January-February 1999
Volume XXIV, Number 1

The Gathered View

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

Winter Board Meeting Highlights

By Barb Dorn, PWSA (USA) President

I hope everyone is surviving his or her winter season. For many of us, cabin fever definitely starts to kick in around this time. I keep telling myself, spring is just around the corner.

I wanted to share a few of the highlights from our winter board meeting that took place on the weekend of January 8. We spent over 12 hours in meetings where a lot of sharing, hard work, and future planning took place.

The PWSA (USA) board of directors and officers meet twice a year, once during national conference time and again in January.

Board members meeting in January welcomed a visit from a Canadian family who were vacationing in Sarasota. Above, with local interpreter Lisa Sabourin (left), Robert, Genevieve, and Luce Bolduc of Quebec. They had never before met another PWS family, and Genevieve, 26, speaks only French.

Changes in Officers

Before I share some of the changes that occurred in our officers, I first want to remind you of how the election of officers takes place: The general membership votes for the 12 board members who all hold three-year terms. Anyone is welcome to nominate themselves or someone they know who is interested and willing to serve our organization. Officers, however, are nominated and elected by the board of directors. The Leadership Committee conducts a search and attempts to come up with a roster of qualified, interested individuals. An officer's term is typically a three-year term with an annual evaluation.

During our January meeting, **Jim Gardner** stepped down as treasurer. He will continue to serve our organization as a board member but felt he needed to leave this position. Jim and his wife, Joan, have been very active in making plans for the 2001 International PWS Organisation (IPWSO) meeting, which will be held in Minneapolis in conjunction with our national conference. Board Chair **Jim Kane** offered his services to replace Jim Gardner as treasurer. Jim is an accountant and former PWSA treasurer, and has worked closely with Jim Gardner on accounting issues for our organization. The board voted and approved this change.

With taking on the treasurer's duties, Jim Kane felt it was necessary to step down as chairman of our board, a position he has held for more than five years. **Ken Smith** was then nominated for and elected to this position. **Don Goranson** also stepped down from his position of vice president. In July, Don had expressed his willingness to remain in that position until another candidate could be found. He will continue to serve us as a board member. **Mary Lynn Larson** was nominated and voted in as our new vice-president. Mary Lynn is the mother of 6-year-old Alex, who has PWS. She is a pediatric speech pathologist for a hospital in Green Bay, Wisconsin. Her husband, Mike, is a PWSA board member.

I want to take a moment to address any questions or concerns that may come to mind with having a husband and wife serve on the national team. Mary Lynn will not be a voting member, and Mike will not be involved in any decisions or voting measures having to do with Mary Lynn. Mary Lynn has been a very active, enthusiastic member of both the national and her state organization since 1992. She is *willing* and *eager* to become a part of our team. For those of you involved in recruiting volunteers, you know how important it is to have that willingness and enthusiasm.

Another concern that may be voiced is the fact that Wisconsin will have three representatives at the national level. With

(Continued on page 3)

inside

The Prader-Willi Syndrome Association (USA)
5700 Midnight Pass Road, Suite 6
Sarasota, Florida 34242
www.pwsausa.org
1-800-926-4797
9:00 a.m.–7:00 p.m. Eastern Time
Local: 941-312-0400
Fax: (941) 312-0142
E-mail: pwsausa@aol.com

Executive Director's View

PWSA (USA) and Volunteerism:
From the Acorn to the Great Oak Tree

By Janalee Heinemann

Recently, I was asked by our new auditor, "*Does PWSA (USA) use volunteers?*" As I began to explain and try to define the role of volunteers in my own mind, I realized that every inch of PWSA (USA) "soil" is permeated with volunteerism. Almost all of the accomplishments by the Association throughout its 24 years have been thanks to the volunteer efforts of hundreds of parents and professionals.

The first seed of the organization began with the efforts of our volunteer founders and PWS parents, Gene and Fausta Deterling. From there sprouted a volunteer board of 12 members and four officers; the first volunteer editor of *The Gathered View*, Shirley Neason; and the first executive director, Marge Wett, who initially was unpaid. Nurtured by caring professionals, and made strong by the determination of families to fight for a better quality life—both for themselves and their children—PWSA (USA) has grown and branched out to more than 30 state and regional chapters, 12 committees, and several teams, also composed entirely of volunteers.

Another major "branch" is the yearly national conference, which is organized and staffed by the host state chapter volunteers under the guidance of the executive director and a volunteer conference committee. The presenters at the conference also volunteer their time and most of their travel expenses. The complex youth program at the national conference (consisting of hundreds of children and adults with the syndrome) is directed by paid consultants, but staffed entirely by volunteers.

All 28 booklets, 12 brochures, and our hardbound management book have been created and edited by volunteers. Many volunteers have burned the midnight oil to complete an article or edit a brochure or booklet for PWSA (USA). Even our new poster was created and produced by a volunteer parent. Fundraisers for PWSA (USA) have been successful thanks to local and national volunteer efforts.

I envision each state chapter as a separate new "branch" that has sprung from the "trunk" of PWSA (USA) and is also under the guidance of volunteer officers and boards, and each family as a "leaf" that needs both the "branch" of the state chapter and the "trunk" of the national office. The "roots" by which we all exist are the basic ethics, bylaws, and policies of all good nonprofit organizations.

As mentioned in the last *Gathered View*, our "blossoms" are our local Sarasota volunteers, who assist the staff with the daily running of the office. They route calls from the crisis hotline, log incoming mail and donations, assist with mail-

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 (keder@erols.com)
Associate Editor: Lota Mitchell (ljecholsm@juno.com)

Communications regarding *The Gathered View* or PWSA membership and services should be directed to the national office of PWSA (USA), 5700 Midnight Pass Rd., 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. Home Page: www.pwsausa.org

Volunteerism—*continued*

ings, label brochures, assist with the bereavement follow-up program, assist with general organization of the office, copy and bind booklets, and work on special projects.

PWSA (USA) could not and would not exist without the thousands of hours of volunteer efforts donated to the Association monthly. Each new "fruit of our labor" is thanks to the many, many parents who stayed up too late on a work night to complete a project for their state chapter or national, or thanks to a professional who went into work on a Saturday to help with a support group or complete an article they promised to write or edit, or thanks to a retired Sarasota resident who chose to spend a day in our office rather than going to the beach.

The spirit of volunteerism is *more* than important to PWSA (USA) — it is the sunshine, water, and soil on which the Association exists and thrives.

Call for Nominations

PWSA (USA) Board of Directors

Each summer at the PWSA Annual Meeting, four seats on the 12-member board of directors are filled through a general election. For the July 1999 elections, the Leadership Committee requests that names of members interested in, or recommended for, a position on the board be submitted no later than March 31, 1999. Board terms are three years.

Recommendations should include a brief description of the candidate's qualifications to serve on the board.

Send recommendations by mail, fax, or e-mail to the attention of the Leadership Committee Chair, c/o PWSA (USA), 5700 Midnight Pass Rd., Suite 6, Sarasota, FL 34242. E-mail: PWSAUSA@aol.com; Fax: 941-312-0142

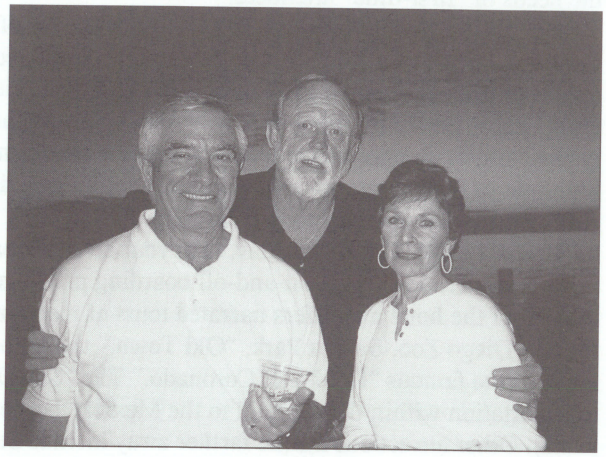
President's Message—*continued from page 1*

Mary Lynn and I located in the same state, I see the opportunity for us to get together for work sessions with little or no cost involved. Planning and coordinating activities for a national organization can be complicated. It really requires a strong team effort. In July of this year, our board will have four vacancies. This will be a great opportunity for others who may be interested in sharing their talents to come forward and take on this challenge. We are especially looking for people from the western part of the country. If you are interested, contact our national office.

