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The Gathered View

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

President's Message

by Jerry Park, PWSA President

Unconditional Love

We as parents constantly struggle with the balance of discipline and love. We seek the eternal approval and adulation from our children as we establish the bumper pads through the long journey of life, hopefully getting only strikes with no spares.

The journey with our children with PWS is not as clear. They rarely understand the discipline, the reason for it, or how to correctly respond to it. This usually leads to retaliation instead of behaving, to the point we often forget what the initial confrontation was about. The behavioral pattern, as they get older, takes unusual twists and turns, and even though the current drugs and medications do help in the defiant behavioral pattern, we all must look out for the unexpected.

Penny and I have experienced behavioral patterns changing course instantly. Whit, our 12-year-old son with PWS, then 9, set our house on fire February of '94. Due to this incident Whit went through the fire department program for fire starting, and we all felt it was an accident. He again set our house on fire in August of the same year, resulting in severe damage. The behavioral pattern was unexpected, and the obsession with fire is still an utmost concern for the safety of Whit and our family. The financial strain, the year or more in and out of alternative living arrangements put our family, as many of you have experienced, to the test! The human spirit is resilient and forgiving, without anger and contempt, and is the strength of each of our families.

> *"It is important as parents and caregivers to take a second look and see the love and innocence these children give without boundaries."*

We, as Prader-Willi parents, are challenged every day by the selfish and relentless pursuit by our children with PWS for their own interest. They will say and do things that are incomprehensible. It is important as parents and caregivers to take a second look and see the love and innocence these children give without boundaries—the expression of emotion that will stir your own emotions like no other. We then begin to realize, once again, that unconditional love may hold more than one meaning. We will always love our children "unconditionally," and our child with PWS will *always* expect us to.

104th Congress Ends in a Blizzard of Legislation

Congress worked frantically before and after its August recess to complete a number of laws that will affect the disability community, including:

The Kassebaum-Kennedy Health Insurance Reform Act of 1996, enacted on August 21, will potentially impact 25 million people currently covered by health insurance plans who (themselves or a family member) have a pre-existing condition by guaranteeing them insurability and portability after a lifetime exclusion of 12 months, as long as they or their employer can afford the premium.

Welfare reform—the Personal Responsibility and Work Opportunity Reconciliation Act of 1996—enacted on August 22, ends federal entitlement to cash assistance and support for poor women and children and replaces it with more limited block grants to the states. The law also repeals the Individual Functional Assessment (IFA) as part of the eligibility determination for children's Supplemental Security Income (SSI), and it requires that children have a "medically determinable physical or mental impairment which results in marked and severe functional limitations." The Social Security Administration must issue regulations by Nov. 22 to interpret this new standard.

While the reauthorization of the Individuals with Disabilities Education Act was not completed and will be taken up again in January, Congress completed a budget that gave IDEA programs a record $784 million funding increase for FY 97. (Source: *Washington Watch*, United Cerebral Palsy Associations)

inside

The Prader-Willi Syndrome Association (USA)

2510 S. Brentwood Blvd., Suite 220
St. Louis, MO 63144-2326

(800) 926-4797 or (314) 962-7644
8:00 a.m.–4:00 p.m. CDLST

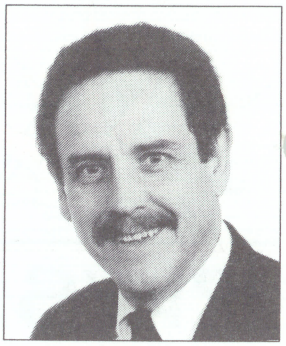
Fax: (314) 962-7869

E-mail: pwsa usa@aol.com

PWSA (USA) World Wide Web site:
http://www.athenet.net/~pwsa_usa/index.html

Out of the Office

by Russ Myler, Executive Director



Fall once again. It really is true that the years seem to go faster as you get older. However, for some the years drag on. I think of those persons living with the syndrome who have not yet had a diagnosis or who have not yet discovered the support and help available through the PWSA community. I believe for those people the years are most difficult.

Fall is time for the Angels Fund Campaign. The program was begun in 1991 under the guidance of then board member Penny Park (Oklahoma). The board recognized that if we were to achieve the goals of the Association we had to have additional resources beyond the annual member dues. Thus the Angels were created.

Member dues pay for the basics: keeping the doors of the National Office open, keeping the 800 number operating, publishing *The Gathered View,* and covering basic operating expenses. For us to reach our dreams of the future we need the added support of our over 8,000 potential Angels.

The Angels program was truly made for those people I mentioned before. A successful effort will give us the ability to further our outreach to those most in need. Angels help us spread awareness to more professionals who haven't a clue about what their client or patient has—or how to help them. Angel donations aid member families facing a crisis secure necessary resources, grant conference scholarships, expand our published material, and assist us in training professionals about management issues.

If you have been a contributor in the past I hope you will continue helping us expand our services and increase public and professional awareness. If you have not contributed in the past, won't you consider a donation this year? Those five years since the beginning of the program have gone by very fast and many persons with the syndrome who could have been identified and helped have had five long years of not knowing what to do. For some of us the years fly by; for others they drag.

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

Communications regarding *The Gathered View* or PWSA membership should be directed to the national office of PWSA (USA), 2510 S. Brentwood Boulevard, Suite 220, St. Louis, MO 63144-2326. Telephone 1-800-926-4797, or (314) 962-7644 in the St. Louis area. Fax (314) 962-7869. E-mail: pwsa usa@aol.com.

