA/index.html> (the Office of Special Education and Rehabilitative Services, Department of Education) or

<http://www.lrp.com/ed/> (Education Administration Online).

For a brief summary of the changes in IDEA, check out:

<http://www.cec.sped.org/pp/idea-a.htm> (the Council for Exceptional Children),

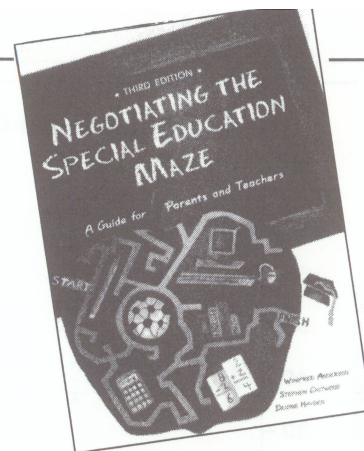
<http://www.fcsn.org/tapp/idea.htm> (the Federation for Children With Special Needs), and

<http://www.npnd.org> (National Parent Network on Disabilities—NPND).

To get a copy of the regulations by mail, call 1-800-USA-LEARN and request a copy, send an e-mail request to laura.blackprice@ed.gov, or order from the U.S. Government Printing Office (\$8.00 each): Superintendent of Documents, PO Box 371954, Pittsburgh, PA 15250-7954, Telephone 202-512-0000.

SSI Alert

By August 30, more than 120,000 children had been dropped from the Supplemental Security Income (SSI) program as a result of eligibility reviews under the revised welfare law and new qualifying rules published by the Social Security Administration last February. Families qualify for SSI if their child meets the criteria for having a disability and if their income and resources are below a certain level. Changes made in the last year make it much more difficult for children to meet the disability criteria. In addition, many children who have lost SSI are also being wrongly denied continued Medicaid coverage in their states. Any family that receives notice that SSI payments will be stopped should file a "Request for Reconsideration" within 10 days, if possible, and call the SSI Children's Project Hotline at 800-772-1213 for free legal help.



Negotiating the Special Education Maze—A Guide for Parents and Teachers (Third Edition), 1997, by Winifred Anderson, Stephen Chitwood, and Deidre Hayden, Woodbine House (800-843-7323), paperback, 264 pages, \$16.95.

One of the best tools available for parents of school-age children with disabilities just got even better. The 1997 revision of *Negotiating the Special Education Maze* contains new chapters on "Early Intervention: Beginning the Journey" and "Nondiscrimination Protection: Alternative Routes through the Maze" (specifically, how two laws besides IDEA—Section 504 of the federal Rehabilitation Act of 1973 and the Americans with Disabilities Act of 1990—provide additional rights and protections in the education arena). In addition the book has a larger, more user-friendly format, with lots of sample charts and worksheets to make the advice concrete.

Although the revision was published before the IDEA Amendments were enacted, its content remains valid: its major focus is on working with the school system to identify the child's specific needs and create a program that will meet those needs. Thirteen chapters cover the entire process—referral & evaluation, eligibility decision, IEP program and meeting, early intervention and IFSPs, transition planning, due process, and monitoring the program. Also included are tips on working with your school system, observing your child at home, interpreting school records about your child, preparing for meetings, and building alliances with other parents for support and to effect changes.

We Count, Too!

Remembering Our Siblings

by Janalee Heinemann

At the 1997 PWSA(USA) national Prader-Willi syndrome conference in Orlando, we tried to place special emphasis on siblings by: 1) placing siblings in separate groups; 2) having two panel sessions where young adult siblings told how living with PWS impacted on them; and 3) having a sharing session for siblings. Leading the sharing session and moderating the panels made me realize that what we did was only a drop in the "bucket of support" we should be giving our siblings. Our siblings do count, too—but do they ever count first?