Above, PWSA Executive Director Janalee Heinemann, Board Member Barbara Whitman, and President Barb Dorn on break at the mid-winter meeting.
Board Member Paul Alterman, center below, keeps a firm grip on our 1999 PWSA National Conference Co-chairs, Frank and Fran Moss. Fran is also a national PWSA board member and executive director of the Prader-Willi California Foundation.

Update on National Conferences

We briefly reviewed the 1998 conference and were pleasantly surprised to learn that we did not experience a deficit, thanks to the extraordinary fund-raising efforts of the Ohio chapter. We continue to take steps to strengthen our financial support and evaluate our expenses for the conference.



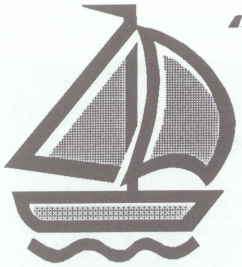
Planning continues for the 1999 conference in San Diego. Many details were discussed and finalized for this year's conference—Providers' Day, Scientific Day, the Youth/Adult Activities Program, as well as the main conference. Many hours of hard work go into this task, and we thank the Prader-Willi California Foundation for all their efforts.

The board of directors approved the Pennsylvania Chapter's conference proposal bid for the year 2000 in Pittsburgh.

In 2001, we will be hosting the International Prader-Willi Syndrome Organisation conference in Minneapolis, Minnesota. Many hours of planning need to be done to accommodate the need for various language translations as well as other cultural and logistical needs.

The board agreed to contract with Mark Eason from Goodwill Industries to help us with a strategic planning process, and Janalee will continue to seek funding for this contract. [See "A Challenge to Chapters, Members," page 5.] Our organization continues to grow at an amazing rate. We need to stop and evaluate what we are doing, how we are doing it, and where we want to go in the years ahead. This consultation will help us document and prepare our organization for the future, a task that must be accomplished in order to seek and obtain national funding and grants.

It was a very productive weekend. Many of us waved a quick goodbye as we raced from the meeting to catch our flights home. It was good to work together as a team, to move forward in order that we may better serve you and all persons who have PWS.



"Sailing into the 21st Century"

21st Annual PWSA (USA) National Conference

Westin Horton Plaza, San Diego, California

July 7 – Scientific and Service Providers Conferences
July 8-10 – Parent-Professional Conference

Make your plans now to attend the 21st Annual Prader-Willi Syndrome Association National Conference in beautiful San Diego! This will be the third national conference in California and the second to be held in San Diego. Schedule your visit to spend some extra time enjoying this beautiful city—its wonderful climate and vacation attractions. (Average July weather: temperature – 76° high, 65° low; humidity – 74%; sunshine average – 68%; rainfall – 0.01 inch.)

Plan also on a great conference experience. This is a wonderful opportunity to learn, but also to renew friendships and make new friends. Our goal for this year's conference is to offer a program to cover areas of most interest to all, but to be especially sensitive to the needs of "first-time" attendees.

Our national conferences are about—and because of—individuals with Prader-Willi syndrome. Each year we plan a professionally organized special day program for them (and their siblings) called the Youth and Adult Activities Program (YAAP). This program begins on the morning of July 8th and continues until noon on July 10th so that the parents and/or caregivers are free to attend the conference sessions. Friday night is the highlight of their program with a special banquet and dancing. (The youngest attendees will have nursery and pre-school care.)

Come Early! On Wednesday, take your own self-paced tour of San Diego on the "Old Town Trolley," with on-and-off boarding privileges at every stop. The trolley stops in front of the hotel and offers narrated tours of many of the major attractions, including the San Diego Zoo, Balboa Park, "Old Town," the harbor and Coronado Island, the home of the famous "Hotel Del Coronado." The "San Diego trolley" provides light-rail transportation within the city and to the Mexican border (for those who wish to visit Tijuana). Other attractions a little farther away include Sea World, the Birch Aquarium, Wild Animal Park, and LegoLand.

HOTEL—WESTIN HORTON PLAZA, 1-800-6-WESTIN

Make your hotel reservations early—The special room rate of $115 during the conference is also available before and after the conference … from June 30 to July 13, subject to room availability. (Don't forget to reserve cribs or rollaway beds, if needed.)

TRANSPORTATION

Make your plane reservations early—remember the weekend prior to the conference is a holiday weekend! You won't need a car during the conference! The Westin is located just 10 minutes from the airport—a $5-per-person shuttle ride or a $7 cab ride. It is within easy walking distance of a wide range of quality restaurants, shopping, the harbor and Seaport Village.

CONFERENCE REGISTRATION

*** Registration notices will be sent out to all PWSA members in March ***

Registration forms will also be available on PWSA (USA)'s web site: www.pwsausa.org.

For updates on the conference, watch *The Gathered View,* the national association's Web site (above), and the California Foundation's Web site (www.pwcf.org).

1999 Awareness Week To Fund Conference Youth Program

Awareness Week Chair Jeannie Dickinson reports that chapters are being asked this year to raise money for conference support—40 percent will go to fund the conference youth program, and the remaining 60 percent can be used by chapters to help families attend the conference or for other local needs. Jeannie adds: "**Most of all, we are asking everyone to work as hard as you can to raise awareness.** Contact your local congressmen and women; ask for their support. Contact your local newspapers and television stations—send them a letter about your personal experience with PWS, as well as PWSA brochures. Ask local hospitals and doctors' offices to put up one of our new posters or to post our brochures. People are willing to help, especially because this is so rare and so devastating. The more awareness there is of PWS, the more people are diagnosed … and that means fewer people dying."

Conference Grants Available

A potential source of funding to attend the conference is your state's Developmental Disability (DD) Council. The grant may need to go through a sponsoring nonprofit organization, such as your local chapter of PWSA, The Arc, Parent-to-Parent, or other DD/MR-related organization. Each state will be different, but the money is there (our tax dollars!) for us to use. The purpose of your request should be expressed in terms of DD Council goals, such as "education and training," "family support," "community participation," or "self-advocacy and empowerment." *(Thanks to Hope Mays, GA, for this tip!)* To find your DD Council, call NICHCY at 1-800-695-0285.

PWSA (USA) helps some families each year who want to attend the national conference but who cannot afford the expenses or find other funding. Applications should be sent by letter to the Executive Director, PWSA (USA), 5700 Midnight Pass Rd., Suite 6, Sarasota, FL 34242, and must be received at the national office by **March 31** to be considered.

Grant application letters must include: 1) the size of your family and ages of your children; 2) an indication of your income and expenses; 3) a brief summary of difficulties your family is experiencing in dealing with PWS; 4) which conference or travel expenses would need to be covered by the grant and the estimated amount you would need; and 5) whether anyone in your family has attended a previous PWSA national conference.

A Life-Saving Experience

A Letter to PWSA from Rick and Connie Loshen, Danville, Illinois

We are already looking toward the 1999 PWSA National Conference in San Diego. For all of those who have never attended a conference, I want to share what our first conference in Columbus, Ohio, meant to us.

First, I would like to tell you a story, one that may be well known to many of you. It is about loneliness and isolation. Our daughter, Erin, is 12 years old and was diagnosed with PWS just a year ago. For 12 years, we obviously knew Erin was different. She loved to eat, was slightly overweight, and had many behavior problems. She underwent extensive testing (including genetic testing for PWS), but all tests were negative. She was classified as autistic, attention deficit, obsessive-compulsive, and just plain "different and complex." We had a child who didn't seem to fit in anywhere. We were stuck in a world that had no place for her or us. We had no idea what caused her problems and had no one to talk to. Our families didn't understand and distanced themselves from the discomfort of Erin's problems. It was a long and lonely 12 years.

When we were approached about retesting for PWS, we again dragged Erin in for yet another test, believing that it was probably a wasted trip. When the results came back positive, we were understandably devastated, but somewhat relieved. We finally had an answer. The diagnosis has brought great changes to our lives.

Luckily, we had a very concerned and caring doctor who immediately hooked us up with the support of the national and state PWS associations. We found out in March about the national conference to be held in July. Before we knew it, we were on our way to Columbus and an experience that "saved our lives."

In Columbus, we met and spoke with many professionals who worked with the Prader-Willi population. They were all extremely helpful and kind. For the absolute first time, someone understood. We told them about experiences that no one else had ever wanted to hear about. They gave us information, insight, and practical advice that have been invaluable to us.

But just as important, we met parents just like us. They were understanding of our problems and challenges with Erin and were more than willing to share their own stories. These parents looked at Erin, not with disdain and disgust, but with love and compassion. For the first time in 12 years, we found "family."