Research

The following new research on imprinting in PWS is being launched at Case Western Reserve University, thanks to a grant from an anonymous donor.

Prader-Willi Syndrome— Imprinting and Mouse Models

Robert D. Nicholls, D.Phil., Associate Professor of Genetics, Case Western Reserve University and Center for Human Genetics, University Hospitals of Cleveland, Cleveland, Ohio

Prader-Willi syndrome (PWS) arises from loss of paternal genetic information in chromosome 15q11-q13, either by paternal deletion, duplication of the maternal (and loss of paternal) chromosome (i.e., uniparental disomy), or a mutation in the imprinting process (known as an "imprinting mutation"). The parent of origin effect is known as imprinting. Genetic abnormalities in imprinting of other genes also result in neurobehavioral disorders or a wide range of cancers, predominantly those affecting children. Very little is known about the imprinting process and what genes are involved. An understanding of imprinting in PWS will allow us to better understand how PWS is caused, and we hope that this genetic knowledge will allow us to devise therapeutic approaches based on an understanding of the genes and mechanism of imprinting.

The "imprinting mutation" class of patients has been recently discovered, and we have identified the specific mutations (very small deletions) in several of these PWS patients. Since these imprinting mutations are inherited through families, our findings now allow "at risk" and prenatal testing in the specific families under study. Further study of PWS patients and families with imprinting mutations is critical to understanding how imprinting occurs at every generation (in the male and female germline). For example, we are finding families with smaller and smaller deletions, allowing us to find the exact DNA region important for this process. Since the imprint is reset at each generation (the nature of this imprint depends on whether you are male or female), we know the process is reversible. If we learn enough about how the process reverses, perhaps we can learn how to turn on this reversal in patients, so that the paternal genetic information now works. These experiments are one of the most important areas of our current and future research.

Although we can learn a lot about PWS and imprinting from our patients, imprinting occurs in the germline and the critical tissues for the syndrome are in the brain. Clearly, we cannot use these tissues in human studies, and therefore a model system is essential. Our discovery of imprinting mutations now gives us a way to create an animal model of PWS. This is possible because the genes in humans that are important for developmental functions are conserved in the evolution of animals, including the mouse, a superb experimental animal model. Studies have already been initiated to generate an animal model of imprinting mutations, which will allow us to not only understand more about the process of imprinting reversal in the germline and the effects of imprinting on the critical genes in the brain, but will also establish a mouse model of PWS with which the basic biochemical and physiological defects can be examined. This has not been possible in human studies. These studies are critical to complete, but due to the innovative nature of these experiments, they have not yet received the funding necessary to complete the work in a timely fashion. The long-term goal of our work is to understand everything about the biological basis of PWS, so that we can devise suitable therapeutic approaches.

The Third International Conference on Prader-Willi Syndrome

May 15 - 18, 1998
Seville, Spain

■ ***Call for Presenters***

Scientists and professionals versed in Prader-Willi syndrome and interested in presenting at this conference, please send name, address and fax number to:

Dr. Ellie Smith
Genetic Department
New Childrens Hospital
P.O. Box 3515 Parramatto
Sydney NSW 2124 Australia

■ ***Registration Packets***

To have your name placed on the mailing list for information regarding this conference, send name and address to:

Jackie Waters, Secretary IPWSO
27 Willetts Road
GB - Chaddesden, Derby
DE 24 NH, England

International Prader-Willi Syndrome Organisation (IPWSO) member countries:

Australia	New Zealand
Belgium	Norway
Canada	Poland
Denmark	South Africa
Finland	Spain
France	Sweden
Germany	Switzerland
Greenland	United Kingdom
Italy	United States
Netherlands	

If you are interested in corresponding with a PWS family from another country, send name and address to:

Mildred Lacy
IPWSO Vice President
P.O. Box 18132
Louisville, KY 40261

Recent Journal Articles About PWS (Jan.-Sept. 1996)

Sources: MEDLINE database, National Library of Medicine, and Uncover database, Colorado Alliance of Research Libraries.
Each cite shows: *Journal, Volume* (Issue), Page. *Cites from the Uncover database list only the first page of an article.

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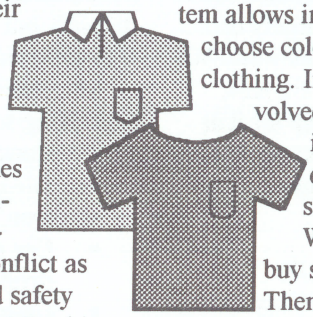
Assisting the Person with Prader-Willi Syndrome in Making Choices About Clothing Selections

by Barb Dorn, R.N.
Verona, Wisconsin

Those of us who care for individuals with Prader-Willi syndrome (PWS) know that there are times when they don't blend in with their peers. Maybe they stand out in a crowd because of their inquisitive personalities, or their pear-shaped bodies, or their rigid behavior. But we can help them blend in by teaching them to dress appropriately. Because of their stubbornness and need for control, teaching a child or young adult with PWS to make appropriate choices and decisions about the clothes they wear can often be a challenging process. Clothing selection is often a source of conflict as well as an issue of health and safety (e.g., frostbite, heat stroke …) Looking appropriate, however, is an important life skill which can enhance a person's self-concept. It is also an area in which persons with PWS can have control and input in decision making. In an attempt to address this issue, I developed two approaches to teach these skills using a visual, concrete format.



