

Down Under Is Not So Far Away

by Louise Greenswag, R.N., Ph.D.

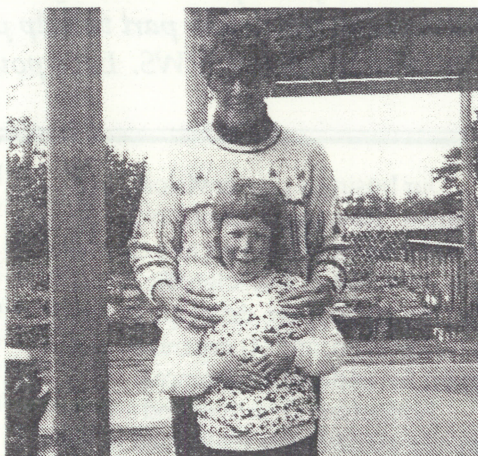
Strictly speaking, "Down Under" is not the name of a place but a term describing New Zealand and Australia, which are on the opposite side of the world from the USA. Last fall, I was invited by the New Zealand PWSA to spend several weeks presenting educational and professional training workshops in Wellington, Christchurch, and Auckland. Since my initial visit to New Zealand in the spring of 1990 merely whetted my appetite (it is a young country brimming with dramatic landscapes and unspoiled beauty), I leapt at the chance to return!

In New Zealand, services to the developmentally disabled are governed by a national agency, The Intellectually Handicapped Society (IHC). My visit consisted of informative and interactive training sessions designed to guide IHC caregivers in the creation of appropriate programming for PWS that fits within the philosophical framework of the country's service system.

Each two-day workshop included an overview of PWS, with extensive discussion of behavioral management. After a question/answer period, participants (both parents and service providers) met in small groups to exchange thoughts and perceptions. This open dialogue heightened the sensitivity of caregivers to the needs and worries of parents. Options for serving the PWS population in New Zealand then were discussed, using hypothetical case studies to create individualized plans of care based on information presented earlier in the day. This case study approach worked very well as a model for those in attendance to use to train others.

The young and vital New Zealand association and its coordinator, Linda

Thornton, are to be congratulated! In just a few years, over 60 children and adults with PWS have been identified, and a program of information and education has resulted in reports of other cases. Recent correspondence with Linda indicates that a designated residence is being prepared for four young men with PWS under the auspices of the IHC. In



her own words, "by establishing a home run by IHC and jointly trained with our input (PWSA of NZ), I now hope we are on our way to achieving good alternative residences for our people."

On a more personal level, Linda — a talented silversmith and writer — shepherded me during my journey with grace, persuasiveness, perseverance, consideration, and a wonderful sense of humor. She and her husband, Nick, have three lovely daughters — Lucy, Charlotte, and the youngest, Francie, who has PWS. The Thornton family lives on a small farm near Masterton in a lovely valley beyond the Rumataka Hills northeast of Wellington. I was welcomed into their lives and kept well fed and cozy during a very damp early spring. Certainly, the warmth of their welcome was characteristic of just about everyone I met in New Zealand. While it

basks in its British heritage and language, it is a delightfully informal country of warm, friendly, and gracious people.

My visit confirms my personal impression that families and caregivers are making a determined commitment to creating purposeful lives for persons with PWS, knowing full well that life is not always fair, that laughter often covers up tears and fears, and that happiness is not an end product but a by-product, measured in daily successes. Worldwide, parents are learning to cope by recognizing they share more commonalities than differences. Down under is *not* so far away!

Louise is a member of the PWSA Board of Directors and its Scientific Advisory Board, co-editor of the book, Management of Prader-Willi Syndrome, and a frequent and popular presenter at PWSA National Conferences and other PWS meetings.

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PWSA (USA) Goals Evolving

By Curt Shacklett, Chairman

The Board of Directors of PWSA met in Atlanta in January for several days of intensive interaction in our ongoing efforts to carve and shape the business plan for our organization. As our Association grows and develops, the Board is continually striving to fine-tune our goals, objectives, and action steps to enable our association to better serve our members and those impacted by the syndrome.

We are a "grass roots" organization ; therefore, every member is part of the "root" system and is vitally important. We must each do our part to help produce "grass," i.e., promote awareness of PWS. Let's pour on the fertilizer.

Our business plan envisions four "essential" function areas of PWSA: (1) communications; (2) acting as a clearinghouse for research and scientific data; (3) State chapter support, development and communication; and (4) organization and support for national conferences.

Numerous objectives and action steps were initiated or reaffirmed as they relate to the various essential function areas, as well as other areas of service such as fund raising, administration, and networking. One area of intense concentration and immediate effort will be that of "communications," particularly the literature and other materials produced by the association. An effort is underway to evaluate, update and edit all currently available brochures, articles and other pieces of information produced by or available through the organization. This is no small undertaking and it will incur significant costs in time and resources. Objectives and action steps are being further developed in the areas of public and governmental relations, and we are moving toward seeking to establish a National PWS Awareness Day. We also want to enhance and continue to collect data and serve as a clearinghouse for residential living alternatives.

hospital, and licensed physician in the country.

Many of our objectives we would like to list as "short term" due to our desire to see them implemented very soon, but due to limitations in time, manpower and financial resources, some objectives must be labeled "long term"

More information regarding our business plan, goals, objectives, and action steps will be presented and from time to time in future issues of *The Gathered View*.

The Board very much appreciates your support and encouragement, and we are profoundly aware of our responsibilities. But because we are a volunteer organization, the efforts of each individual member are needed, particularly at the state and local levels. Though we wish to see strength in the National Office, we also want to encourage the growth and strengthening of state and regional PWS chapters, associations, and support groups. We are a "grass roots" organization; therefore, every member is part of the "root" system and is vitally important. We must each do our part to help produce "grass," i.e. promote awareness of PWS.