Perhaps one of the greatest benefits of the conference was watching Erin gain a feeling of belonging and friendship. She had a wonderful time and has not stopped talking about all the friends she made. She could be herself and be totally accepted by her peers. In Columbus, she finally entered a world where she was welcomed with smiles and open arms.

We want to sincerely thank all of those who participated in the 1998 national PWSA conference. We are looking forward to attending the 1999 conference in San Diego and again seeing the many friends we made in Columbus.

A Challenge to Chapters, Members

The Michigan Chapter recognizes the leadership PWSA (USA) has undertaken in increasing awareness and supporting families and individuals with PWS. In doing so, it has grown from the simple grass-roots support group that was founded in 1975 to a multinational organization with 30-plus state chapters. This growth has not always been easy and has its costs. PWSA (USA) is currently running a $30,000-a-year deficit. In order to make the organization solvent, the board of directors has approved a contract with Mark Eason, Vice President for Research and Development at Goodwill Industries. Mr. Eason has over 25 years' experience with the administration and development of non-profit and for-profit businesses and will help PWSA with strategic planning and development of alternative funding sources. With his guidance, our national association should be able to continue its current endeavors and also launch new programs and fund research to help all those with Prader-Willi syndrome and their families. Our chapter has sent $2,000 to PWSA to help cover the cost of Mr. Eason's contract ($5,000 total), and we issue a challenge to other state chapters and/or individuals to come to the aid of PWSA (USA).

I clearly remember the day when I received the diagnosis on our daughter and was very grateful to have a kind voice on the other end of the toll-free number. I wouldn't want others to have to go through this ordeal alone. Thank you for your consideration.

Jim Loker, M.D.
President, Michigan Chapter
Prader Willi Syndrome Association (USA)

Grant Needed for Crisis Intervention

Our national PWSA office receives crisis calls every day. Most are from families who are in serious need of help, whether for information, support, or advocacy. All need a caring and knowledgeable person on the other end of the phone—with plenty of time to explore solutions.

Executive Director Janalee Heinemann is seeking a grant that would fund a full-time crisis intervention counselor to handle these calls.

If anyone has a lead on a national grant that might cover salary costs for a crisis counselor, please send the grant application forms to Janalee, or call her at 1-800-926-4797.

Ask the Professionals

Q Does the new diet drug, Meridia (sibutramine), work for people with PWS?

A **Dr. Zipf** responds: "There have been no clinical trials with Meridia in PWS children or adults. We don't know." **Dr. Cassidy** and **Dr. Wharton** reported no experience with the drug, and **Dr. Driscoll** says his only attempt to use it with PWS patients was stopped after two months by the parent.

Dr. Lee cautions against making any broad recommendations on diet medications: "There are no data to support the use of ANY pharmacologic agents [drugs] for the control of weight in PWS. Most of the agents used for weight control have serious potential side-effects (remember fen-phen?) which have not been studied in PWS. Furthermore, the FDA has not approved specific labeling of any weight control agent for PWS adults or children; therefore, physicians and manufacturers are legally prohibited from recommending such therapy except on a case-by-case level."

Q Is there any experience with using the Dr. Atkins Diet, or similar low carbohydrate/high protein diets, for weight loss in individuals with PWS? If so, with what results?

A **Dr. Cassidy:** I know of no medical trials [of the Atkins diet].

Dr. Driscoll: Our feeling is that any diet works. What we do is try to find the one that our patients will comply with. This can vary. There is no one magical diet that works for everyone. Diligence is the key.

Dr. Lee: As cited in my chapter in Greenswag and Alexander (*Management of Prader-Willi* Syndrome, page 48), there is one scientifically sound published article using a protein-sparing fast for treatment of PWS-related obesity. This article, by Bistrian et al, was published in 1977. To my knowledge, there have not been any meaningful published scientific data since that time.

A true protein-sparing fast, which we used on occasion at Texas Children's Hospital, is associated with potentially severe metabolic complications and even death. Therefore, such a fast should always be started on an inpatient unit and carefully monitored for the duration of the fast.

The Atkins diet, the "sugar busters" diet, and many other "carbohydrate-limited, higher protein" diets are basically less severe versions of a protein-sparing fast. I have used my own modification of this approach quite successfully for treatment of childhood obesity, with or without PWS, but I certainly would not make any general recommendations based on anecdotal experience.

Dr. Wharton: The Dr. Atkins diet ... is a takeoff on a diet that was experimented with in the early 1980s at Massachusetts Institute of Technology. It was called the "protein-sparing modified fast." This diet calls for the dieter to eat only protein and—unlike Atkins'—*no carbohydrates.* When people do this, after about two to three days their body has no carbohydrate available as a fuel, so it breaks down fat and uses the byproduct called ketones as the body's principal fuel. This diet appears to help people not be hungry, which is clearly a great benefit. It could be that this "anorexic effect" is due to the fact that since the body doesn't take in any carbohydrates, the insulin in the person's bloodstream falls to very small amounts. It is quite possible that either lowering insulin or the ketones themselves, or both, decreases appetite.

I have tried this diet with quite a few individuals with Prader-Willi syndrome, and there are a few problems but considerable potential benefits. In terms of the benefits, I have had a couple of patients lose over 100 pounds while living at home. Additional benefits are that it helps rid the lungs of excess fluid, so anyone very overweight with heart or lung problems is often helped. Also, it can result in early rapid weight loss, which can be of benefit for people with medical conditions. Another benefit to someone supervising the diet is that the ketone bodies that the body uses are disposed of in the urine. A simple daily test will tell whether someone is sticking to the diet or has found extra food.

In terms of some of the difficulties, the first is that it's difficult to adhere to for a variety of reasons: it's boring, it has limited food choices, it's hard for kids who are in school or others who depend on cafeteria food. It's also expensive.

There are additional cautions: First, young children should not go on this diet.

The Experts Quoted Here:

Suzanne B. Cassidy, M.D.
Pediatrician and geneticist
Case Western Reserve University, Cleveland
PWSA Scientific Advisory Board, Chair

Daniel J. Driscoll, M.D., Ph.D.
Pediatrician and geneticist
University of Florida, Gainesville
PWSA Board of Directors

Phillip D. K. Lee, M.D.
Endocrinologist
Children's Hospital of Orange County, CA
PWSA Scientific Advisory Board

Robert Wharton, M.D.
Pediatrician
Spaulding Rehabilitation Institute, Boston
Massachusetts General Hospital

William B. Zipf, M.D.
Endocrinologist
Ohio State University, Columbus
PWSA Scientific Advisory Board

❖

Medical Advisory Board To Be Established

At the January meeting of the PWSA board of directors, the concept of a Medical Advisory Board was discussed and approved. While the current Scientific Advisory Board reviews and advises on research proposals, the new Medical Advisory Board would prepare treatment guidelines and respond to questions such as those posed here. Drs. Dan Driscoll and Jim Loker have been assigned responsibility for development of this new board.

❖

Please send your questions for "Ask the Professionals" to the PWSA (USA) office.

It should be for teens and up. (The exception is for a morbidly obese child, i.e., >200% Ideal Body Weight with a medical complication such as sleep apnea or diabetes.) While on the diet, the person needs to take specific supplements—calcium, potassium, and multivitamins. In addition, prior to starting, the person should have an electrocardiogram to check their heart. Finally, the person needs to be under a doctor's supervision, as there should be periodic blood tests and other evaluations.

So, in summary, the [protein-sparing modified fast] works well if followed, may have specific benefits of helping someone feel less hungry, and is generally safe. However, it needs to be used with caution under the supervision of a doctor.

Dr. Zipf: There are no clinical trials with the Atkins diet, but we *know* that a balanced low-calorie diet *does* work for PWS, very well. This has been proven over and over again. Children in general need a balance of nutrients; any restricted diet needs to be balanced. There is *no reason* to use a special low-calorie diet over a balanced diet in PWS, and it may impair growth and development of the child.

"Our youngest child, Victoria, was diagnosed at 11 days old with PWS. I discovered a "thunk" in her right hip three weeks after her nine-month appointment and took her to the pediatrician, who diagnosed her with a dislocated hip. We are now in the cast process, and she will follow the cast with a harness. If the odds are good that a normal baby can get a dislocated hip, then why aren't the children that are diagnosed with PWS and have hypotonia and loose joints screened more closely for hip dislocation through either ultrasound or X-ray? I feel very strongly that we would not have to be going through this whole casting process if my daughter had been more thoroughly monitored.

—Charlene Devine, New York

Q What do we know about hip dislocation in infants with PWS?