Color Coordination

My son, like others with PWS, has diffulty choosing clothes that match and are color-coordinated. The following system allows individuals to independently choose color-coordinated pieces of clothing. Inititally, there is work involved in marking items, but there is time saved by decreasing disagreements and/or discussions.

When buying clothing, try to buy solid colored shorts or pants. Then pants and shorts are sorted by color. For example, blue pants and shorts are placed in one pile, green in another pile, and so on. Each pile is then assigned a color coding and marked on the inner aspect of the waist band. This can be done using small pieces of iron-on seam binding, fabric paints, or permanent color markers. These markings do not need to be large; a small tag or decal is sufficient. You do, however, want to use a color that can be readily seen on the item of clothing (e.g., red marking on blue pants, green on black, pink on red).

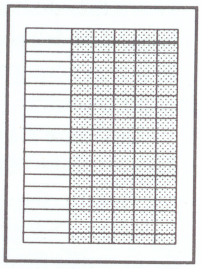
After marking all shorts and pants, shirts and sweatshirts are then marked. Most shirts can have two or three different markings which coordinate with one or more pairs of pants/shorts. Using the examples above, a red, white, and blue USA T-shirt could receive both a red and a pink marking. In this case, the person could choose either blue or red pants/shorts to wear with this T-shirt.

These markings give the person with PWS a visual tool to use to match their shirt with their pants/shorts. In shirts, these marks are placed on the inside of the shirt collar, usually near the label.

Color choices for socks can be made simple by purchasing white and black socks only. These two colors coordinate with most other colors.

Appropriate Clothes for Weather Conditions

Many children and adults also make inappropriate choices in selecting the proper clothing for the temperature or weather they may face. A clothing chart that I developed helps guide persons with PWS in making choices about appropriate clothing and shoes for weather and temperatures most often encountered in the Midwest. Once again, this chart provides a visual, concrete tool to assist them in this daily living skill.



(The laminated chart lists 14 articles of clothing down the left column. The next five columns indicate the appropriateness of each clothing item for a specific temperature range—from "Below 32 Degrees" to "Above 76 Degrees." Temperature columns are color coded for easy identification.)

In order to limit choices, parents and care providers may need to remove out-of-season clothing. For example, remove sweatshirts, sweaters, turtleneck shirts, etc., in the spring and return them again in the fall. This process can sometimes be made easier and more acceptable if "new" spring and summer clothing is exchanged for these items.

Summary

As parents and care providers for persons with PWS, it is our job to try to teach and guide them in making appropriate choices in their clothing selections. The challenge is finding a concrete, visual tool to assist them with this process. This article has presented two ways to address these concerns. These approaches allow the individual with PWS to make independent decisions in choosing the type and color clothes they wish to wear. By providing them with this assistance, we can also reduce disagreements. Hopefully, independent, appropriate clothing selection will become an automatic daily living skill, and it will help them blend in with their peers.

Barb's Clothing Chart is available in two sizes: 8 x 11 inches ($7.00) and 11 x 14 ($13.50). Prices include pen for marking chart and shipping cost. Mail request with payment to: Barb Dorn, 305 Amanda Way, Verona WI 53593. Telephone: 608-845-9597

Journal Articles—*continued*

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Editor's note: This listing does not include articles in languages other than English nor articles published in *Prader-Willi Perspectives*, a quarterly journal devoted to PWS. For an index to articles in *Prader-Willi Perspectives*, contact the publisher, Visible Ink, Incorporated, at 1-800-358-0682.

By Linda Keder, based on notes taken at the PWSA (USA) national conference in St. Louis last July and at the Prader-Willi Service Providers' Conference in Bethesda, Md., in June.

Voices of

A Round-up of Wisdom and Management Tips

On Early Nutrition

Gail Overton (nutritionist): There's little published about infant feeding in PWS; normal nutrition guidelines are very appropriate. Get a handle on it early. Be aware that developmentally delayed children have different feeding issues—e.g., they may require special feeding devices or positioning.

Suzanne Cassidy, M.D.: The biggest danger in the "failure to thrive" period is not feeding the hypotonic infant enough. Families who get an early diagnosis sometimes make this mistake. Fat is extremely important for brain development and should not be restricted in the first months of life. Also, the hypotonic infant can use too much energy trying to suck. It's important to work closely with a medical person during this period to monitor growth, feeding time, and energy requirements. Appropriate feeding requires careful balance and frequent checking. It may be necessary to use a gavage feeding tube (through the nose into the stomach) or a G-tube (directly into the stomach) until the child can suck well. No studies have correlated difficulty in feeding with problems later on.

Find a good doctor who's willing to learn about PWS, if they don't already know about it. Attitude is more important than past experience with the syndrome.

On Diet

Ms. Overton: Good patterns are established from the beginning of life. What the child is exposed to, and the preferences and dislikes of the parents, have an impact. Parents need to be role models for good eating habits. Calcium, zinc, and iron are frequently deficient in people with PWS, says Ms. Overton, noting that she gives prenatal vitamins (the kind pregnant women take) to all the people with PWS in her practice.