Let's pour on the fertilizer.

Tending the Marriage Garden

by Janalee Tomaseski-Heinemann

Recently, Al and I have been watching the marriage of some close friends self-destruct. They were both married before and brought children into this relationship, so they blame it on the kids. They state that most conflicts arise over disagreements in how to raise and discipline the children. They also state they have no time to work on their relationship because all of their time and energy goes to driving the kids to events, attending activities, and getting to appointments. This scenario sounds similar to that of many parents of children with Prader-Willi syndrome.

Eleven years ago, when we began our relationship with five children between us — including 8-year-old Matt, who had PWS — the question Al and I had to ponder was, "Is there a way to keep a marriage all you want it to be in spite of children, time constraints, and PWS?" In some ways, we went into the relationship blindly optimistic, like most people in love. On the other hand, what we had going for us was an equal commitment to work on the relationship and to put it as a priority — above all. We realized in the long run that it would be better for our family as a whole if we did not put our relationship on hold to meet our children's every need. We found the best way to keep ourselves nurtured, happy, and sane was to have a supportive relationship. It helps to go hand in hand into the waves of frustrations, worries, and crises of raising children.

We have stopped to reflect on how it is that we have managed to come through more years, more children, and more real life crises than our friends and yet feel so good about our relationship. The following are guidelines we have established for ourselves that we feel have made the difference.



Make your time together a priority over the kids, over other people, over recreation, over work.



Be slow to criticize, quick to compliment. Don't criticize the other in public.



When one works, the other works. When you play, play together.



Keep talking about the little things. Don't go to bed without resolving differences.



Be creative in finding fun time and romantic time together.



Touch a lot. Sleep with lots of body contact. Schedule love making at least once a week, whether you feel you need it or not. (Yes, even if you're too tired, the kids are beating on the door, or you don't feel the least bit sexy.)



Never, ever comment on or make fun of your spouse's weight or any characteristic they are self-conscious about. Learn to laugh at yourself — not at each other.



Be cautious of relationships with the opposite sex. Be aware of the signals of attraction you might be giving off, and don't deny to yourself when an outside relationship is becoming detrimental to your marriage.



Discuss in private, and present a unified front on, issues of discipline and privileges with children. In a second marriage, be extra cautious in disciplining each other's children, as your spouse will be sensitive about criticism of their biological child. It is hard to weigh whether you are being too strict or too lenient with any child, but this is especially true of a child with disabilities.

Having a child with Prader-Willi syndrome does not have to mean that your relationship will automatically deteriorate. On the other hand, only a conscious and daily effort will keep your relationship whole and alive. Like a beautiful garden, it takes daily tending. (Unlike a beautiful garden, you can't hire someone else to maintain it for you.) Our grown children now tell us that it was by our example that they learned to "tend their gardens." A favorite quote of ours from an unknown author states:

"The most important thing a father can do for his children is to love their mother."

by Tere Schaefer

Public Relations

● **Impact magazine**, published by St. Coletta's of Massachusetts, Inc., ran a lengthy article in its Winter 1993 issue on Prader-Willi syndrome and on its specialized program for two students with PWS at the Cardinal Cushing School and Training Center. Included in the article was a discussion of PWS based on an interview I gave, but which incorrectly states that only one researcher is doing the majority of work on the syndrome. In fact, research on PWS is being conducted throughout the country and is an area of great interest at present. Many of these researchers are also members of our organization, which indicates that their concerns go far beyond the research lab. Their dedication and convictions are a source of hope and encouragement to us all.

● **Information packets** continue to roll out of the National Office, sometimes up to 10 a day, in response to first-time inquiries about PWS and/or the association. Interestingly enough, since we've moved the office, we have gotten more walk-in visitors for information than we ever had in the five years at our previous location. Now we need people to walk in and say, "I'd like to volunteer my time. Could you use my help?"

Research

● **Scientific journal articles** inform us of the recent progress being made in understanding the syndrome. Research reports on the use of synthetic growth hormone in the treatment of PWS have been published in the *Journal of Pediatric Endocrinology* by Dr. Moris Angulo and colleagues at Winthrop-University Hospital in New York (Vol 4:3, pp. 167-173) and by Dr. Phillip Lee of Baylor University in Texas (Vol 2:1, pp. 31-4). Studies of IQ and behavior issues by Dr. Elisabeth Dykens and colleagues at Yale are reported in the *Journal of the American Academy of Child and Adolescent Psychiatry* (Vol 31:6, Nov. 1992, pp. 1131-1136). (See review of Dr. Dykens' work on page 6 of this *Gathered View* issue.) And Dr. Vanja Holm and colleagues at the University of Washington report the diagnostic criteria for PWS in the recent issue of *Pediatrics* (Vol. 91:2).

Our deepest sympathy to the Spates family of Maryland.

Melinda, their 27-year-old daughter with PWS, passed away on March 17. Melinda wrote a beautiful monologue, which she was to present to her class on March 23rd. Unfortunately, it is too long to publish in *The Gathered View*, but we hope to share it with PWSA members in a future publication.

Conference 1993

Hotel Reservations

First, a note on the hotel arrangements: We've discovered that some of you did not receive a hotel reservation card in your pre-conference packets; our apologies for the error. For those of you who did not receive the card, the special conference rate at the Registry Resort Hotel is \$55 per night, and reservations can be made by telephoning 800-247-9810. (Although some members have reported difficulties trying to make reservations through the hotel's 800 number, we've since talked the hotel management and believe the problem is now cleared up.) **Please make sure when you call the hotel's 800 line that you tell them you are with the PWSA conference.** If you'd prefer to make your reservations by mail, call the National Office, and we'll send you a hotel reservation card.