A **Dr. Cassidy:** Hypotonia can predispose to hip dislocation, although it isn't very common. There is a standard pediatric maneuver called an *Ortoloni* that identifies this, or X-rays should be done. Certainly, the level of suspicion for hip dislocation in PWS should be high. No incidence studies have been done, and I have not seen it in a young infant with PWS.

Dr. Driscoll: I have not seen it very often in PWS. All pediatricians should be checking *all* their patients for hip clicks/dislocation. It is a standard part of the exam beginning as a newborn. PWS may be more susceptible because of the hypotonia, but again, I have not seen it that often in PWS versus other infants.

Dr. Wharton: Recently one of my young patients underwent surgery for hip dysplasia (a poorly formed joint). I'm not sure it's more common in PWS, as this type of thing happens regularly in typical kids as well, but I'd be interested in hearing from readers about their experience. My recent patient is the only one from a fairly large clinic that has had this problem. Hip dysplasia can be seen with individuals who are late walkers, but that's not the reason.

Editor's Note: We'd like to hear from you if your child has had a problem with hip displasia or hip dislocation. It so happens that both of us editors had experience with this in our children, and we're wondering if a broader survey needs to be done. Please send your story to PWSA's national office.

—Linda Keder and Lota Mitchell

The Haberman Feeder: Successful for Many Infants with PWS

Carolyn and Dr. Jim Loker, Michigan Chapter presidents, have been talking with many parents and doctors about early infant feeding and found that a unique bottle and nipple system called the Haberman Feeder has been quite successful for a number of families. The Haberman was first designed for babies with cleft lip and palate, but it has proven effective for babies with low muscle tone as well. Jim recommends it to parents and doctors who call them about young ones with feeding issues.

Before even considering G-tube placement, families need to try this type of feeder, the Lokers advise.

Here's some information from the Medela Company on how the Haberman Feeder works:

- "In conventional bottles, much of the baby's effort is wasted on compression of air within the bottle and movement of feed within the system. However, the unique Haberman teat is separated from the bottle by a one-way valve. Once filled, milk is held in the teat and cannot flow back into the bottle. All your baby's sucking effort is directed onto the contents of the teat, so even the weakest suck gets results."
- "The Haberman Feeder has a slit-valve in the mouthpiece which only opens when your baby sucks. Your baby has complete control and is never overwhelmed. It also has the advantage of variable flow. Graduation from a large opening, down to a shut position, can be achieved to suit the individual needs of your baby. Whichever opening you select, it will always shut between sucks, similar to the breast while breast-feeding."
- "The Haberman Feeder may reduce air-swallowing. It has a special air inlet groove which allows air in at the same rate as feed is released from the system. There is no vacuum to work against your baby's sucking efforts, and air-swallowing is avoided."
- "If your baby needs some additional help, you can gently squeeze and release the reservoir of the teat. This will deliver a controlled amount of feed into your baby's mouth. The full teat has a natural resistance that prevents you from squeezing too hard."

The Haberman feeder is available with an 80-ml or 150-ml bottle. There is also a mini-Haberman for smaller babies or preemies. Contact the Medela Company at 1-800-435-8316 or 815-363-1166 for more information or to order the feeder.

Research

Towards a New Understanding of Prader-Willi Syndrome: Analysis and Treatment Recommendations

By Robert H. Wharton, M.D., Spaulding Rehabilitation Hospital and Massachusetts General Hospital, and Karen Levine, Ph.D., Spaulding Rehabilitation Hospital and Children's Hospital, Boston, Mass.

Introduction

Chromosomal variations challenge the imagination of the molecular biologists and behavioral geneticists who try to correlate genetic disorders and behavioral phenotypes (common characteristics). Their challenge is to discover the pathways that lead from gene to phenotype. These are the critical pathways that convert biology into behavior. In this paper we will present new ideas supporting a unified theory behind the classic behaviors of individuals with Prader-Willi syndrome. The central theme is that individuals with PWS lack the early intrinsic biological and reflexive mechanisms to support newborn survival. The absence of these factors can be most readily seen in the newborn with PWS and serves as the basis for some of the critical behavioral features that are responsible for many of the difficulties encountered in later life.

Appetite disturbance has been highlighted as the cardinal feature in individuals with Prader-Willi syndrome. However, while excessive appetite is clearly the most dramatic neurobehavioral feature associated with PWS, it is neither the initial nor is it always the most important neurobehavioral challenge. In fact, an intense and narrow focus on excessive appetite diverts attention and investigation from other neurobehavioral issues that could better provide a key to understanding this most complex neurobehavioral disorder.

We suggest that appetite is only one of a cluster of neurobehavioral features that stem from a common unifying system: namely, the body's system for self-protection and survival. Within this system are the following additional features that are also disordered in individuals with PWS: arousal or sleepiness, attention, pain response, and social attachment. Further, while we do not discount the critical role of the appetite disorder, we will present the difficulties around the issue of social attachment as an important cause for much of the challenge of the syndrome for both individuals with PWS and their families.

Considering the impact of PWS on individuals affected by the disorder from a single hypothesis has several distinct advantages. First, providing a unified construct facilitates a better understanding for families and providers. Second, researchers can target their investigations on the basis of more specific hypotheses. And, third, specific areas for medical and behavioral treatment that enhance quality of life for individuals with PWS and their families can be more directly targeted. In this paper we will present some new ideas regarding some of the behaviors of individuals with PWS. We will then provide a brief introduction of a therapy that we believe can assist parents and providers in enhancing the interactional successes for children with PWS and therefore improve their quality of life.

Neurobehavioral Features

Appetite

"The human body," according to Jean Anthelme Brillat-Savarin, "that highly complicated machine, would soon be useless if Providence had not placed in it a sentinel which sounds a warning the moment its resources are no longer in perfect balance with its needs. This guardian is appetite, by which is meant the first warning of the need to eat."

It is important to consider an individual's opportunities for eating in order to be able to understand the role of hunger. First, humans benefit by feeling hunger when food is either present or potentially available. The most critical time to be hungry is directly after birth. However, although hunger at this time is certainly necessary for survival, it will not be sufficient. To successfully feed and be nourished, the infant must simultaneously accomplish several complex motor and behavioral tasks. Specifically, the newborn, with assistance from its mother who has her own programmed nurturing instincts, must both locate the mother's food source and be located by the mother, latch onto the food source, coordinate a suck and swallow to utilize the source, and have the ability to repeat this activity on a regular basis.

However, with respect to this system, newborn infants with PWS lack the required skills needed to promote survival. They fail to root to locate the food source, do not cry to alert the mother to their presence and need, nor do they cry in hungry protest if feeding is prematurely withdrawn. In addition, they sleep excessively in the newborn period, thereby diminishing their availability to feed, and in fact are likely to sleep through the night within the first week after delivery, thus further failing to demonstrate hunger.

The first indication of abnormal hunger in children with PWS occurs at around 18 months to 2 years of age of age. Subtle changes are veiled in what appears to be normal hunger. Children at this time become, as one father described his child, "serious eaters." By this, the father meant that his child had started to eat vegetables! In fact, the child not only ate vegetables but would eat all foods offered.

The next indication of a disordered appetite is when the young child with PWS finishes everything on his or her plate. However, what makes this appetite behavior abnormal is the comparison with the eating patterns of typical young children. Typical toddlers neither eat a broad range of foods nor demonstrate a consistent eating pattern. Rather, their eating pattern reflects the fact the food is frequently a secondary rather than a primary concern for them.

As much of the literature on PWS

suggests, children with PWS typically demonstrate traditional signs of an increased or disordered appetite between the ages of 3 and 5. For the individual, however, although the behavior itself may be considered abnormal, the behavior serves an important biological goal. This enhanced appetite ensures that individuals with PWS will accumulate food sufficient to survive prolonged periods of starvation. Further, even should their ability to find food yield inadequate supplies, their "energy efficiency," that is, their propensity to store food as fat rather than to directly utilize foods they have ingested, also serves to stave off starvation. Hence, individuals with PWS appear to be uniquely capable through appetite, behavior, and metabolism to tolerate starvation.

This transition of appetite over time, from anorexia (lack of hunger) to insatiability (constant hunger), is one of the more intriguing aspects of the neurobiology of the disorder. Moreover, it is a unique aspect of the neurobiology of PWS: No other neurobehavioral feature goes through this type of evolutionary change.

No amount of sleep alters the need for sleep, just as no amount of eating alters the need for food.