The sharing session was attended primarily by siblings ages 12-14 years—ages difficult even in ideal circumstances. I was touched with how open they all were in expressing their feelings. When I asked the group if they ever shared like this with their friends at home, they all said emphatically, "No!" They were too embarrassed to tell their friends all that went on in their homes, leaving them isolated with their feelings. Let's face it, at that age you don't want to be different, and although their friends complain about their brothers and sisters, those friends don't usually have locks on their refrigerators, siblings who steal from them, siblings who have dramatic tantrums in public, siblings who pick at sores, etc.

Resentment and guilt were common feelings discussed. One sibling complained that she had to be responsible for her young sister who has PWS. The rest of the group all "jumped" on that complaint and said, "How would you like to be us?"—they all had older siblings with PWS, and all felt they were expected to be responsible for them. They also felt that their brother or sister with PWS often got by with much more than they did, and they were acutely aware of their parents' double standard on discipline, chores, achievements, and tolerated behavior. We discussed how it was much easier for a parent to reprimand them versus their PWS sib. They had to admit that the consequences of coming down hard on their sibling would be something they

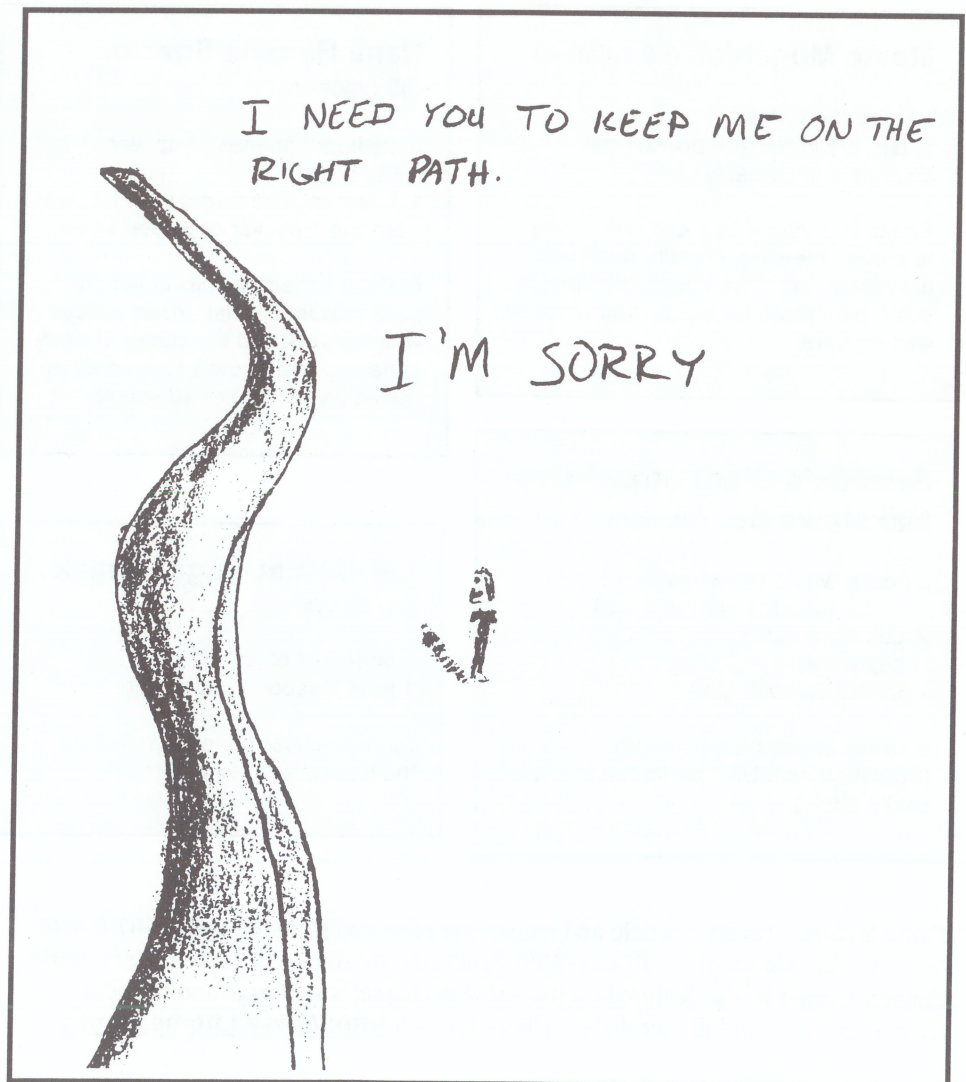
wouldn't want to deal with, either.