Site Visit - 'This will be the nicest hotel we have ever stayed at'

When I made a site visit to the hotel last month, I knew this would be the best place we have ever stayed for a conference. Maybe my judgment got clouded by going from 10 degrees Fahrenheit (and my down coat) to 85 (wearing shorts and getting sunburned). However, I really have to say there is lots of room to sit, relax, and congregate around a beautiful, huge pool. In addition, the hotel offers two smaller pools, 24 tennis courts, a health club, and even special rates on a professional massage. The Registry Resort is located along beautiful Scottsdale Road, which has many resorts, several restaurants, and ends up at the

Scottsdale Fashion Mall. The power shoppers among you will be delighted to see Saks Fifth Avenue, Nieman-Marcus, and plenty of hometown flavor as well. And, of course, there's a golf course for those of you who want to take in a sunrise round before the meetings start!

Southwestern Banquet This Year

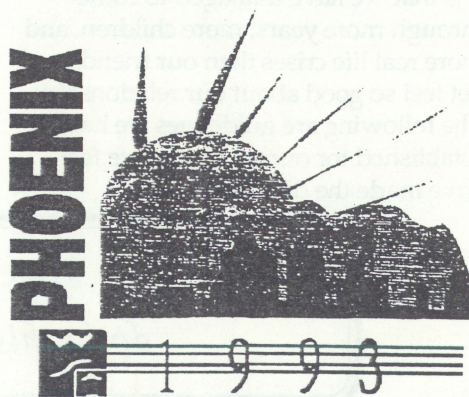
The traditional banquet will take on a new look: Southwestern. So don't bring those formals; bring your boots, checked shirts, and denim skirts. Your payments and PWSA (USA) can cover the basics for this southwestern meal, but you could help make it extra special.

Are you, your chapter, your employer, your local VFW, Kiwanis Club, Knights of Columbus, or maybe just a bunch of your friends, willing to sponsor an extra treat for the banquet?

Possible extras are:

- \$200 for a country band;
- \$200 for Guy, a storyteller, and his ox, Fuzz;
- \$250 for a square dance caller and instructors;
- \$300 for Indian dancers;
- \$350 for authentic western hats for the Youth/Adult Activity Program participants;
- \$500 for sheriff's badges for the men and garters for the women; or
- \$500 for a commemorative bandanna for everyone.

Sure, the banquet would be fine without these things. For many, however, these special touches would make the evening under the stars much more memorable. Think -- for many participants, this will be the *only* PWSA conference they ever attend.



Conference Grants Available

Each year, PWSA offers grants to those who wish to attend the national conference, but whose finances prohibit this type of expense.

How do you apply for a conference grant?

Send PWSA (USA) a letter with the following information:

1. the size of your family and age of your child(ren);
2. an indication of your income and expenses;
3. a brief summary of what the greatest difficulties you, as a family, are experiencing in dealing with PWS;
4. whether your family would need all your conference expenses paid or if a partial grant would be sufficient (e.g., for travel or lodging only); and
5. whether anyone in your family has ever attended a PWSA national conference.

What is the process?

Your letter will be reviewed by the executive director. Traditionally, there are two grants available to cover up to \$2,000 in expenses. However, based on the number of requests and the amount of money requested, the executive director may then apply to the CIT [Crisis Intervention and Training] committee for additional funding. In 1992, five families were able to attend the conference through grant funding. PWSA members contributed \$300 toward these grants with their registration fees.

Request Deadline

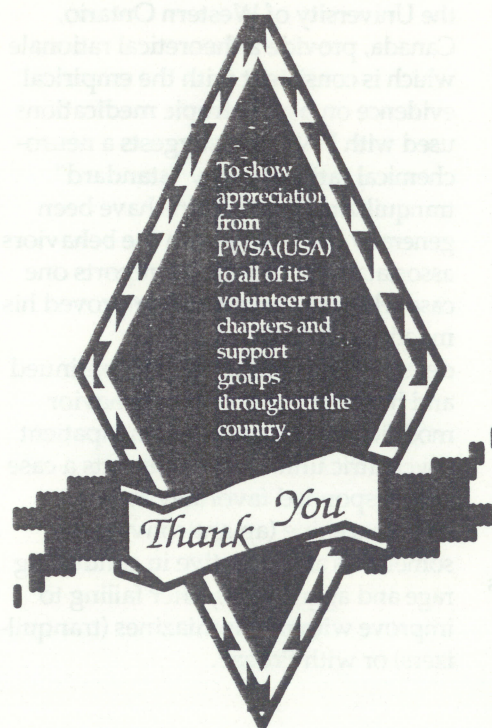
Letters requesting conference grants must be received at the National Office by **Friday, May 14, 1993**. Applications received after this date will not be considered for funding.

Chapter Notes

Bright Idea: North Carolina has established a Lending Library, which includes PWSA(USA) publications (articles, audio and video tapes, books, conference proceedings, newsletters); PWSANC brochures, articles and newsletters; and miscellaneous articles and brochures relating to PWS. For information, contact Doris Oakley, President, (919)599-3120.

Good News: Chapter efforts do pay off! Both New York and Missouri report opening of group homes within the past six months. Congratulations! North Carolina also reports real progress toward this goal.