Arousal

According to the poet John Keats, sleep is meant to "enliven all the cheerful eyes/That glance so brightly at the new sunrise." Neuroscientists, only somewhat less romantically, see sleep as a suspension of consciousness, a respite for the mind, whereby the motionless human lies is a state of neuronal disconnection and deactivation. Wakefulness, a neurological state competing with sleep for the domain of activity, lies at the other end of consciousness from sleep. While most individuals alternate their time between these two complementary states, some spend their waking hours in a persistent state of sleepiness.

Following delivery, the infant with PWS fails to demonstrate the level of arousal seen in typical newborns. Whereas newborns, in general, demonstrate heightened arousal and attention that enable them to respond preferentially to a mother's voice, imitate facial expressions, and explore facial features, the newborn infant with PWS fails to demonstrate these essential behavioral features. Once again, newborns without these abilities will be less able to survive, as they will be unlikely to attend to their need to locate their mother or persist in their efforts to be supported by the mother.

Diminished arousal seen in the neonate confirms its presence in PWS as it continues to some degree throughout the individual's life. This diminished arousal is most easily recognized by observing the several features consistent with sleepiness. First, individuals with PWS will generally sleep when there is minimal stimulation. This is manifest in car rides, sitting watching TV, waiting in doctor's offices, or in other settings of boredom. Further, their entry into sleep is characterized by a dramatic rapid descent from an awake state into a sleep state, similar to a sleep attack often seen with narcolepsy. This sleep behavior, rather than demonstrating continuous sleepiness, instead suggests an altered baseline level of arousal.

The disordered sleep and appetite can be conceptualized as an alteration in biologically determined "set points." A lowered set point for arousal fits with a raised set point for appetite. Whereas individuals with PWS cannot achieve satiety with respect to appetite, neither can they achieve satiety for sleep. No amount of sleep alters the need for sleep, just as no amount of eating alters the need for food. Therefore, what appears to be sleepiness is rather diminished arousal.

Attention

Diminished arousal also impacts attention, another important survival mechanism. Children with PWS are typically not hyperactive and therefore the quality of their attention is seldom called into question. On the contrary, rather than hyperactive they present as quite hypoactive (underactive) and, rather than showing a poor attention, they demonstrate what could be described as a strong attention. Young children can sit with and apparently enjoy the same book, toy, or video for a prolonged period of time. Older children and adults can work with word puzzles for hours. Therefore, the likelihood of an attention deficit can seem remote, as their persistence in specific tasks appears to reflect a strong attention.

They are not merely attending well to their particular interest, they are unable to transition out of this interest.

However, what one often considers to be "good" attention in these situations is instead a manifestation of a somewhat abnormal or "sticky" attention. This apparently strong attention may more accurately be considered an obsessive or perseverating behavior. It is not merely that the individual is strongly focused on, or engaged in their book or puzzle. Instead, they are "locked into" a particular satisfying activity which is rewarding to them. Moreover, the reward is not the pursuit of novelty present in typical infants and toddlers, nor the educational satisfaction they achieve from their work; rather it is the repetitive and sustained nature of the interaction, the sameness of the work they are doing. They are not merely attending well to their particular interest, they are unable to transition out of this interest.

Another likely reflection of their attention is apparent in their memory profile. Individuals with PWS are known to have an excellent long-term memory. Once information is learned, it is carefully and successfully stored to be made accessible for the future. However, another aspect of memory, namely auditory short-term memory (the ability to remember what was just heard), is clearly deficient. Neuropsychological testing consistently demonstrates this to be an area of weakness for individuals with PWS. Further, this specific difficulty is not associated with the general degree of cognitive strengths or weaknesses. While auditory short-term memory can reflect difficulties in a variety of areas, it can specifically reflect diminished attention. Individuals

(Continued on page 10)

Attention—*continued*

who are not able to attend to presented material because of their own distractibility, inability to focus, sleepiness, or other characteristic will not be able to able to store that information. If, however, they are able to store the information, that storage is generally successful. Therefore, with respect to attention, individuals with PWS have a "sticky attention" that interferes with cognitive flexibility and a shortened attention that precludes the easy incorporation of information.

Pain

An analysis of the system of pain perception and response demonstrates behavioral characteristics parallel to those of the other survival systems. Parents of individuals with PWS consistently report that their children have a high pain threshold. They relate anecdotes of their children not reacting to blood tests and immunizations that typically provoke pain responses in their other children. Pain, like hunger, is a feeling that is meant to generate a response in the individual that will ensure the body's physiological integrity or survival. Whereas hunger is a sensation designed to tell an individual that energy supplies critical to maintaining the body's working ability need to be restored, the sensation of pain warns the individual that the body's general integrity is in some way threatened.

However, there are also times when feeling pain would be a disadvantage. Consider the classically described situation of a mother suddenly finding her young child's body trapped under a piano. The mother, believing only that her child will die if she is unable to free him, spontaneously lifts the piano enough to allow her child to escape. If the same mother had wanted to lift the piano for a more neutral purpose, like freeing her sheet music, she would not succeed. What enables her to lift the piano off her child is not the immaculate presence of new-found strength but rather the lack of the sensation of the pain involved in the experience. The stress she experiences seeing her child trapped causes the release of chemical mediators—endorphins—that temporarily mask the sensation of pain. Following the removal of the piano, however, the body will recognize the importance of attending to the muscles that have been torn through her effort and will release an additional hormone, cholecystokinen (CCK), to alert her to the need to repair her injuries.

This example illustrates that pain, like hunger, arousal, and the other neurobehavioral features that have been described, is another sensation that promotes a person's survival. The key for the organism is to survive. When pain is the chief signal of a threat to the body's integrity, the person will feel the pain and have an option to respond. However, should a person be in a situation where the body does not sense anything they perceive as a threat, there will not be a response.

Pain, although likely felt, does not call for a response because the pain itself does not produce in the individual the critical sense that their body is threatened and needs protection.

The initial results from our survey on pain appear to confirm parents' impressions of their children's high pain threshold. Specifically, parents consistently report that their children have not reacted to some of childhood's typically painful experiences, such as teething, colic, immunizations, or blood tests. Where there is a response to an injury, this is more likely to be a response to the surprise or other emotional discomfort, such as that rendered by a fall or involved in the trauma. Pain itself, however, does not elicit a call for maternal assistance, a call for protection. Pain, although likely felt, does not call for a response because the pain itself does not produce in the individual the critical sense that their body is threatened and needs protection.

Social Attachment

Arousal, pain, appetite, and attention are all systems negatively affected in individuals with PWS. Furthermore, each of these behaviors is not bound by affect or emotions. They are instead systems that the individual instinctively employs in routine and stressful situations to ensure protection and survival. An additional behavior, social attachment, is less obviously but, nevertheless, significantly affected by the neurobiology responsible for the Prader-Willi syndrome phenotype. While it is critical to understand that individuals with PWS clearly are attached to their parents and others and form close and meaningful bonds, nevertheless, there are some disturbances with respect to their attachments that are key to the understanding of the disorder.

The writer Elie Wiesel expressed social attachment in a straightforward and simple manner: "Smile all you want, Dr. Freud," he wrote, "but I was attached to my mother, maybe too attached ... I search my mind for my earliest memory, and I see a little boy sitting on his bed, calling for his mother." The psychologist John Bowlby, more scientifically, maintained that attachment was both a collection of observable *instinctual* behaviors, such as holding, vocalizing and greeting, that infants direct to their caregivers; and a corresponding *affective* state that the infant expresses through these behaviors. The behaviors enhance an infant's ability to remain physically close to the target of attachment, especially during times of distress, or emotional stress; while the psychological state permits the infant to feel safe and secure when in the presence of the familiar.

At birth, and in the immediate postnatal period, instinct for protection and survival is the predominant force responsible for an infant's behaviors. Infants demonstrate their sophisticated survival skills by their visual, auditory, and olfactory responses—even newborns can demonstrate preference for mother's face, voice, and milk.

However, these early instinctual skills, similar to other reflexes available to all newborns, serve a transitional function, protecting the child while the infant's brain matures and develops a process for learning that is able to provide the predominant life-sustaining and life-enhancing behaviors. What is key about the transition from instinctual to cognitively driven affective behaviors is the gradually developing shift from the in-

... individuals with PWS clearly are attached to their parents and others and form close and meaningful bonds, nevertheless, there are some disturbances with respect to their attachments that are key to the understanding of the disorder.

stinctual drive for the physical closeness of infancy and the perinatal period to the more affectively based need for emotional closeness in later infancy and childhood.