On Thick Saliva

Dr. Cassidy: It is common for people with PWS to have thick saliva, dry mouth, and crusting on the lips, due to an autonomic nervous system problem. Reduced saliva can impede speech development and cause other oral problems. Products designed for people with decreased saliva flow due to head and neck surgery can increase saliva flow in persons with PWS as well. One such product is Biotene, available over the counter (ask for it at the pharmacy) in three forms: toothpaste, mouthwash, and sugarless gum.

On Exercise

Dr. Cassidy: Exercise is the "cornerstone of treatment in PWS." Once a child is mobile, they should be doing 30 minutes of exercise a day. The more muscle a person has, the more calories they burn. Dr. Cassidy mentioned one adult in a Prader-Willi group home who was able to eat 2,400 calories a day because of consistent exercise.

Margaret Johnson, R.N.: Working with adults in a group home setting, she recommends starting with 20 minutes of daily exercise and working up to an hour a day. Expectations for muscle development need to be different for those with PWS—she has not seen significant muscles development in males even with an hour a day of exercise and testosterone treatment. She urges caution in using some exercise equipment—specifically, stair-climbing machines have caused "popped patellas" (kneecap damage) in a few cases.

Jennifer Deau, exercise physiologist: Exercise should emphasize cardiopulmonary fitness level and keeping moving. She recommends one hour of "continuous and vigorous" exercise three times a week. She also stresses working on balance and on developing shoulder girdle muscles for full range of motion in the arms. In a two page handout entitled "No Such Thing as a Free Lunch," Ms. Deau recommends: walking (swinging arms in opposition to legs), lap swimming or pool walking, aerobic dancing, stationary biking, nature hiking (if balance allows), skiing and snow play. Use small weights (½–1 pound) to build upper extremity strength, but limit it to three times a week with supervison.

Ms. Deau offers the following tips for motivating someone to exercise:

- Couch it in terms of play
- Give them choices
- Don't make it a punishment
- Use rewards such as a calorie-free drink
- Commit a time for it; structure the session
- Use firm, consistent guidelines and expectations
- Use a chart or notebook to record progress
- Believe your child can accomplish the goals
- Have fun!

On Feet and Shoes

Dr. Cassidy: There are many feet problems in PWS. It's important to protect the feet so the person can exercise.

Ms. Deau: A quality athletic shoe with flared sole and good lateral support can help with the balance problems in PWS. She recommends New Balance brand (now available for young children too—model 996).

Kelly Riley, parent and physical therapy assistant: Buy good shoes! For young children, she recommends high tops and says she has always been successful at Stride Rite stores. Some young children with PWS may have foot problems that require a type of orthotic or shoe insert. The physical therapist should key in on any foot problems and make a referral to an orthopedist.

On Growth Hormone

Merlin Butler, M.D.: Metabolism is the one factor in weight management that

Prader-Willi Syndrome Association (USA)
Scientific Advisory Board

Policy Statement:

Growth Hormone Treatment and Prader-Willi Syndrome

Since the commercial release of recombinant human growth hormone (GH) in 1985, therapeutic use of this medication has been studied in a variety of medical conditions and genetic syndromes. Based on current medical knowledge, the Scientific Advisory Board of the Prader-Willi Syndrome Association (USA) has drafted and approved this policy statement to guide health care providers in the use of GH treatment in patients with Prader-Willi syndrome (PWS).

Current considerations regarding the use of GH treatment in PWS can be divided into the following categories:

1. Treatment of GH deficiency in children with PWS.
2. Treatment of GH deficiency in adults with PWS.
3. Treatment of body composition abnormalities in patients with PWS.

CHILDHOOD GROWTH HORMONE DEFICIENCY

As of this date, the United States Food and Drug Administration has approved routine therapeutic use of GH for only 2 indications:

1. Growth hormone deficiency in children.
2. Growth failure associated with chronic renal failure in children.

Recent studies indicate that GH deficiency occurs frequently in children with PWS and that treatment with GH is efficacious in improving the growth rate of these affected children.[1] The Scientific Advisory Board recommends that routine clinical use of GH treatment only be used in cases of documented GH deficiency. Moreover, the Board recognizes that routine provocative GH testing is not ideal in children with PWS because: (1) the results may be falsely low due to obesity, (2) the test procedure is associated with a relatively high risk for morbidity and mortality, (3) different GH assays give widely discrepant results, (4) the diagnostic boundary for a normal/abnormal GH result is largely arbitrary, (5) there is lack of agreement on ideal testing protocols, and (6) the test procedure is costly and requires a several hour period in the outpatient clinic or hospital.

The Board agrees with the recent recommendations of an international panel of pediatric endocrinologists[2] that the diagnosis of childhood GH deficiency be based on a combination of auxological and biochemical criteria:

1. Short stature.
2. Abnormally low height velocity.
3. Abnormally low serum levels of insulin-like growth factor-I (IGF-I) and/or IGF-binding protein-3 (IGFBP-3).
4. Absence of other conditions which may account for 1-3.
5. Radiographic documentation of open epiphyseal growth plates.

The pros and cons of GH treatment should be thoroughly discussed with the child's parents or guardians before making a decision to treat. Treatment should commence using standard dose guidelines (~0.3 mg/kg/wk divided as a daily subcutaneous injection) with careful monitoring of clinical status at 3-4 month intervals.

Standard GH treatment includes dose adjustments based on weight. However, there is some evidence that lean mass is a better indicator of GH requirements. Therefore, the Board recommends that the GH

dose in children with PWS and GH deficiency be adjusted based on clinical growth response rather than on strict weight-based criteria.