The group was able to laugh when relating their most embarrassing moments, and all were able to admit that there were times they did or said mean things to their brother or sister with PWS—yet at times stuck up for them around other kids. All expressed relief that they were not alone in their feelings and living in such "unique" circumstances. They said they would like more sharing sessions and suggested a "chat room" on the Internet—which we are pursuing.

On the adult sibling panel, we had three outstanding young women (a male sibling had to cancel): Denise Miller and Melissa and Jennifer Robertson. Af-

terwards, reflecting on the session, Denise wrote, "Guilt in my opinion is what siblings suffer with the most." She also said that she was glad to see that the younger generation of siblings is being informed about the syndrome at a younger age and wrote, "This knowledge will be what helps them understand the situation better and will help open up hearts and minds to feel and give more compassion to their siblings with PWS and also to themselves."

There is more to share from our sessions, but I will save it for another article. In closing, I would like to share with you a card one of our siblings wrote which expresses far more in a few words than I could ever say:



Calorie Control

While Karie was growing up, I kept her weight under control with strict calorie counting. My favorite calorie book is the one called "Food Counts," which is available at many Walgreen's stores. Karie is a master mathematician and has become a calorie-counting expert. This keeps her busy during meal preparation. I am aware that Karie thinks about food all the time and often asks about the next meal and recounts what she has eaten for recently consumed meals. Accepting her preoccupation with food, I try to make her restricted diet easier to follow by allowing her the freedom of having something to eat whenever she feels hungry. All she has to do is ask, and I'll give her a snack. There are so many items to offer, like dill pickles, air-popped popcorn, sugar-free popsicles, and mini rice cakes. As long as Karie knows that she can have something to eat if she only asks, this seems to minimize incidents of unauthorized food consumption and seems to lessen her obsession with food. Using this philosophy (along with locked cupboards and an alarm on the refrigerator), I was able to help Karie keep her weight under 100 lbs. up to the age of 16, when she entered a group home. Even now, she is slim and trim at 114 lbs., following a diet of 1,400 calories a day and a daily exercise program.

—Teresa Kellerman

A Sampling of Karie's Special Treats

Movie Munchies (50 calories)

1 cup puffed wheat
½ tsp. dry lemonade powder mix
1 sprinkle of Sweet'N Low

Shake in a zipper bag and take along to movie, meeting, church, doctor appointment, etc. This keeps little fingers busy, minimizes tantrums, and reduces waiting time!

Sweetie's Giant Strawberry Milkshake (100 calories)

5 frozen whole strawberries
(about 1 cup, or 5 oz.)
½ cup skim milk
½ cup water
1 packet Sweet'N Low

Blend in blender until smooth.
(Strawberries MUST be frozen to make shake thick.)

Nana Banana Snacks (30 calories)

3 medium bananas (4 oz. each), cut into quarters
1 T. lemon juice mixed with 1 T. water and 1 packet of Sweet'N Low

Roll the banana pieces in lemon juice mixture to coat. Insert a clean wooden stick into the center of each banana piece. Freeze on a sheet of waxed paper. Makes 12 snacks.

Let-Us-Eat-Sugar Snack (20 calories)

1 large leaf of lettuce
1 level teaspoon real sugar

Sprinkle sugar on lettuce. Roll up the lettuce leaf. Nibble away!

Avoiding Food Rewards in the Classroom

Sometimes it seems there's junk food lurking around every corner of the school. If it's not birthday or holiday parties, it's teachers giving candy as prizes or incentives for good work and behavior. The following wisdom, from the mother of a child with serious food allergies, can be helpful to Prader-Willi parents who are trying to curb the edible rewards at school:

"It has been my experience that children cherish the concrete treasures they earn and have long-lasting memories which they attach to them. When they have something to show for their hard work, it is an opportunity for a conversation with Mom and Dad. They may not save the wrapper from the candy, but they do bring home the stickers. ...

