

# The Gathered View

*Newsletter of the Prader-Willi Syndrome Association*

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## *Prader-Willi Syndrome Association Conference "now history"*

As Dr. Beltran writes in his "President's Message," (see page two) the 11th Annual conference is "now history." We are happy to report that attendance followed fairly close to our usual attendance with 248 registered adult attendees and 101 in our activity program. Comments made and evaluation sheets shared gives us the impression that efforts were worthwhile and we held another successful conference. We have asked a "first time attendee" to write his impressions of the meeting, which is included in this issue along with some photos that we are happy to share. The evaluations always share that the opportunity to meet and talk with other parents is the greatest part of the planning. We are all looking forward to our next conference in July, 1990 in Salt Lake City.

Door prizes were offered at the conference to assist in supplementing the conference budget. The round trip airline ticket to anywhere in the U.S. was won by Dr. P. Wilkinson of Lethbridge, Alberta, and his wife, Joyce won the 2nd prize of one of our autographed, numbered, Management books. The 3rd prize, another book, was won by Rhett Eleazer of South Carolina.

One of the board actions was accepting the bid from the Illinois chapter to host the conference in 1991. Some confusion has developed because of the plans being made to hold a meeting in the Netherlands. We would like to clarify the fact the 13th Annual Conference on PWS will be held in the Chicago area. The Netherlands meeting, which is still very much in the planning stage, will be financially sponsored by PWSA but this meeting will not take the place of our usual annual conference. Further information will be shared with the membership as plans develop. At the present time, Dr. Cassidy and committee are working with Henk Moezelaar of the Netherlands and Jean Phillips-Martinson of Sweden in the development of this meeting.

*A window on the conference . . . their smiles say it all!*





## PWSA

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## President's Message:

The Eleventh Annual PWSA Conference was held in Calgary, Alberta, Canada this year and is now history. Each year the conference demonstrates the maturing nature of the organization. The Scientific Sessions have developed into reports of fundamental research as well as the reports of observations of the Prader-Willi individual's characteristics. The main meeting provided a look into the social aspects of the lives of individuals with PW that had not previously been touched. But it was the meeting of the Chapters leaders that brought to my attention some changing aspects of organization that cause me concern.

The lifeblood of any organization and the reason for its existence is the people who want to be identified as members of the organization. It is stated clearly in the By-laws that the PWSA exists for the improvement of the lives of people with Prader-Willi syndrome. The goal of increasing awareness and educating the public and professions is being met on a grand scale. This is easily demonstrated by the fact that the new text edited by Director Louise Greenswag and her associates has gone into a second printing. It is also demonstrated by the decreasing number of state legislators that do not know of the syndrome.

The lives of many persons with Prader-Willi has been greatly improved by the development of over twenty dedicated residential facilities. There are more and more reports of earlier diagnoses of persons with the syndrome, evidence that the medical community is more aware, largely due to the efforts of the association and its membership.

Why then did our chapter presidents report a large disaffection for supporting local chapter efforts. Almost every report indicated that meetings were attended by the same few individuals that held the waning group together. That there were parents who would not make the effort to help in the efforts of the group to raise funds, or engage in projects, or to accept the responsibility of holding an office. Even more devastating is the unwillingness to participate in voting for officers and directors of the board for the local as well as for the national meeting. This is not unlike our state and national political elections where fewer and fewer people partake of the fundamental right of a democracy, to elect their own representatives.

When apathy at any level of organization is evidenced by indifference of the voting membership the organization can be threatened by various forms of destructive energy ranging from corruption of power to insensibility. An even more subtle destructive force that can develop is evidenced by loss of communication between the members and the development of factions fostered on rumors and innuendo. These are unpleasant thoughts and no more easy to write

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## Banquet Honorees

A few years ago PWSA started a tradition of honoring people during our conference banquet. Naturally, the first people thought of were those instrumental in founding the organization. Seeking information on PWS, the Deterlings journeyed to Seattle, WA. It was there, with the help of Dr. Holm, staff, and a few other parents, that PWSA was founded in 1975. We were happy in 1984 to be able to honor Gene Deterling as our first president and Shirley Neason as our first newsletter editor. (Fausta Deterling certainly deserved "honorable mention" also as a founder.)

The next year we chose Dr. Vanja Holm, who Dr. Beltran lovingly refers to as our "Mother Superior." Her establishment of the PW clinic in Seattle, working and caring all these many years, has benefitted us greatly.

In 1986 I felt it a privilege to award our thank you plaque to a dear friend, Dorothy Thompson. Dorothy's story is that two people with PW in her 103-bed facility took up 90% of her staff's time. It was then she decided these people needed help. Luckily she has been giving that help ever since through our organization. To the list of founders and Mother Superior, we add "Guardian Angel."

In 1987 the tables were turned on me--the person scheduled to be honored by usurped by D.J. Miller and with the help of many others surprised me with that honor. I have a beautiful memory book containing letters from our membership that is one of the most treasured gifts I have ever received. The following year we followed through with the plans and did honor Dr. Delfin Beltran. Sam, as he is better known, has been involved in our group since the first parent-professional meeting in Washington in 1976. He served as one of our first board members, as Board Chairman, and stepped into the role of President many years back. His many contributions to PWSA have contributed to our successful growth.

This year I chose to embarrassingly keep this honor "in the family." To ignore the fact that the person being honored is my husband and to place his name on this deserving people list is done because he is just that--he is deserving of this honor. Dick was the first replacement on the original Board of Directors, he served many years as Chairman and still remains on the board. In 1979 he willingly gave up a room in his home for the establishment of an office for the national headquarters and the office remained there until a year ago. He has always been my right hand when he was needed. Particularly in the past few years, he has been indispensable as our computer consultant. He has taken over the duties of running the GV labels, membership listings, and all of the many things we previously paid hundreds of dollars to have done. We also have another computer consultant, Loren Goff, Judy's husband. Without the help of Dick and Loren many of these money and hour saving innovations would not be operating.