Once started, GH treatment may be continued until natural epiphyseal closure or when the child has reached an acceptable adult height. Clinical monitoring should include accurate height and weight measurements, physical examination and, if feasible, estimation of body composition.

Children with PWS may be at increased risk for spinal curvature abnormalities, including scoliosis and kyphosis. There is no evidence that GH itself causes these abnormalities. However, such abnormalities often become more apparent and may worsen during rapid growth. Therefore, children with PWS, whether or not they are treated with GH, should receive a careful back examination at regular intervals. Radiographic studies and orthopedic consultation should be obtained if clinically indicated. The decision to treat or continue GH treatment in a child with spinal curvature abnormalities should be made in consultation with an experienced orthopedic surgeon and after full discussion with the child's parents or guardians.

Children with PWS are prone to developing obesity and its associated complications, including glucose intolerance and non-insulin-dependent diabetes mellitus. GH is a glucocounter-regulatory hormone. Therefore, children with PWS and GH deficiency should be carefully monitored for signs and symptoms of glucose intolerance during GH treatment, particularly if they are massively obese (e.g. >200% of ideal body weight) or have a family history of diabetes mellitus. Routine biochemical screening tests may include a fasting blood glucose, urine glucose dipstick or a total glycated hemoglobin measured by boronate-affinity chromatography. If glucose intolerance or diabetes mellitus occurs, the GH treatment should be stopped. If treatment is restarted, the dose of GH should be substantially reduced.

ADULT GROWTH HORMONE DEFICIENCY

Recent studies indicate that adults with GH deficiency may benefit from replacement therapy. However, the occurrence of GH deficiency in adults with PWS has not been well documented. Furthermore, the therapeutic use of GH in adults regardless of clinical condition has not yet been approved by the U.S. Food and Drug Administration.

BODY COMPOSITION ABNORMALITIES

Treatment with GH has been shown to positively affect nitrogen balance, increase lean body mass and reduce body fat in several conditions. Moreover, treatment with GH may preserve lean body mass[3] during caloric restriction. However, these effects have not been well documented in individuals with PWS. Furthermore, the therapeutic use of GH in body composition disorders other than those associated with childhood GH deficiency or childhood chronic renal failure has not been approved by the U.S. Food and Drug Administration.

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Experience

from Summer 1996 Prader-Willi Conferences

we can't control (exercise and amount of food being the two we can change). GH, however, can affect metabolism. In "normal" individuals, GH increases metabolism by 20 percent. Prader-Willi metabolism is reduced compared with both lean and obese controls, due to decreased muscle mass. (As part of the PW research at Vanderbilt University, metabolism is measured in a special chamber that can monitor a person's movement and oxygen/carbon dioxide exchange during an 8-to-24-hour period.)

Dr. Cassidy: No one knows for sure at what age to start growth hormone or in fact whether GH is the best thing for children with PWS. It does seem to increase final height. The age for use of GH in kids without PWS is 3. If a child is growing below the normal growth curve and at a slow rate, then GH should be investigated. Note that GH can worsen scoliosis.

Jim Boyle, M.D., parent: GH helped his son grow from below the 5th percentile at age 3 to above the 95th percentile at age 5. He also developed a more normal appearance and is now (at age 9) able to eat 2,000–2,200 calories a day. The ability to increase calorie intake and alleviate stress from food issues can be a tremendous benefit to the entire family from GH treatment.

Jeanne Hanchett, M.D.: It is "very well established" now that GH has many advantages for persons with PWS, but it is important to consider each case individually. One disadvantage is scoliosis (she has seen two cases of rapid progression with GH); an x-ray is the best way to detect scoliosis, especially in obese patients. There has always been a concern about tumors; GH probably does not cause tumors, but may accelerate existing ones. Diabetes also is a concern and is usually a contraindication for use of GH.

In Dr. Hanchett's experience, the chief concerns for families of those with PWS are obesity and behavior problems, not short stature. She cites two parents who opted not to increase their children's height: one—a single mom—felt she could no longer control her son's behavior if he exceeded her height; the other parent felt her child would be treated "more gently" by society with shorter stature.

Jim Kane, parent: Because of the great expense, getting your child on growth hormone can be a struggle. The keys are to find the right professional to help you, know your insurance policy, and be persistent. In his daughter's case, the right person proved to be a specialist in diabetes who was willing to work with the family and with an endocrinologist in another state. The doctor was able to convince the insurance company to provide GH based on the child's obesity problem. In his daughter's case, GH significantly worsened existing scoliosis, necessitating back surgery last year. The surgery was very successful, and recovery may have been aided by continued GH treatment.

On Behavior

Barbara Whitman, Ph.D.: Parents need to become knowledgable early on. If you let the child manipulate you, you've lost the battle. The deadliest issue in a family is when mom and dad can't agree on an approach—this sets up a pattern that escalates over time. Get your disagreements resolved (go to family therapy, if necessary)—it's critical for the child's mental health that the parents present a united stand. All kids push the limits; kids with PW take it to "an art form."