At delivery, the infant, having just endured a stressful birth experience, needs to attach and bond to its mother to ensure survival. Behaviors necessary to promote survival are heightened sensory awareness, ability to feel and react to pain, hunger, alertness, and sustained attention. In addition, and of considerable importance, the initiation and development of a social attachment to the mother will enhance the child's potential for survival.

The typical infant's attachment to its mother continues to grow during the first few months in what is typically called the maternal-infant bond. At around six months, infants begin to demonstrate that losing sight of their mother is a cause for alarm. As described by Bowlby, when a child at this age loses visual contact with his mother, the initial response is one of protest and urgent attempts to recover his lost mother. According to Bowlby, "he will often cry loudly, shake his cot, and look eagerly towards any sight or sound which might prove to be his missing mother." Not only has the infant's memory expanded to permit recognition of what he is missing, but his social attachment has reached a peak whereby he recognizes that his needs are completely tied into his mother's. This discovery becomes the cardinal manifestation of childhood attachment or parent-infant bonding.

The newborn with PWS typically lacks these essential instinctual, and therefore inborn, reflexive survival skills. During infancy, their lack of strong search for social attachment continues. In a recent survey, the vast majority of parents of children with PWS indicated that their infants and children did not demonstrate any specific response to separation. Parents were asked the question whether their children demonstrated concern when the mother left their sight. These children, for the most part, did not cry, scream, yell, or make any dramatic protest when their mothers left the room they were in and withdrew from their immediate worlds. The child with PWS, though able to connect to his mother and feel a sense of the mother's importance, nevertheless, cannot see her critical need to nourish and protect. Therefore, her departure may cause some emotion related to the loss of an important person, but not to the threshold that suggests to the child a sense that his integrity or survival is somewhat threatened.

This somewhat restricted social interaction carries over to the child's involvement with friends as well as family. Although the child with PWS may enjoy playing with other children and adults their play is marked by certain characteristics: first, they infrequently initiate these interactions, opting instead to be the recipient of the attention of others; second, when they are playing and the play stops, they generally do not have the social skills or desire to maintain the play situation; and, while they have friends, their attachments to their friends frequently falls somewhat short of having a best friend, someone they depend on for physical play or emotional support.

Young children with PWS are often content to sit and play alone. Parents, teachers, and others may interpret this behavior as a lack of social interest, or perhaps a preference for solitary play, and be hesitant to intercede. We believe, on the contrary, that aggressive (but fun!) "interference" will help the child develop successful social skills.

To better assess the issues around social attachment, we are now conducting a study using what is known as a "Strange Situation." This psychological paradigm employs the observation of a child with his mother and also with a "stranger" to assess the strengths of a child's attachment to his mother. We anticipate that what we learn from doing this study will not only help our understanding of PWS, but will also help in the development of strategies to enhance the social attachments and interactions of individuals with PWS.

Treatment

We have been recommending and using a particular treatment with the families of some of the young children with PWS in our program. This intervention is an adapted form of "Floor Time," a play-based intervention developed by Stanley Greenspan, M.D., child psychiatrist, and described in some detail in his most recent book, *The Child with Special Needs* (1998). The principle of Floor Time is that increasing a child's arousal increases social interaction, communication, and learning capacity.

Young children with PWS are often content to sit and play alone. Parents, teachers, and others may interpret this behavior as a lack of social interest, or perhaps a preference for solitary play, and be hesitant to intercede. We believe, on the contrary, that aggressive (but fun!) "interference" will help the child develop successful social skills. One family described that until they began Floor Time their family would all be piled up roughhousing, and their child with PWS would be playing off to one side. They had previously interpreted this to mean the child did not want to play with them. Once they began actively working to engage the child, however, he began to initiate play with his siblings.

The first step in a Floor Time approach is to get the child excited. With very young children this is often most effectively done with happy physical play such as tickling, or roughhousing. Once the child is engaged and excited, frequent small "pauses" in the play allow the child to signal that he wants "more." For very young children this requesting more may

(Continued on page 12)

Happy Birthday, David!

Vicky Knopf of Salem, Connecticut, shared her son David's story and photos with Gathered View readers nearly two years ago (April 1997). As he passed his fourth birthday, she sent PWSA this update on life with David.

Well, it's that time again. Four years ago today (December 1), we were dumped in Holland with no explanation whatsoever [a reference to Emily Perl Kingsley's essay, "Welcome to Holland," *The Gathered View*, February 1997].

David's birth and the events following are etched in my mind forever. I see the faces of the doctors telling us that they don't know what's wrong with our baby, I hear the constant hum of the NICU, and I can still smell the smells. And lastly, I can still remember the diagnosis ringing in my ears: "Prader-Willi syndrome." What on earth is that?? "Expect your child to be mentally retarded, obese, and have horrible behavior problems."

Well, we have come a long way since that day four years ago. We have survived 11 hospitalizations, six surgeries, endless amounts of therapy. David has blossomed into a beautiful little boy who has met and exceeded all our expectations. He has a wicked sense of humor and unconditionally loves everyone. A delivery man who came to the door got a very unexpected hug yesterday … he said it made his day! David has a knack for knowing when someone needs a hug … whether he knows them or not.

I still feel the twinge of pain when I allow myself to think about the "what ifs?" but that happens so seldom now. I think more of "what if he hadn't come into our lives?" For one thing, I would never have met all you wonderful people who have become more of a family to me than I ever thought possible. I rejoice in all your triumphs, I grieve with your losses. My other four children are much more aware that people are only different on the outside, that it's what inside that counts.

Our lives are so much richer because of David, and David wouldn't be David if he didn't have PWS … it's part of who he is. He is one special, terrific little guy! HAPPY BIRTHDAY, DAVID … I LOVE YOU! LOVE, MOMMY

—Vicky Knopf

David Knopf at age 4.

"Trouble Beads" can help stop some troubles with skin-picking, according to the case worker of a young man with PWS. In a conversation with Janalee, this provider explained that the young man keeps a small set of chosen beads on a heavy duty chain in his pocket. When he's tempted to pick at his skin, he rolls the beads instead. It works for him!

Treatment—*continued from page 11*

come through subtle responses such as a glance or a giggle, or signaling by moving his arms or legs. The adult continues this interaction by responding happily and expressively to any social initiation attempt, then pausing again. By playing "slowly" enough and reading the child's cues carefully in order to respond to them, the adult will help young children with PWS respond very happily to this type play.

In preschool, children with PWS are often socially isolated due to diminished social initiations. For these children, having playtimes begin with "silly" games that increase arousal can maximize their capacity to initiate and sustain social play. Teachers can readily facilitate social play by increasing the children's arousal through excited fun social play. In larger preschools, an individual aide to assist with social facilitation using Floor Time techniques is very beneficial.

Summary

This manuscript presents a new profile of individuals with PWS. Rather than highlighting excessive eating characteristics and the medical consequences of morbid obesity, the manuscript presents observations suggesting that behavioral characteristics responsible for the protection and survival of the individual are at the core of the disorder. These characteristics consist of diminished arousal, decreased pain perception, excessive sleepiness, and decreased attention. Finally, social attachment, one of the key behaviors associated with an infant's survival and a child's quality of life, is presented as a new significantly affected behavior in individuals with PWS.

We hope that with this new understanding will emerge new strategies for exploring some of the biology responsible for the difficulties involved in the disorder. We also anticipate that the play-based strategy that we present will enable significant numbers of young children to enhance their social and emotional well-being and therefore their successes in life.

▲▲▲

Editor's Note: This article is adapted from a longer piece published last year in *Prader-Willi Perspectives*. Drs. Wharton and Levine also presented their observations, including videotape of the "Strange Situation" research, at the PWSA (USA) National Conference in Columbus, Ohio.

Parents vs. Providers: A Losing Battle

By Barbara J. Goff

Last year at the annual conference in Ohio, I had the opportunity to participate in a panel discussion regarding residential options for adults with PWS. The panel consisted of parents and providers, with me straddling the middle. I used to be a provider of residential, respite, and crisis services for kids and adults with PWS, but for the past four years or so, I have been working independently on behalf of both parents and providers: consulting, training, working with schools, evaluating services, providing advocacy, etc.

I have always been a big fan of families and have prided myself on working hard to establish positive relationships with the families of the individuals I have served over the years. Having been a provider for 20+ years, I also have a deep appreciation for the struggle of trying to create positive, warm living environments while meeting endless rules and regulations, maintaining high quality staff, and facilitating relationships among groups of individuals who were strangers one day and roommates the next.