Best wishes: Although the Prader-Willi California Foundation is not an official chapter and is structured differently from a chapter, PWSA(USA) would be remiss not to recognize Bill and Shirley Dingley. Bill and Shirley are retiring from many years of devotion and work as publishers/editors of the PWCF News, support group leaders, Board members and lobbyists in the state of California.



Summer PW Program at the Pittsburgh Rehabilitation Institute

Bea Maier, Ph.D., Program Coordinator, announces that The Rehabilitation Institute of Pittsburgh is planning two short special summer treatment programs for children and adolescents with PWS:

"Our programs will focus on improving their muscle tone and cardiopulmonary fitness, achieving weight loss, and increasing their understanding of their special dietary and life style needs, as well as enhancing their social skills and self-esteem.

The programs provide a high level of individual attention from a multidisciplinary team, consisting of a physician, unit team leader, psychologist, dietician, exercise physiologist, speech language pathologist, occupational therapist, social worker, computer literacy specialist, counselors and coordination staff. We have daily use of a swimming pool and recreational facilities. We want to make the experience away from home fun and comfortable as well as insuring that the goals are achieved. Parent guidance and training is an important aspect.

Sponsorship for the program is usually obtained through health insurance or Medicare and Medicaid. We will work with parents and referring physicians to facilitate obtaining sponsorship. Our program dates for 1993 are as follows:

June 14 - July 9, ages 6 - 13;
July 19 - August 20, age 14 and up.

We will continue to admit children, adolescents and adults in crisis at any time."

For more information, contact Bea at The Rehabilitation Institute, 6301 Northumberland St., Pittsburgh PA 15217, (412)521-9000.

Review of Recent Psychological Research Reports

by Stephen Sulzbacher, Ph.D., University of Washington, PWSA Scientific Advisory Board

Two reports on psychological development and one on psychopharmacology in Prader-Willi syndrome appeared in the November 1992 issue of the Journal of the American Academy of Child and Adolescent Psychiatry. These articles all have implications for current treatment of persons with PWS.

Psychological Development

The two articles on psychological development are by the same authors and report data on the same 21 cases, ages 13 to 46, in the northeastern U.S. One study reports IQ data, and the other examines how well these older adolescents and adults function in daily living and socialization.

Dr. Elisabeth Dykens and her Yale University colleagues provide a much-needed extension of the very early work done by myself and Dr. Vanja Holm more than 10 years ago at the University of Washington. At that time, we suggested that, in children under 8 years old, IQ seemed higher in those whose weight was well controlled, relative to children who were obese. In Dykens' study, weight and degree of intellectual impairment were not related. Taken together, these results suggest that IQ and weight are not related directly, but that psychosocial variables, like consistent parenting and behavior management, probably accounted for *both* the good weight control and the improved cognitive performance in the earlier study and may continue to account for differences in psychosocial adjustment in adulthood.

Dykens' results are encouraging because they imply little intellectual deterioration into adulthood and also suggest specific educational approaches uniquely suited to persons with PWS. A further implication of Dykens' research is that persons with PWS can certainly continue to profit from educational community college courses as constructive adult leisure time activities (just like the rest of us). *This article should certainly be brought to the attention of teachers looking to adapt curriculum to the person with PWS.*

Dykens' report on adaptive and maladaptive behavior of older persons with PWS is less definitive and also less encouraging. She found that, while her sample continued to exhibit relative strength in daily living skills, their socialization and temperament continued to be problems into adulthood. Weakness is particularly noted in judgment and coping strategies. Temper tantrums and disobedience continued to be problems in late adolescence and throughout the adult years. Tiredness, lethargy, and liking to be left alone were behaviors reported to increase in persons with PWS in their 20s and 30s.

Psychopharmacology

In the third paper, "Psychopharmacogenetic aspects of PWS," results of two uncontrolled case studies are used to illustrate an innovative way to think about medications for affective and behavioral disorders associated with PWS. Dr. Tu and his colleagues at the University of Western Ontario, Canada, provide a theoretical rationale which is consistent with the empirical evidence on psychotropic medications used with PWS. He suggests a neurochemical rationale why "standard" tranquilizing medications have been generally ineffective with the behaviors associated with PWS. Tu reports one case who lost weight and improved his mood and behavior when dextroamphetamine was discontinued and he was treated with a behavior modification program on an inpatient psychiatric unit. Tu also reports a case who responded favorably to carbamazepine (an anticonvulsant sometimes also effective in controlling rage and aggression), after failing to improve with phenothiazines (tranquilizers) or with Prozac.

An earlier article, entitled "Use of psychotropic medications in persons with Prader-Willi Syndrome" summarizes results of a survey on usage of these medications conducted by Drs. Barbara Whitman and Louise Greenswag. This study, partly funded by the PWSA, was reported in Dr. Suzanne Cassidy's recently published book of abstracts from the 1991 International PWS Conference (*Prader-Willi Syndrome and Other Chromosome 15Q Deletion Disorders*, 1992, Springer-Verlag). In this study, 37 percent of the 111 persons surveyed had been tried on a psychotropic medication, and 32 percent (n=36) were taking this medication at the time of the survey. This chapter doesn't list which medications were being used, nor what behavior might have been affected. It is also unclear whether a psychiatric diagnosis had been made, in addition to Prader-Willi syndrome. Nevertheless, if indeed one-third of adults with PWS are taking one or another medication for behavior, one must agree with the conclusion of Whitman and Greenswag that more carefully controlled studies should be done, and that a theoretical framework like the one Dr. Tu introduces would be a useful way to structure such studies.

Conclusions

These four articles represent progress in our understanding of the behavioral side of PWS. One could conclude from these studies that there are steps that can be taken, which may include adult education or a trial of a psychotropic medication, to help persons with Prader-Willi syndrome control their moods better and achieve a more satisfying life for themselves into adulthood.