"There are so many alternatives to food rewards. ... Stickers and stars work very well for younger children. As they get older, youngsters appreciate interesting pencils, pens, erasers, and coupons for special treats. These might include lunch with the teacher, extra computer time, no homework for a night, or a lesson in a favorite area such as airplane-making. Homemade badges, pins and refrigerator magnets can be made up and distributed as appropriate. Computer-made certificates of merit are also good choices. Promotional items from stores, agencies, radio stations and other retail dealers can be used. Consider also recycled products such as books, left-over party favors, and small toys. There are several books available listing "free" items for kids. Writing letters to companies requesting their free products can be part of the language arts lesson.

"Make a pledge this year to become involved with your school on a treasure hunt for non-food rewards. Work with other parents, grandparents, school personnel (especially the PTA) and do not forget the children themselves. You will be surprised at some of the great ideas they come up with, and they are the ones who benefit the most!"

("Back to School," by Wrenn Reynolds, *New Developments*, Vol. 2, No. 1, The Developmental Delay Registry, Chevy Chase, MD)

Editor's Note: Teresa's article and recipes are reprinted from the *Prader-Willi Nilli News*, the newsletter of the Prader-Willi Syndrome Arizona Association. Her Calorie Counter Corner is also featured on the Arizona chapter's web page and includes recipes for lunch and dinner dishes. Check it out at <http://pwsaa.home.ml.org>

On Hoarding

In the last issue of *The Gathered View*, the mother of a 34-year-old sought advice on how to reduce her daughter's mounting collection of "stuff" without causing serious setbacks in behavior. Her daughter, who lives in a group residence with three young men with PWS, has filled her room with piles of things she's intent on keeping.

We heard from only one family who's had a similar experience with their daughter in a group home—What worked best for them, they reported, was to go to their daughter's group home and clean out her room when she's gone. "We pitch and pitch; she never misses it at all," they wrote.

While apparently there were no serious repercussions in this case, such action could result in serious consequences. We asked **Dr. Barbara Whitman**, a behavior expert on our Scientific Advisory Board, if she could suggest an alternative solution that would recognize the rights of an adult while at the same time accomplishing the goal of reducing clutter.

Dr. Whitman recommends an approach based on establishing clearcut and logical rules. She writes:

"I have dealt with this with several adults, and the way I do it is to talk when they are calm about how the rules allow either so much space or so many items. If any person reaches the 'allowable limit,' then the rule is that before another item can be brought in, something has to go.

"For example, one adult collected magazines. She had two rooms full. We sat down and said: Number one, that's a fire hazard, and the fire marshal has limited the space (that wasn't an untruth), so only one of those areas could be used. In addition, the rules would be that when the area was full, something had to go before more was brought in.

"Of course there is always initial grouching, and the first clean-out can bring tears and tantrums, but once through the initial time, the rules can be regularly enforced."

Would any residential providers care to share their house rules concerning storage of personal possessions?

A Lesson in Expectations ...

Andrew Never Listens!

by Brian Norton

My son Andrew is as hard-headed as a Missouri mule. He will argue about anything!

Andrew's grandparents have an inground swimming pool and my wife, Cheri, has made sure all her children can swim. As a matter of fact, Cheri was a lifeguard in her younger days, as my daughter is now. So Cheri taught Andrew to jump off the diving board and dog paddle to the ladder at a distance of three feet.

One typical Sunday afternoon all the grandchildren were at Grandma's swimming in her pool. All of the older grandchildren were jumping off the diving board, so Andrew started jumping off also. Suddenly, instead of going to the ladder, Andrew swam for the shallow end, which is about 20 feet. I held my breath as he paddled and swam.