While rifts between parents and providers are to be expected to some degree, I believe there is special cause for concern when it comes to services for individuals with PWS. I don't have figures, but I know that some providers have voluntarily stopped serving individuals with PWS as a direct result of "parental dissatisfaction and interference," and I know some parents whose children are in residential situations that could stand some major improvement, but the parents don't want to rock the boat.

Some of this conflict and tension came out at that workshop last year, and it keeps nagging at me. So, in an effort to promote some healing dialogue, I am asking that you—parents and providers—fill out the attached survey, the results of which I hope to present at the conference in San Diego and in a future article for *The Gathered View*.

If you are the parent of an adult living in a residential program or a provider of such, please take a few moments to say what's on your mind. Let it all out! Your answers are completely confidential and will never be reported in a way that would identify you or your situation.

Perhaps when we come together in San Diego we can tackle some of the biggest issues, whether it's socks that don't match or the parents who won't let their kid learn to clean his own room. I NEED LOTS OF RESPONSES TO MAKE THIS INTERESTING AND WORTHWHILE. … I thank you in advance.

PARENT AND RESIDENTIAL PROVIDER SURVEY

Deadline: As soon as possible (But no later than May 15).

Return this survey to B.J. Goff, 33 Benz St, Springfield, MA 01118
or e-mail your answers to galagof@compuserve.com

Please feel free to attach additional sheets and add any comments you think would be helpful in addressing the issue of parent/provider relationships.

Parent ___ Provider ___ *(check one)*

Please identify your state: _____ *(optional)*

Parents Only:

My child resides in *(check the one that best applies)*:

___ Intermediate Care Facility (ICF)
___ Community residence/Group home
___ Supervised apartment (24-hour supervision)
___ Shared living (shares home with nondisabled person)
___ Semi-independent (less than 24-hour supervision)
___ Other *(describe briefly)*: _____
_____

There are a total of _____ *(number)* individuals served in this residence.
Of these, _____ *(number)* have PWS.

My child has been living at this residence for _____ years/months *(circle one)*
He/she has a single room: Yes ___ No ___
If No, with how many others is the room shared? _____.

Providers Only:

We have been serving individuals with PWS for _____ years/months *(circle one)*.

We currently do so in the following residential settings *(check all that apply)*:

Mixed	*PWS only*	
___	___	Intermediate Care Facility (ICF)
___	___	Community residence/Group home
___	___	Supervised apartment (24-hour supervision)
___	___	Shared living (shares home with nondisabled person)
___	___	Semi-independent (less than 24-hour supervision)
___	___	Other *(describe briefly)*: _____

_____

Both Parents and Providers: *(If parent, address issues with provider and vice versa)*

My biggest concerns about the provider/parents are:

1.

2.

3.

(continued on next page)

Parent/Provider Survey—*continued*

Three things I really like about the provider/parents are:

1.

2.

3.

Parents:
If I had an alternative, I'd remove my child from this residence: Yes ___ No ___

Providers:
My job would be great if only the parents would stop/start (whichever):

Both Parents and Providers:
Why do you think your relationship with the parent/provider is so good/bad?

What do you recommend to change/improve/enhance your relationship?
(provide solutions or alternatives to your greatest concerns, and add any other recommendations)

If you are willing to be contacted regarding this issue, please include your name and phone number: *(optional)*

Name _____ Phone _____

THANKS AND SEE YOU IN SAN DIEGO!

WANTED:

E-Mail Pen Pals

Hi! My name is Brittany, and I would like e-mail pen-pals. I'm interested in someone who is around my same age (16) and who likes to do puzzles in books. I live way out in the West Texas desert, but I like it here. I like the quietness and the coyotes howling at night except when they get my dog agitated. I want to be a veterinarian when I grow up. I also like to do cross-stitch on plastic canvas a whole lot, read and color. My e-mail address: avalonb@nwol.net

Family of Same Faith

The mother of an 8-year-old girl with PWS would like to know if there is another PWS family out there who are **Christians of the Independent Missionary Baptist** faith. If so, she would very much like to hear from you. Contact: Cathy Pendleton, 2551 Essman Sugar Camp Rd., South Webster, OH 45682, Tel.: 740-778-2933.

Extra-Wide Shoes

A mother is looking for a source of extra-wide shoes for her child. If anyone knows of a particular shoe manufacturer or distributor to recommend, please call the national PWSA office (1-800-926-4797).

Palm Prints for Study

Miriam Livny, a sibling and student in New Jersey, is looking for participants for a research project on palm prints. She writes: "I collected fingerprints and palm prints from people with PWS and compared them with the common abnormal print patterns [such as those of other syndromes]. I concluded that the majority of the palm prints were abnormal, of a type similar to the one that characterizes Down syndrome, yet containing an extra line … I would like to continue my research, in order to substantiate my results.

"If anyone with PWS would be willing to send me his or her palm prints, it would be greatly appreciated. If you have a stamp pad at home, simply coat your entire palm with the ink (preferably black or blue and not too much). Then, press your hand onto a white sheet of paper, being careful not to move it while on the paper. Hold your hand on the paper a few seconds, and then lift. Do the same for each hand. Make sure the print is clear and not smudged. On this paper, be sure to include your name, age, gender, and birthday. Please also indicate whether you have a chromosome deletion or disomy (if you know). *Thank you!*"

Send your palm prints to: Miriam Livny, 42 Thomas Dr., Manalapan, NJ 07726.

More 1998-99 Angel Donors

CONTRIBUTIONS RECEIVED AS OF DECEMBER 31, 1998

Gabriel
($1,000 or more)

James E & Suzanne L. Fuller
Jim and Joan Gardner
James and Mary Kane
Robert & Wauneta Lehman
Esther Plank
John Seibel
Tom and Michelle Torbert

Heavenly Angel
($500–$999)

David K. Kaugher
Donald Minsk
Versa Handling Company
Francis H. Williams

Arch Angel
($250–$499)

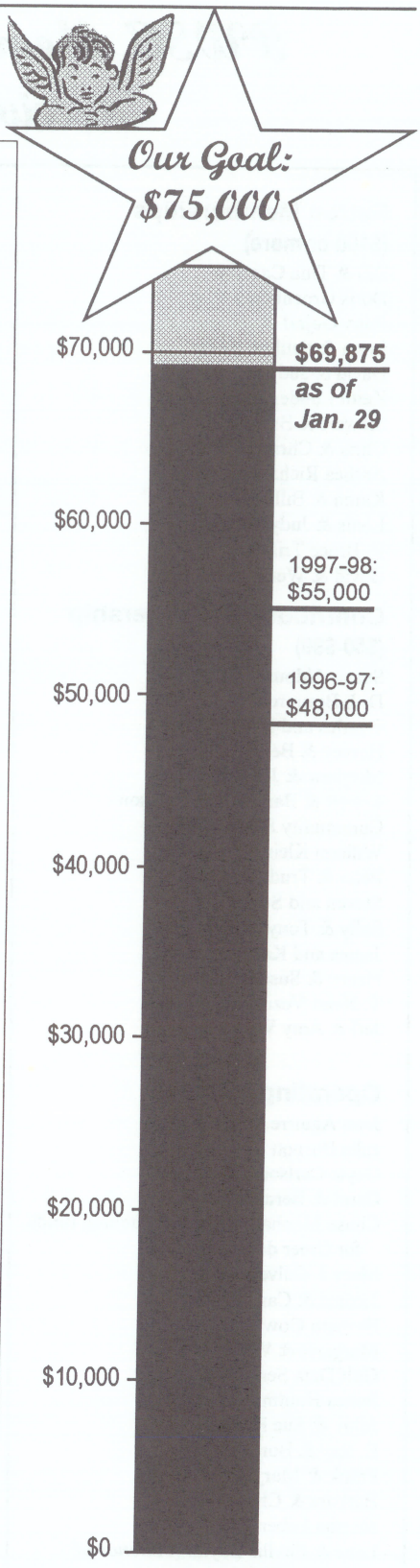
Delfin J. Beltran, M.D.
Fredna & Marler Bennett
David & Michelle Campbell
Craftsmen Interiors, Inc
Lota Echols
Phil & Becky Gulling
Richard & Darlene Heinricy
Mr. and Mrs. Lionel Lamoureux
Cyr & Anne Linonis
James & Carolyn Loker
Betsy McCool
Robert Muirhead
Jacques & Pauline Parent
Jack Roe
William C. Stege
Sandra Stone
Earle (Roo) & Susie Wood, III
Soo J. Yim