Ask the Professionals

Responses to the following two questions were provided by Suzanne Cassidy, M.D., Associate Professor of Pediatrics at The University of Arizona and member of PWSA's Scientific Advisory Board and Board of Directors.

Q: *My four-year-old daughter has never liked to drink water and rarely expresses thirst. Another parent told me that this is common in PWS. How common is it? What causes it? How much water does she need in a day? Will inadequate water intake result in any serious medical or dental problems?*

A: It is very common for people with PWS of all ages to shun drinking of water, even though they may drink the normal amount of fluid for the day by drinking other things. Like many of the abnormalities of PWS, control of thirst appears to be due to the functioning of the hypothalamus, which is altered in PWS. People and animals who have damage to the hypothalamus for other reasons also drink very little water, which confirms that this is part of the deficiency of this part of the brain in PWS.

No one has done an objective study of how many people with PWS don't like to drink water, so we don't know how common it is. Many of the patients I follow refuse to drink water.

Obviously, water is essential to life. The amount of water needed on a daily basis depends upon the size of the individual, the amount of activity they do, and the temperature of the air around them. The fluid requirement can be calculated for each individual when these factors are known. For most adults, approximately 50 cc/kg/day [cubic centimeters per kilogram of weight per day] is usually used; young children often need 100 cc/kg/day.

If one doesn't drink any fluids, dehydration occurs and can cause serious medical problems, especially kidney problems. However, the fluid can be taken in as any liquid, not only water. Coffee, tea, soda, juices, etc., all contain a lot of water. Also, foods contain water, especially fruits. I have never known of a patient with PWS who became dehydrated or had any consequences of decreased water intake unless they had fever, uncontrolled diabetes, diarrhea, or other excess fluid loss.

Q: *I've heard that PWS occurs one in 15,000 births and that 90 percent of the cases go undiagnosed. How is the incidence rate determined? If it's true that only 10 percent of the cases have been identified, is it possible that many of the undiagnosed cases are less severe or differ in some other way from those that are diagnosed?*

A: A recent study based on ascertaining diagnosed patients with PWS in the state of North Dakota determined that the prevalence of PWS was approximately 1/15,000, based on the number of individuals who carry the diagnosis and the population of the state. A similar figure was reported at the International PWS Conference in 1991 by individuals from Norway, where an epidemiological study was done. Several other countries also reported similar figures.

Unfortunately, no one has done a study based on following large numbers of consecutive newborns who were tested for PWS, and, therefore, a precise figure is unknown. Such a study would be extremely expensive, tedious and difficult to do, and is unlikely to ever be done.

I am told that a group in Sweden feels that the incidence of PWS is more like 1/8,000, but this is based on estimating the number of diagnosed patients and the likelihood of diagnosis, and is less accurate.

I am unsure of where the "90 percent undiagnosed" figure comes from. I have never read any studies documenting that, and the incidence of 1/15,000 was based on diagnosed patients, and, therefore, does not take into account those who have not been diagnosed. Many workers in the field believe that there are a number of people who are affected and do not carry the diagnosis, however. This is based on people who get diagnosed at all ages (in my experience, up to 40 years of age) for the first time. Probably, there are more older undiagnosed individuals than

younger ones, since most patients get evaluated as children and the condition has only been described for about 35 years. However, there is no way to know whether there are individuals who are mildly affected and, therefore, not diagnosed. I was part of a study attempting to determine the clinical spectrum (variability) of PWS by doing molecular genetic studies (for the deletion or maternal disomy seen in PWS) on patients who lacked some of the features of PWS. While all typical patients have one of these molecular findings, those 15 patients studied who were not typical — because they lacked some of the features, or had somewhat different features — did not have either of the molecular findings. This suggests that the clinical spectrum may not be significantly broader than we now believe.

Parents:

We are still looking for responses to last issue's "Ask the Parents" question:

What do you do when your child with PWS has destroyed property — either their own or someone else's — in a tantrum?

If you have any experience or wisdom to offer on this subject, please drop us a line at the National PWSA Office or call the 800 number. Also, let us know what questions are on your mind regarding your son or daughter with PWS. Sharing both your questions and your experiences through *The Gathered View* can benefit all of our members, families and professionals alike.

Professionals:

We invite your participation in this column and throughout *The Gathered View*. We would be happy to receive for publication your additional responses to questions in this column, your suggestions for new question-and-answer topics, or your suggestions for feature articles for the newsletter. Both other professionals and families could benefit from reading about your experience or research.

From the Home Front

From a distant "home front" in Victoria, Australia., Mrs. N.B. McNamara shared many insights of the things she has learned in her 68 years, 30 of which have been spent caring for her son Jan Erick. Here is a condensed version of her letter:

In '92 I was forced to put my son into a CRU (community residential unit) because I had a hip replacement, and then the flu for six weeks, and then I returned home to find it had taken more out of my system than I realized. My doctors thought something would 'give way' and it sure did in the form of duodenal ulcers, which I knew had been present for years. Jan Erick kept having his temper tantrums, which upset the ulcers a great deal. I was forced to tell my doctor the truth about the ulcers, and he hit the nail on the head when he said to me "You live for that boy, don't you?" After giving my life to him for 30 years I suppose I need a bit of a break, but I know as his guardian I still have a say in his care. As much as I don't like it, at least he is getting looked after, and not everything he wants to do. By the way, I have him on a hypoglycaemic food intake. I find that both he and I dislike the word diet, as it gives him a false sense of being different - yet food is a diet.