Janalee Tomaseski-Heinemann, M.S.W., parent: Be specific about rules, options, and limits. Avoid problem situations, such as a buffet meal where you keep having to say "No." Don't give the opportunity to lie (i.e., don't ask "Did you … ?"). Use a kitchen timer to manage choices (when the timer goes off, they must stick with the latest choice)—it won't work on the first or second try, she notes, but it soon becomes very effective. Food consequences (withholding or delaying food) can be a very powerful tool. Know "when to shut up"—back off if your child's really upset.

Louise Greenswag, R.N., Ph.D.: Provide opportunities for success. Model and rehearse social interactions and situations. Because kids with PWS may want to be in charge when playing with peers, practice taking turns.

Karl Sieg, M.D.: Behavioral problems often begin between ages 6 and 11. Diet is an important component of behavior, since food has neurotransmitters and affects mood. Also, the effectiveness of medications can be impaired by vitamin deficiencies in the diet.

Growing awareness of their differences from peers can lead to depression and irritability. Having choices is important to self-esteem.

Some recent studies are showing a crossover between PWS and obsessive-compulsive (OC) behavior. Some medications can increase OC and irritability.

Attention deficit disorder (ADD) is not typical in PWS, but there are some similar problems.

Ivy Boyle, M.D., parent and child psychiatrist: People with PWS have problems with attention, both *over-* and *under-*attention. Many kids with PWS are now on Ritalin, which is a relatively safe drug that requires no monitoring. There are many medications now being used and tried for the psychological and behavioral problems associated with PWS. Some have proven very effective, though experience varies among individuals and even between separate trials in the same individual. Be willing to try something again. It's important to be aware of the potential side effects of specific drugs, monitoring requirements, and the possible need for adjusted doses. Abrupt withdrawal of any drug can have significant effects and should be avoided.

Dr. Hanchett and various parents: So-called "rectal picking" (self-probing to the point of internal injury) is not uncommon in PWS and may be mistaken for inflammatory bowel disease. Investigate rectal bleeding as a possible behavior issue.

My Son's Road to Greater Independence

A Transition Diary —by Pat Shiley, Parma, Ohio

Pat Shiley, Executive Director of the PWSA of Ohio, responded to the "Diary of a Placement" in the February issue of The Gathered View with her own account of how her son, Tim, created a new life for himself at age 30 and how the family adjusted to the change.

December 1994

Jack (my husband), Tim, and I attended a PW group home open house in our county (Cuyahoga). The day was cold and blustery, but the hospitality was warm. Many of the people who came we knew, and while we talked with our friends, Tim wandered around the house, enjoying himself thoroughly. Several commented that he acted like he was the host of the party. On the way home, Tim said he wanted to live in a house like that—meaning no parents, but staff and other residents.

The next morning, Tim went to his workshop supervisor and asked to be put in touch with his case manager, Kim, so he could talk with her about moving into a group home.

January 1995

Kim met with Tim and then with me. At that time, Cuyahoga County was developing a second PW home. Funded through the Supported Living Program, it was tentatively scheduled to open in September 1995. That seemed so far away. I started the process of obtaining guardianship. Tim's spirits soared. He was 28½ and NEVER had had the option of leaving home to live elsewhere. He was so excited. Our feelings soared with Tim's, because we deeply felt that he needed his independence, even though it wasn't total independence.

May 1995

As months passed, there were papers to complete, interviews, and in May, a dinner visit for Tim to Moreland House. Afterwards, he chattered on and on about who he saw, what they did, what they wore, etc. Then he received an invitation for an overnighter at Moreland House. He was so happy, and it was a big success. Again, he talked endlessly about his stay.

We received a letter stating Tim was accepted for residency, and he was thrilled. We met with the agency staff, case management, the three other house residents and their families—a little like meeting your child's future in-laws for the first time. Plans were discussed, including payment of utilities, chores, behavior, and all the details involved in the move. Tim was so excited. We knew in our hearts this was the right thing to do at the right time. Tim made it very easy for us to accept HIS decision to move out of our home.

> *"Because Tim wanted to change his life, he accepted all of the challenges along the way—and he has been successful. He has a new house, a new job at a new workshop, a healthy new body, and a life independent of his parents."*

July 1995

Suddenly Kim told us that the house provider, REM Consulting of Ohio, would like Tim to attend The Rehabilitation Institute of Pittsburgh before moving into Orange House because it will be operated on the program set up in Pittsburgh. Tim needed to learn about the diet and exercise. This was our one hesitation, because we didn't think it necessary, but we decided to leave the decision to Tim. Tim said he'd try it. Kim called to say that Tim would be admitted to "The Rehab" on August 8, and his last day at the workshop, where he had been one of the top producers, would be in July. Already? Where did the time go? I thought we were ready! So soon?

The Friday before we left for Pittsburgh, we had a date in court for the guardianship hearing. All of a sudden our lives were in a whirl, going faster and faster. Guardianship was granted in 15 minutes after seven months of preparation! Tim, finished with workshop, helped us shop for a new bedroom set for his new home.

August 1995

Monday evening we headed to Pittsburgh, and Tim was admitted the next morning. We spent the day in meetings, catching up with Tim at lunch.

Food and PW. When Tim was born, he weighed 3 lbs., 6½ oz. Feedings were long and often. Every ¼ oz. he lost or gained was critical. The Rehab told us Tim, who is 4'6", needed to lose 74 to 94 lbs. Were they kidding? We shared such disbelief at the numbers.