Angel
($100–$249)

Mary Baldwin
Roger & Beth Barnett
Larry & Diane Baylor *(in honor of Bob, Wauneta, & Barbara Lehman)*
Byron & Gretchen Braun
Gregory and Elizabeth Britton
Nicholas Brown
Julie Burnett
Patrick Casey
Christian Perspective, Inc.
John Cooper
James Davis
Ron & Randi DeHaan
Gladys & William Faherty
Jerry & Suzanne Fawbush and the Fawbush Family
First Congregational Church
John A. Forster, Jr.
John & Barbara Gillen
Jack & Karyn Goldberger
Jack Green
Paul & Roda Guenther
Robert D. Harrison
Thomas Hauser
Ralph Heller
Willie Hinson
Mary Lou James
Al and Linda Kennett
Richard K. Lamberson
Kristin Lay
Monroe & Constance Levy
Fred & Pat Lipp
C. W. Lonnquist
Patricia Lueders
Ralph Magalee
Diane Masterson
Patrick and Erin McCarthy
Angus & Nancy McDonald
Steven & Joanne McMaster
John and Karen Meslow
Eugene & Jean Alice Metzger
Lyle Millan
Terry & Debbie Mleczewski
Fran & Frank Moss
Regina R. Murphy
Joseph H. Nook, III
Arvind F. Patel
Family of William Richardson
Vaunne B. Schell
Kathleen Schloessinger
Janet M. Schuman
Nell Shapiro
David & Jane Shoemaker
Eugene Smith
Kelly & Amy Soncarty
Early & Jackie Spiars
Finis & Loretta Stafford
Cliff & Wanda Strassenburg
Chris Sweeney and Ruthie Duescher
Richard & Eleanor Weiner
Karl & Denise Westenfield
Richard & Marge Wett
Elwood & Mary Jane Wissmann

Cherub
(Up to $99)

Charles & Gladys Allred
Max & Rosalyn Alterman
Dolores Bachen
Josette Barrett
A. L. Bayless
Kate Beaver
Laverne Becker
William and Susan Beecheler
Daniel & Linda Bennett
John Bintz
Mary B. Boyum
Timothy & Penny Brandon
Darlene & Dwain Buer
D.P. Burleigh
Richard & Susan Butts
Brenda Calabrese
Mary Carlson
Kathleen & Louis Cillo
Ray & Mary Clawson
Chris & Linda Cooper
Alan & Diane Crone
Salvatore & Lillian DelSesto
Lorraine Diamond
Ray & Elsie Dorn
Byron & Sharon Eager
Debbie & Michael Fabio
Ed & Mary Farley
Jason & Nancy Finegold
Levy Gales
Del & Mary Lou Gerdes
Roger & Tracy Goatcher
Marion Goodman
Jim & Joni Gorman
Steve & Loni Hamilton
Cornelius Hanley
Jody Hanus
John R. Harrington
Marie Harvey
Greg & Cindy Heinemann
Janice Henry
Ronald & Ethel Hirshon
Volena A. Howe
Jean T. Janes
Charles & Barbara Johnson
Tom & Bonnie Jones
James C. Karras, Sr.
John Kline
Ken & Maria Knox
Evelyn Krebsbach
Leona Lacroix
Jean R Larosa
James Lazarus
George and Ruth Lockwood
Andres Lopez
Lane and Phyllis Loyko *(in honor of Amanda Diaz)*
Wanda Manchel
The Marciniak Family
Ernest & Alice Marek
Phyllis Martin
Jacque McGuire
LaVerne Mikeska *(in honor of Chelsea Ocker)*
Kathryn & Glenn Moffett
Len & Karen Moore
Michael Mosteller
Lorraine Okuda
Lisa Oppenheim
Allan & Catherine Partridge
Richard Ruzicka, Jr
Dennis & Sharon Saacks
Cynthia Sajid
Timothy & Cindy Schmehl
Mary Schneider and John Beck
Donna J. Schramm
Jeff & Angela Schroeder
Hazel Schuster
Jack Sherman
George & Margaret Smith *(in honor of Lota & David Mitchell)*
Richard & Darlene Smith
Robert Snow
Cis Speer
Edward & Ruth St. John
Edward J & Eleanor Stepanik
Jan Sundell
Ewina B. Thompson
Stanley & Tracy Thorp
Anna Tomasicchio
Andrew Trentacosta
Christina & Michael Turcola
Virginia G. Turner
Mary H. Valliere
Ray & Susan Wagner
Ernest & Gloria Walters
Patricia Wasmund
Shirley Weger
Barry & Lisa Whoric
James A. Wilmott
Harry & Dortha Wilt
Ray Zimmerman



Our goal is in reach, thanks to PWSA's Angels!

PWSA Membership Gifts and Other Contributions Received Through December 31, 1998

Patron Memberships ($100 or more)

Bill & Tina Capraro
Doris Groenboom
Rajiv Gujral
Erich & Pauline Haller
David & Judy Lehman
Keith Louden
Stewart & Bronnie Maurer
Chris & Christy O'Gara
Anthea Richardson
Karen & Bill Ripley
Louis & Judy Schutz
R. Bruce Trimble
David & Wendy Young

Contributing Membership ($50-$99)

Susan Aldous
D. J. Braunreiter
Charles and Margaret Bush
Harvey & Beverly Bush
Matthew & Jennifer Chase
Robert & Barbara Christenson
Community Systems, Inc.
William Klein, Sr.
Peter & Trudie Lehman
Steven and Susan Lundh
Sally & Tony Mancheno
James and Kathy Remski
Henry & Susanne Singer
T. Nejat Veziroglu
Jeff & Amy Willinsky

Operating Fund

Joan Aguirre
Julie Burnett
Gayle Carlson
Carol & Bernard Charles
Chase Manhattan Bank (matching funds for Greer donation)
Mary J. Culver
Jerome & Carol Glorkler
Barbara Gow
Margaret & William Greer
Gulf Data Services
James Houtman
John & Sue Hruska
E. Ray & Bonnie Johnson
Frank & Mary Keenan
Barbara & Charlie King
Martha Lebenstein
Lane & Phyllis Loyko *(in honor of Amanda Diaz)*

Operating Fund—*continued*

Ruth Masters
Bernice & James McKean
The Mentholatum Company, Inc.
John Miller IV
Dorothea Mokrejs
Christy Peak
Skip & Nancy Potter
Louise Robinson *(in honor of Tracy Nichols)*
Laverne Sidlo
St. Paul's Lutheran Church *(in honor of Rachel Forster)*
Phillip Stehling
United Way of Champaign County
United Way of Northeast Louisiana
United Way of Orange County
United Way of Southeastern Pennsylvania

Research Fund

Stuart R. Boyd
David Byers
Shirley R. Schwartz

Chapter Donations

PWSA of Maryland and Metropolitan Washington DC
PWSA of Missouri
PWSA of New Jersey
PWSA Northwest Association
PWSA of Ohio
PWSA of Wisconsin, Inc

Restricted Donations

Chippie Alterman Trust
Paul Alterman
R.B. Galloway
Jim Gardner

Gifts In-Kind

Don & Peg Goranson—reproduction of 300 PWSA videos
Russell & Daphne Mosley—production of 2,000 restaurant wallet cards
Joyce Welch—used Mac computer system with printer

REMEMBRANCES

DONATIONS IN MEMORY OF

ROSEMARY BELL
Dick & Judy Drewry
Patricia Kerr
William & Marjorie Shoopman

JASON BOND
(and in honor of Jeff Bond)
Robert Andercyk, Jr.
Bernard & Joan Bena
Ray & Dorothy Duerling
Monica Foos
Fallston United Methodist Church Friendship Players
Elisabeth Hunsinger
Charles Kerr
Celestin & Alex Kurian
Wayne & Martha Marr
William & Carolyn Matschulat
Lyle Millan
Julie and Rick Morrison
Michael & Ellen Murphy
John & Karen Pieper
Wilson Russell
Karyn & Steven Schulz
Mary & King Smith
David Zajano

VERONICA CAILLE
Jeffrey & Rose Walters

PAUL LEONARD
Thomas & Dorothy Pickette

GRACE LIST
Edward List

DAVID MORGAN
Beverly Durtsche
Paul Hillman
Phyllis S. Morgan
Rodney & Shawn Morgan
Pat Nichols

JOHN AND LOUIS JAY RIBES
Mary Lou James

WALLACE LEE TALBOT
Angus & Nancy McDonald

LYNN TILL
Jean Till

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.