I most certainly have lived my life for him all his life, because his father was killed prior to my knowing I was pregnant. I have been his Mum, Dad, confidant, and friend or whatever all in one. In hindsight I see just where and how that was a big mistake on my part, but one doesn't always think of that while doing it. It is only when I stood back, in my case while in the hospital, that I realized this. In looking back, one of the most vivid memories is an incident in '90 when my son suffered many attacks of hypothermia, and on one occasion his heart rate was 32 per minute with a temperature of 27.5 degrees Celsius (about 50 degrees Fahrenheit). Fortunately he was saved by the BAIR HUGGER. — an American invention which is like a great big hair dryer which pumps warm air all over the patient. It is made by Augustine Medical Inc., Eden Prairie MIN (800)733-7775. I have been pleading with our hospital to purchase one permanently, but here at the moment money is very tight, believe me, the cuts going on galore for our loved ones. The stupid administrators are cutting the 'hands-on' staff in lieu of the top-heavy administrative staff . . . about which I have been writing many letters and going hither and yon. But one can only work within the means they are given, and thankfully I do manage.

Hope I haven't "bashed" your eyes too much with this mail. I'm endeavoring to help — even if it is only regarding the BAIR HUGGER — just in case you know some soul over there, with your winters, it may possibly assist.

Kind regards,

(Mrs) N.B. McNamara

Victoria, Australia

Many Thanks to Our Contributors

Heavenly Angels (\$250-\$999):

Schmidt, Whitehead, Kane

Arch Angels (\$100 - \$249):

Patterson, Notbohm, Ripley,
Eisen, Our Lady of the Lakes,
Toby, Kaugher, Schertz, Judge

Angels (\$50 - \$99):

Abramek, Dorsey, Menzer-Wolf,
Krautwurst, Schneider, Mitchell,
Schertz

Cherubs (\$1 - \$49):

Kitzman-Kelley, Lorenz,
Dibattista, Strongitharm, Bauer,
Hannuksela, Cohen, Posch,
Vermeulen, Schneider, Powell,
Bacigalupo, Boyle, Stutts,
Parent, Chase

Conference Grants:

Kellerman, Westinghouse Electric, Corp.

Louise Greenswag's B-Day:

Chausow

Patron Dues Members (\$100+):

Greenswag, Goranson,
Williams, Metzger, Miller, Trepanier

Contributing Dues (\$40-99):

Burton, Rose, Marik, Sidbury,
Morris, Hamill, Welch

CIT: Olson

Research: Boyd

Contributions through United Way:

Grand Rapids UW - Barney
Monroe County UW - Vermeulen
Hamilton Life - Hilgenberg
KPMG Peat Marwick Co. - Horowitz,
Mahoney, Nussbaum, Ralston

Memorials:

(In Memory of - Contributor)

Wilma Pearson - Maurer

Kathy Robbins- PWA KY Chapter

Milton Singer - Levine, Gail, Clement,
Geiger, Sabatos,

Corrine Rachel & Clarence Erdahl &
Grace Ebena - Van Zomeren

Adam Hanson- Duddy, Haley, Bartlett
Learning Center, Cocheran

Melvin Granger - Quinn

P. Richardson - Schneider

Carmen Ciancio-Sheeran

Mitchell - Margaret Hood

Margaret "Peggy" Ashbach -
Kolle; Barron; Swanson; Carlson;
Crestfield Morgans; Hockin; Swanson;
Magnuson; Scherer; Northwest Lasers,
Inc.; Campbell; Mid-States Morgan
Horses; Koenig; Laser Control, Inc.;
Young; Off The Road Tire Service, Inc.;
Ziegler Inc.; Commerical Asphalt, Inc.;
Asphalt Recycling and Reclaiming Assoc.

by Melanie Grace, B.S., C.R.C., C.W.A.
and Anna Marie Saporito, M.Ed., C.R.C.,
C.V.E., The Rehabilitation Institute,
Pittsburgh, Pa.

Welcome to the wonderful world of work! Or at least it *can* be. However, adults with Prader-Willi syndrome and their families often find that this world of work isn't always so wonderful. In an attempt to address some common work-related concerns, this is the first in a series of articles that will offer practical information, suggestions, and helpful hints to make the vocational process as successful as possible.

The National Prader-Willi Syndrome Association estimates that approximately 70 percent of adults with the syndrome participate in sheltered work, 15 percent are in therapeutic activity programs, and 13 percent are employed in the community. Before we offer practical suggestions for improving work situations, however, it is important for adults with Prader-Willi syndrome and their families to know about the various levels of employment, how they differ, and the type of "client," or worker, appropriate for each.

In general, it's important to know that rehabilitation professionals define the levels of employment according to an individual's level of functioning and the support systems necessary to ensure successful programming. Another important point to understand is the funding source. Funding sources are necessary for all levels of work except non-supported, competitive employment. Some examples of the sources of funds are the office of mental health/mental retardation and the state office of vocational rehabilitation.

Following are descriptions of the many forms of employment, beginning with the most structured and ending with the least structured:

● **Therapeutic Activity Centers (TAC)** emphasize basic self-care skills, socialization, and interpersonal skills. Basic pre-vocational skills, such as simple sorting, simple assembly, and collating are used in a limited fashion in this highly structured and supervised setting. Group size is generally 5-6 clients with one staff person.

● **A Work Activities Center (WAC)** uses work tasks to focus on adaptive behavior and basic skill development. Clients in a WAC may work half or full days. They may do work tasks similar to those found in a TAC, but the work expectations are somewhat higher. Money or tokens may be used as reinforcement. The group size is larger than a TAC, but smaller than those in sheltered work programs.