Tim is unusual in that he is a discriminating eater—translation: he doesn't like fresh fruits, vegetables, salad, and most anything on the diet plan. We left Tim with very heavy hearts, knowing he was going to be hungry, and I cried on the way home. That was the hardest part of this entire process. We stopped to eat at a restaurant. The hostess asked, "How many?" Jack said, "Two." The word stuck in his throat, and I had tears in my eyes. I couldn't eat my meal. How could I? My child was hungry and 200 miles from home.

The next day I had a job interview,

Continued on page 9

was hired, and started work the following day. Thank God, or I would have sat at home and cried thinking about Tim. That weekend in church, the gospel reading was about the Father sending his only Son to suffer and die for us. I started to cry. I thought of sending my only son to Pittsburgh to be hungry. I'm crying again as I write now.

We traveled to Pittsburgh Saturday and came home Sunday evening every weekend of the 5½ weeks Tim was there. We took him bowling because he is an excellent bowler (has carried a 151 average in two leagues, with a high of 204). What could we do that didn't involve food? The staff was good with him. He called home to say that he exercised so much he wore his socks out! Could we bring him new socks?—but they need to be thick and cushioned!!!

Work really helped me. Jack noticed the emptiness in the house, too. Tim's little hands used to empty the dishwasher every day. He would fold and put away the clean clothes, take out the trash, carry out the newspapers—all without being told and he'd do it gladly. The house was just so empty without him.

But his new home, Orange House, was getting ready for the arrival of all four residents. We took pictures of the house, which is 15 miles from us, and sent them to Tim.

September 1995

Tim left Pittsburgh, 18½ lbs. lighter, with the Orange House staff and went directly to his new home. That evening we visited him; he was tired but happy. After 30 days, Tim would be able to stay overnight at our house. In the meantime, he could visit us during the day but return to Orange House in the evening. We kept him in a Saturday bowling league, and he could attend church with us, so he had a bridge between his old life and his new.

I got panicky trying to plan his meals. What to prepare? How much? How to make it? If he's still hungry, what do I do? There were very few complaints from Tim. He's learned to eat a few new foods, but he still generally doesn't like fresh fruits and vegetables.

Thanksgiving 1995

All he wanted for Thanksgiving was a frozen fish dinner and a piece of pumpkin pie. He still lost 2 lbs.!

Keep On Keeping On ...

"I guess my own insight began when I decided that personal suffering was not going to get me down.

"It was endurance and courage that got me through.

"Where does the strength we need come from? I think—it comes from who we are and from within ... The ability to go forward in spite of what has happened is amazing.

"I think that many people take setbacks too seriously. We've got to let go of things and carry on, realizing that it's not the arrival that matters—it's the journey itself. Life is not always easy, and for some of us, it may take the rest of our lives to learn how to live it.

"In our journey, we must just keep going along as if each step is the best step. That's what keeps us Keeping On. That's what makes us the great people we are (when we're finished)!!

"When we're going through a challenge, we shouldn't just sit there. We go out and talk to people. We go here, there, wherever, whether it's church, group therapy, or (for us) PWFA (the Prader-Willi Florida Association, a PWSA chapter). We seek out the people we relate with—analyze—talk. WE GET OUR SPIRIT BACK ... We keep the doors to our hearts and souls open, so that we feel free to talk about everything and to be who we really are. Closing ourselves off from others can be the cause of great pain.

"So many people are struggling with what we should or shouldn't do. Well, I say, do what's good for you. Be around people with healthy perspectives. Eat good, exercise, go to bed early. That's a simple formula. Insight comes from looking within. Spirit is what we see.

"Keep On Keeping On. You have everything to gain, and life becomes a beautiful adventure."

—Roda Guenther, President, PWFA

(Excerpted from the *Gator-Willi News*, newsletter of the Florida Chapter)

Christmas 1995

Tim had a relaxed holiday and didn't gain any weight.

March 1996

It's been 7 months since Tim went to Pittsburgh. He's lost 74½ lbs. His waist has gone from 46" to 32". He's at his goal weight of 100 lbs. Friends have told me the face is different, the body is different, just the voice is the same! He is so proud. His self esteem is very high. He's accomplished what a lot of us just talk about—but he's done it! He's also learned to live with other people and share—a very difficult task for him.

He comes home almost every weekend for the whole weekend. I've learned not to get panicky about meals, but I really plan. Orange House staff is a big help. They will pack a meal for him, if necessary, and give me suggestions on "how much" or "what" or anything else to help Tim continue to be successful. He exercises every day—walking, swimming, using a video tape.

Because Tim wanted to change his life, he accepted all of the challenges along the way—and he has been successful. He has a new house, a new job at a new workshop, a healthy new body, and a life independent of his parents. All of this was done because HE wanted the changes. His will power is amazing. And HE made it easier for us. Many people close to Tim thought ***we*** were wrong to have him move out. ***Tim*** explained to them ***he wanted*** to move and he didn't care if they didn't like it! After everyone has seen his house and how happy he is, they know he made the right decision!

One last word: I would like to salute the staff of REM Consulting of Ohio, who operate Orange House. Because of their consistent successful implementation of procedures, Tim is a success story. ▲

A Letter From Dr. Beltran

Dr. Delfin J. (Sam) Beltran was the chairman of the board of PWSA from its founding until July of 1980, when he succeeded Gene Deterling as president of the Association. Dr. Beltran served as PWSA's president for 10 years and wrote many wonderful columns for *The Gathered View*. More recently, he has written or contributed to several articles in *Prader-Willi Perspectives*. In a letter received this summer by former board member and officer Stewart Maurer, Sam and his wife, Linda, update PWSA friends on their move to Kansas and on the progress of their daughters— Sarah (24), who has PWS, and her younger sister, Heather. Following is an abbreviated version of that letter.