When he finally made it to the shallow end, I told him he couldn't make it and don't ever do it again. He said, "I can do whatever I want." That sunk in my head ... I thought to myself: Why insist he couldn't make it when he obviously could! After 10 times from diving board to the shallow end, I finally gave up trying to tell Andrew he couldn't.

Because he never listens, anyway, thank God.

(Editor's note: Brian Norton has written for The Gathered View about "being a dad," and he often writes humorous and hopeful articles about his son with PWS in the Missouri chapter newsletter, The Missouri View. This article is reprinted from the September 1997 issue.)

Medical Problems of Aging with PWS

The mother of a 43-year old man with PWS contacted the national office regarding age-related medical problems her son was developing, particularly nighttime incontinence problems. She would like to talk with other parents of older adults with the syndrome to compare medical and physical issues. Interested families are urged to contact the national PWSA office to be put in touch with this parent.

Editor's Note: At a 1996 conference, Dr. Suzanne Cassidy, chair of PWSA's Scientific Advisory Board, reported findings of a study of 22 persons with PWS over age 30. Although bedwetting was common, she reported, this problem usually improves with age.

Send questions and answers for this feature to *The Gathered View* c/o the national PWSA office. See page 2 for our address and phone.

People with PWS Who are Physically Aggressive

The aunt of someone with PWS, "who has numerous problems because of his physically aggressive behavior," would like to see an article about individuals who have this behavior problem. "I would like the comfort and support of knowing that there are other physically aggressive people out there," she writes. Families who are willing to share their stories—and solutions for curbing this behavior—are urged to write us in care of the national office.

Speech Therapy Techniques That Don't Involve Food

Several families of young children have asked for alternative activities to the many early intervention "games" that involve food. Parents and professionals: Do you have some good ones to share?

In Recognition of Our Contributors

(JULY 4—NOVEMBER 1, 1997)

1996-1997 ANGEL FUND

Heavenly Angel (\$501-\$999)

Paine Webber (Mr. & Mrs. Hal L. Burnett, Doris Jane H. Miller)

Arch Angel (\$251-\$500)

Jim & Jane Carlson

Cherub (Up to \$100)

Melissa Janquart
Raymond Mears
Regina R. Murphy
Peggy J. Smith
John Talanian
The Sunlighters First Congregational Church



OTHER DONATIONS

Patron Membership (\$100 or more)

Mr. & Mrs. D. P. Burleigh
Anne & Steve Manning
Terry & Debbie Mleczewski
Mrs. R. Lorraine Notbohm
Steve & Verla Percy
Dr. Maurice Schenk
Robert & Marjorie Stickle
Maria Subira-Nadai

Contributing Membership (\$50-\$99)

Edward & Amanda Kavanaugh
Mr. & Mrs. Thomas LaBossiere
Karen Lewis
Greg & Sandy Olson
Redford Opportunity House
Maureen Serrano
Henry & Susanne Singer
Mrs. Emery Wertz
Liz Whigham

Research Fund

Stuart & Marnie Boyd

In memory of Mrs. Chippie Alterman:

Abe & Elizabeth Alterman
Nan & Bill Curlee
Rita Davis Doyne & Family
Sandra & Burt Epstein
Georgia Association for Prader-Willi Syndrome, Inc.
Lana & Richard Krebs
Mr. & Mrs. Irving Libowsky
Hal & Jo Meeks
Roy & Mable Meeks
Pat & Don Mendel
Fred & Jeannette Nichols

In memory of Sam P. Alterman:

Jayne & Irving Adair
Abe & Elizabeth Alterman
Bruce & Sally Alterman
Louis & Julie Alterman
Mr. & Mrs. Neil L. Aronstam & Family
BM&M—Bruce Kaufman, Richard Mendel, Donald Minsk and Partners & Staff
Julius & Vicki Blahut
David & Robin Blass
Fannie M. Carl
James & Carolyn Caswell
Champion Building Services—William C. Lomel II, President
Mr. & Mrs. Gerald Cohen
Pauline & Gilbert Cohen
Nan & Bill Curlee
Debbie & Don Davidson & Family
Mr. & Mrs. Marvin Davis
Rita Davis Doyne & Family
Sandra & Burt Epstein
Doris & Harold Frank
Laurie Freeman Gidlow
Mr. & Mrs. Jonathan Golden
Ellen & David Herold
Jack & Gladys Hirsch
Bruce M. Kauffman
Rita & Sol Kimerling
Lana & Richard Krebs
Joy G. Kunian
Mr. & Mrs. Irving Libowsky
Frances & Sylvan Makover
Lenore S. Maslia
Bronnie & Stewart Maurer
Hal & Jo Meeks
Roy & Mable Meeks
Don & Pat Mendel
Russ & Pat Myler
Mr. & Mrs. Gene Oberdorfer
Sanford & Barbara Orkin
Zac Pasmanick
Frank Pridgen, Jr.
Mr. & Mrs. Charles H. Shepherd
Abe & Linda Schear
Mr. & Mrs. Harry Solomon
Mr. & Mrs. Nelson Stewart
Beth & Edward Sugarman
SunTrust Bank—Trust & Investment Services
Noam & Susan Zelman
Robert R. Zerden

Operating Fund

William & Janet Arner (*in memory of Susan Sides*)
Robert Baker (United Way of Central Maryland)
Shirley & Hal Burnett (*in honor of Ben Keyfetz' 95th Birthday; in honor of Margie Oberfelder's Special Birthday; in honor of Jo Lane; in honor of Audrey Feldman*)

Schelle Childress (*in memory of Renee Davis*)

Computer Sciences Corp. (United Way Los Angeles Area)
Steven A. Diaz (United Way of Washington DC)
Catherine M. Feerrar (United Way of Washington DC)
KPMG Peat Marwick (United Way Los Angeles Area)
Aulga & James Mahar (*in memory of Marion Dancey*)
Carol Matza (*in memory of Julien Remy Jolivet*)
Eric Miller
Linda Panagoulis (United Way of Washington DC)
Mr. & Mrs. Andrew Russo (*in memory of Mae Ceppos*)
Curtis & Marion Shacklett, Inez Shacklett (*in memory of Janie Hughes*)
Virginia Sheeron (*in memory of Danny Robin*)
Linda M. Steppe (United Way of Southeastern PA)
John & Marie Thomas (*in memory of Gertrude Baugh*)
Stephen G. Timney (United Way of Southeastern PA)
Jean Till (*in loving memory of Lynn*)
Roger West

Ronda West

In memory of Kendrick J. Smith (Bronnie Maurer's father):

Zoe E. Barker
Mrs. Robert S. Bloomer
Dr. & Mrs. Gerald J. Duffner
S. Dudley Hallagan, Jr.
Helen Hamilton
The Highlands at Pittsford
Rev. Walter R. Hobkirk
Mr. & Mrs. James McDonald, Jr.
Eleanor D. Pennington
Robert & Patricia Place
Prop-Care, Inc.
Evelyn P. Rayfield
Benson & Edna Shultz
Eleanor Stuart
Ann L. Warner
Norman L. Waterman Funeral Home

Conference Contributions

Jennifer A. Gomez
Paul & Donna Weisner (*in memory of Frederick R. Krause*)
Lucille Meredith

1997 Awareness Week

Georgia Association for Prader-Willi Syndrome, Inc.

Many Ways to Contribute

PWSA (USA) has an ongoing need for financial support. There are many ways for members to help:

- ◆ Participate in our annual Angel Fund campaign, and invite relatives and friends to become part of this once-a-year major fund-raiser. (You'll be receiving our 1997-98 request soon.)
- ◆ Include a donation with your membership renewal at the Contributing or Patron level.
- ◆ Send a contribution in honor of a special person or event or in memory of a loved one. (We'll send a tribute card notifying an individual or family of your donation.)
- ◆ Designate your United Way or other community fund donation for PWSA. (Ask your fund drive organizers how you can do this.)
- ◆ Include a bequest to PWSA (USA) in your will.

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