● **Sheltered Work Programs** focus on adaptive behavior and productivity. The work tasks originate from job contracts with outside companies. These jobs can vary in the number of steps and skills required to complete them. Sorting, assembling, and manufacturing tasks are typical assignments used both to train employees and to provide extended employment. Clients are paid piece rate or a pro-rated salary, depending on the nature of the contract with the outside company. Salaries will vary according to the productivity of the individual. The client-to-staff ratio ranges from 15:1 to 20:1.

● **Transitional Work** is usually the next level in the progression. These programs also emphasize adaptive behavior but focus, in addition, on job readiness skills that are needed for working in the community. Specific job training and job trials are used at this level. This work is actually performed out in the community, but with a high level of structure and supervision provided. "Enclaves" and "mobile work crews" are terms used for transitional types of employment.

An enclave is a group of workers with a supervisor from a sheltered work program who work in a designated area within an outside company. They generally work in small groups, (6 to 15 workers per supervisor) completing basic clerical, sorting, assembly, and packaging types of tasks. The supervisor from the sheltered work program manages or makes accommodations to ensure quality and work completion.

Mobile work crews are groups of clients that may go to several businesses to perform a specific job duty, e.g., janitorial services. A work supervisor from the sheltered work program trains a small group of employees and monitors

their work performance as they move from one site to another. Transportation generally is provided by the work program.

● **Competitive employment** is work in the community that is paid for directly by the employer. Entry level, semi-skilled, skilled, technical, and professional are job classifications under the umbrella of competitive employment. Within this level of employment, "supported work programs" are available for individuals who otherwise would be unable to work competitively because of difficulties in adjusting to a work setting, or who need additional training time but have exceeded the production demands of sheltered/transitional employment.

Supported work programs allow individuals to develop good work behaviors and job skills in a competitive setting. This is accomplished by providing extended on-the-job training and work accommodations. Ongoing support by a job coach is available for a few months on a full-time basis during the training phase. As the employee learns the job, the involvement of the job coach fades from weekly to monthly visits, or, in some cases, even less frequent intervals. However, if the need for additional support recurs, the job coach can become more actively involved again.

As one can see, a variety of employment options can be available in the community to individuals with Prader-Willi syndrome. However, availability of vocational programs and the specific program guidelines will vary from community to community. The current trend in rehabilitation is to provide the least restrictive vocational option and offer individuals the best opportunity to work in the community.

The next article of this series, will cover the vocational evaluation process needed to determine the appropriate placement.

The authors invite and would appreciate feedback and questions concerning any vocational issues for adults with PWS. They can be reached at The Rehabilitation Institute, 6301 North Umlerland St., Pittsburgh, PA 15217 or telephone (412) 521-9000.

Alternatives to Buying Out the Toy Store

Toys are an important aspect of play and learning for young children with disabilities, but they can also be a source of great frustration if a child's muscles can't meet the strength and coordination challenges of an interesting plaything. It's often hard to predict which toys will make a hit with the child and be physically manageable as well. Rather than spend a small fortune in the search, parents may want to explore borrowing toys, either instead of, or before, buying them. Here are some possible ways to do that:

* A Lekotek Center — If you're fortunate enough to live near one of the 52 Lekoteks in the U.S. (24 states have them), take your child for a visit. Originating in Sweden, the centers provide individual consultation in selecting and adapting toys for children with special needs to develop their sensory and motor skills. For a small membership fee, families can visit for monthly play sessions, work with early childhood and family support specialists to determine their child's particular play needs, and borrow several toys until the next session. For more information, contact the National Lekotek Center, 2100 Ridge Ave., Evanston, Ill. 60201, telephone: 1-800-366-7529 or 1-708-328-0001.

* A toy-lending library — Some communities operate nonprofit toy loan programs open to any resident. Check to see if your community has one. Or write to the USA Toy Library Association, 2530 Crawford Ave., Suite 111, Evanston, Ill. 60201.

* Your child's education program or therapist — Someone who's already working with your child in the area of motor skills may be able to let you borrow a new toy for a short period to see if it's something worth purchasing for home. It doesn't hurt to ask.

* A local parent group — If nothing else is available, try starting your own toy-lending program with a group of friends, your child's playgroup, or groups of parents from school or church who have children of similar ages.



Family Network

A family with a lower-functioning child with PWS (age 9) has asked to be put in touch with other families who have lower-functioning children with the syndrome. If you would like to share experiences with this family, please call the National Office at 800-926-4797.

Beads for Profits

Ms. Billye Schmidt makes crafts and donates her proceeds to the Missouri chapter of PWSA(USA). She has used up her supply of beads, crystals, pins, etc., and is asking for help. If anyone has broken strands of beads, single earrings, broken pins, old or unwanted jewelry of any kind. Please send them to Billye Schmidt, 9432 Tiber Dr., Afton MO 63123. She'll recycle them to benefit PWS!

Pre-Registration Agenda Packets

Any overseas or Canadian members wishing to obtain registration information for this year's PWSA (USA) conference, send a fax (612)641-1952, or a note with your request to the National Office.

Support is Needed for Conference Grants

When you register for this year's conference, don't forget the line to add a few bucks for conference grants. These grants enable several additional families to attend the conference, who otherwise would never be able to have this experience. Please give generously !!!!!

Material Review Volunteers

The PWSA Publications could use three to four volunteer parents (of any aged child) to review the materials currently published by PWSA(USA) please let the National Office know.

Our Own PW Poster Boy

by Lota Mitchell, M.S.W.