To all of my old friends at PWSA:

From here in the depths of Kansas tornado country—greetings. It has been six years since Sarah first came to Wichita to attend the Institute of Logopedics. Now they have renamed it HeartSpring and are in the process of relocating out of shooting range of the 21st Street gangs. Three years ago she entered as a full-time client of the Kansas Elks Training Center for the Handicapped (KETCH). This project provides vocational training and residential management for the handicapped. At the present time Sarah is living in a newly built four-bedroom house with two other clients. One is verbally and mobility impaired, and the other girl is mildly mentally retarded and nonverbal.

Sarah loves her work, both because it generates income to supplement her SSI and also because her work supervisor is a kind, understanding, and intelligent person who gets Sarah to work hard and well. The unit is about a block square space of well-lighted light industrial construction—everything from living skills and physiotherapy to education, mailroom work, and light industrial production such as air filters for Boeing or reconstructing the sheet metal clips or power cables for Beech and Cessna production lines. From time to time the workers will be sent to business locations in the community such as Pizza Hut or a bank to practice their skills in the real world. These assignments have proven difficult for Sarah because of the rows of office desks well laced with snacks. Most of the time she works at a production facility that makes ring binders, and she has become productive in all phases of that line.

This summer Sarah's sister Heather is home from the University of Pennsylvania where she is majoring in linguistics, psychology as related to the handicapped, and philosophy and languages. This summer she again is working as a residential careworker for KETCH but is also working as a classroom aide for the HeartSpring summer sessions. She plans to go on for her masters and possibly a doctorate in language and learning disabilities, with possibly a longer-range goal of coordinating that interest with genetic studies as that field continues to blossom.

Linda continues to work nearly full time as a recovery room nurse at our hospital's same-day surgery unit. I was fortunate to find a position in an outstanding medical community. I am back to doing some obstetrical anesthesia … and it never ceases to amaze me how many first-time young mothers come to delivery with weights well in excess of 200 pounds. I used to think that the dairy-fed of Wisconsin were large, but there are more take-out quick food parlors per capita in Wichita than any other similar community in the country, and all of the young families express gross ignorance of home cooking or dieting. …

God Bless and Good Luck to you all, and maybe after I retire I'll return to going to meetings. Until then, there is enough farm work to fill the spare time, and then I still have my favorite toy—the computer.

Sam & Linda Beltran
Andover, Kansas

A Mother's Plea to Conference Planners and Attendees

Our 2-year-old daughter, Anna, was diagnosed last January with Prader-Willi syndrome. After the initial shock, we voraciously read anything we could find. As you know, the literature paints a bleak picture concentrating on the negative aspects of the syndrome. However, we read everything we could about hypotonia, developmental delays, hyperphagia, and the hot topic of the year, rectal picking.

We were both excited and somewhat nervous about attending the conference in St. Louis. We didn't know quite what to expect. We had met other older children with Prader-Willi through our chapter but didn't realize that the national conference could be so overwhelming. We enjoyed the sessions on the first two days. They were informative and accomplished what we came to the conference for. The session for the "Mothers Only" started off on a good note, however, quickly became the same thing that I had been reading about in the articles, concentrating on the negative aspects of the syndrome. This left me with a very pessimistic attitude. In talking with other mothers in the same session, they too left distressed. This is an unfortunate way to end a conference such as this, especially for parents at their first meeting. What I am afraid is that it will dissuade parents of newly diagnosed children from attending future conferences.

Certainly I am not idealistic and realize that there will be problems ahead. It just may not be the best time to drag the horror stories out in the last sessions, just as everyone is going home. These may be best left in their age-appropriate sessions. As more physicians are becoming aware of the syndrome, there are going to be more first-time parents attending the national conference, as was shown in St. Louis. They do not need to be confronted with all the problems at their first meeting. I hope that future sessions can be planned to give a more positive approach to the problems and encourage other parents to contribute their suggestions.

Carolyn Loker, Kalamazoo, Michigan

In loving memory of Claire

Claire Gallagher was just fifteen years old when she lost her life aboard TWA flight 800 along with twenty of her friends from our High School French club while they were pursuing their dreams to visit Paris. Claire was a wonderful daughter, a loving sister, a cherished friend. She was incredibly gifted both artistically and academically.

The photo above was taken several years ago while we were on vacation in Canada. Claire has her arm around her brother, Kyle, [who has PWS] and is tenderly helping him through a very difficult moment. I cherish that picture and all the many times she showed love and compassion to her family and others.

She will forever be so dearly loved and sadly missed .

Barbara, Bob, Patrick and Kyle Gallagher

Montoursville, Pennsylvania

Holiday Greeting Cards for Sale to Benefit PWSA (USA)

The national PWSA office has beautiful, full-color greeting cards, decorated with artwork by some of our young people with Prader-Willi syndrome. They are available in an assortment of five cards, one each of the following themes:

Hanukkah
Santa
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Merry Christmas and Happy New Year

$5.00 per 5-card set, plus $1.50 shipping

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