The ranks of Prader-Willi syndrome can now boast a "poster child"! Three-year-old Stephen Immekus, son of Paul and Sandy Immekus, has his picture on numerous billboards throughout the city of Pittsburgh, Pa., as well as in direct-mail appeals and TV spots.

As part of the 1992-93 fundraising campaign for St. Peter's Child Development Centers, Stephen and three other children from Centers classrooms were selected to pose with Pittsburgh Pirate Don Slaught, who co-chaired the campaign.



Stephen, Don, and Stephen's brother, Mark; At Home Plate, Three Rivers Stadium

St. Peter's is a community agency operating in four areas of the city to provide free developmental services for infants, toddlers, and preschoolers who are mentally retarded and/or developmentally delayed, plus counseling and education for their families.

Stephen's Story

Born November 15, 1989, Stephen began life in typical PWS fashion. He didn't cry, was hypotonic, wouldn't wake up, and couldn't eat. At 8 days old he was back in the hospital, dehydrated. Much less typical is that when Stephen was *only 10 days old*, geneticist Angela Linn, M.D., made the diagnosis of Prader-Willi syndrome on the basis of chromosome tests.

A feeding specialist helped Sandy and Paul learn how to feed their baby, and at 13 days old Stephen was back home again. At six weeks he started physical therapy, and at 3

months he became a "student" at St. Peter's for infant stimulation one hour a week. Today he attends St. Peter's five days a week, four hours a day.

Stephen walked at 2 years 7 months and began talking at age 3. Until then he used sign language exclusively to communicate; now he combines speech and signs, with a preference for speech unless he has trouble being understood. He says two- or three-word phrases such as "Don't want to" or "Steve eat." Sandy says that, although he talks about eating, Stephen is not a big eater and does not search for food. He has very definite food preferences: pizza and spaghetti top his list, while lunchmeat rates a flat refusal.

Stephen likes having books read to him and listening to children's tapes, but his favorite activity is playing with shoestrings! Sandy, now the president of the Prader-Willi Western Pennsylvania Association, was very surprised to find that another little boy from the group also loves shoestrings. The two mothers began wondering if this was yet another PW characteristic!

Family Reactions

Big brothers Raymond, 7, and Mark, 5, treat Stephen just like any little brother – wrestling, chasing, pushing him away. The older boys know he has weak muscles, but Sandy and Paul haven't felt the need to try to explain PWS to them yet.

Their pediatrician (whom Sandy says they educated about the syndrome by using PWSA brochures) recommended they consult a registered dietician shortly after Stephen's first birthday because he was not gaining enough weight. The dietician provided them with weight sheets, recipes, serving size recommendations, and nutritional guidance that led them to change the whole family's eating habits.

How has having a child with PWS affected Paul and Sandy? At first, they were totally shocked, and both cried. Next, denial came – and went. Then they wanted to find out everything they could about the syndrome and what they could do. When Stephen was just a few weeks old, his family visited mine, because we live in a nearby township and have a 19-year-old daughter with PWS. Fortunately, meeting our Julie "to see their future" was an encouraging experience for Sandy and Paul after hearing some bleak predictions from doctors at the time Stephen was diagnosed.

Sandy relates that the first six months were the worst for her. She was disappointed at the baby's lack of response. She says she thought of him as a "useless baby, such a waste." Paul's attitude, after his initial grief, was to be positive and practical: "He's going to be OK. We won't let him put on the weight. We'll make him the best we can. Together we can handle this."

By Stephen's first birthday, Sandy says, they "had come to terms with the diagnosis," and today they truly enjoy him as he is. Sandy says that her little poster boy makes her laugh every day and that, although it may sound strange, "Everyone should have a special child, because it makes you really think about what's important."

Lota is the former chair of PWSA's Board of Directors, a member of PWSA's Publications and Education Committee, and an active member of the Prader-Willi Western Pennsylvania Assoc.

Calendar of Events

If you have a date and event you want posted in the Calendar of Events, please let the National Office know by May 14th to be included in the next issue.

April 30 -May 1, 1993: New York-New England Conference, Albany NY.
contact Shelly Tarakan at (516)621-2445.

May 1, 1993: PWSA of Wisconsin, contact Barb Dorn (608)838-9535.

May 22, 1993: PW Florida Assoc., contact Sandy Stone (904)423-9378.
Meeting will be at the Ramada Resort in Kissimmee.

June 5, 1993: Prader-Willi Northwest Assoc., contact Susan and Steve Lundt (206)285-7679.

July 14, 1993: National Conference Care Providers Day, Scottsdale, AZ.

July 14, 1993: National Chapter President's Day, Scottsdale, AZ

July 14, 1993: Eighth Annual Scientific Conference, Scottsdale, AZ

July 15 - 17, 1993: 15th Annual PWSA(USA) Conference, Scottsdale, AZ

June 30 - July 3, 1994: Second International PWS Conference, Cambridge, England

The Gathered View is the official newsletter of the Prader-Willi Syndrome Association(USA) and is sent to all members. The opinions expressed in *The Gathered View* represent those of the authors of the articles published, and do not necessarily reflect the opinion or position of the officers and Board of Directors of PWSA(USA). Duplication of this newsletter for publication is prohibited. Quotations may be used upon credit given to PWSA(USA). **The Association gratefully acknowledges its volunteer editors, Lota Mitchell and Linda Keder** If you are interested in a membership dues are \$21 for an individual, \$26 per family, \$31 per agency/professional. Send dues, change of address, or letters to: 1821 University Ave. W. #N356, St. Paul, MN 55104. Questions or comments regarding this publication or PWSA(USA) call: 800-926-4797 or 612-641-1955 or Fax 612-641-1952.

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