But most of all, I believe Dick deserves this honor because he is always there to listen. A person could not remain in my job for the past ten years without a sounding board. It makes it a lot easier to work through problems when you have someone to share them with. Luckily, the rewards of this job far exceed the problems, and some of that can be contributed to the many good people who are members of the organization. So we honored Dick at the banquet and at the same time said thank you to the rest of you also.

Calgary has a special ceremony to honor people and all of the attendees enjoyed becoming "Honorary Calgarians." Dick's certificate stated, "This here paper proves Dr. Richard Wett had the purely pleasurable experience of visiting the only genuine cowtown in Canada...namely Calgary, and was duly exposed to exceptional amounts of 'heart warmin', 'hand shakin', 'tongue-loosinin', 'back-slappin', 'neighbor-lovin', western spirit...is hereby deputized as an honorary CALGARY WHITE HATTER said title means from this day on, the owner of this certificate accepts responsibility for extendin' this brand of western hospitality to all folks and critters."

-- Marge A. Wett





## *A Letter to the Prader-Willi Syndrome Association*

The illness and death of our darling daughter, Lynnel this past February has left me with an urge to notify other parents of certain signs and symptoms that they may not be any more aware of than I was. She was diagnosed with PWS at age 8. Her symptoms were primarily classic. She was on as good a diet as can be maintained with a child living at home, semi-independent and very active. She was in a hospital for a month at age 18 for a fasting diet. This worked pretty good on 500 calories a day. However, this was not encouraged at home with all her numerous physical activities. Up until her years' illness, her weight remained around 180-210. I'm sure she was 250 lbs. at the time of her death. Although her calories were watched closer then, all her physical activities were stopped (sheltered workshop, swimming, recreational activities), busy every minute of every day. Naturally as she became less active her weight increased. Walking was the only activity she was allowed, and this had to be carefully monitored. She began having respiratory problems in April, 1988. She could not breath lying down -- she had to sit up or be elevated on pillows. After 10 days of antibiotics, it did not improve and she was hospitalized. She also began a menstrual period two days prior to hospitalization which she had never before had. Immediately upon hospitalization, hypoxia was suggested and confirmed and oxygen administered along with lasix, theophylline and an antibiotic. Pneumonia was guessed but, due to the inability to get a clear chest x-ray was never confirmed. An enlarged right side of the heart indicating congestive heart failure was confirmed. Menstrual bleeding was confirmed and she was started on a hormone therapy to regulate the periods and possibly help with the fluid retention. The therapy worked to a degree but she began bleeding every time she became congested throughout her illness no matter when it was. My theory was that this was the body's way of attempting to rid itself of excessive fluid buildup.

After three weeks, it was decided no more could be done. This was after every test conceivable had been administered. She was brought home and was on an in-home program of 3-5 liters of oxygen 24 hrs. per day plus theophylline and lasix. These were regulated by visiting nurses until the theophylline could be adjusted to her system. Unfortunately, the bouts of respiratory distress became more frequent and meds had to be continuously increased as did the oxygen. She died February 12th peacefully at home with us. She was 24 yrs. old.

My suggestion to other parents is to watch for signs of hypoxia! Lynnel periodically turned blue

(her lips, her fingertips, her toes) all through her life usually during times of excitement or stress. This was never mentioned to me as her possibly being hypoxic, and it was so very difficult for anyone to draw blood that they seldom did when she was in the hospital or just to see a doctor. Fortunately, during this last hospital stay, there was a doctor there who could draw blood and they did numerous tests by being able to do this. I firmly believe she had a degree of hypoxia all her life--even before she became so heavy. The doctors stressed the fact that a person's body usually always maintains the proper amount of oxygen no matter what their size is, and they never figured out the problem as her lungs seemed clear. Also, there is an overall deterioration of the lungs due to PWS children always being very shallow breathers. Lynnel was never capable of taking a deep breath as a normal person is, and this caused the lungs to atrophy over the years. Lynnel was also confirmed as having sleep apnea several years ago, and the test was repeated in 1988 and confirmed that the sleep apnea had worsened. The doctors also suggested this really has nothing to do with a person's weight. Although, through the years, it certainly has something to do with the oxygen saturation in the body.

Lynnel was first on a waiting list to go into a PWS home three years ago which was just three miles from our home. Naturally, after she became sick, it was too late. Lynnel was an only child, her step-father is a paramedic, she was an extremely happy-go-lucky young adult, always had a smile and never had the temper tantrums that are one of the PWS symptoms. Consequently, having her at home was a joy for us. However, when she was encouraged by her school to go into the PWS home and was all for it, we went along all the way. Unfortunately, she was not chosen at that time.

In summary, parents must be more aware in the signs and symptoms mentioned above. When the PWS child goes into heart failure, it comes very rapidly. Up until the Thursday night before Lynnel's death on Saturday night, she was not in enough distress to really be noticeable. She kept her smiley personality and never once complained right up to her final breath. Her optimism throughout her lifetime would put normal people to shame. She never complained about all her activities being curtailed--when she got better, she would resume them. Their positive attitudes are as strong as their "angelic qualities" in securing food from strangers.

Perhaps more research should be done concerning the oxygen saturation level, the sleep apnea and the shallow breathing in PWS children.



## *A "First-Timer" Comments on the Conference*

It's a long way from New York to Calgary. It's a long way both in terms of miles flown and years passed since the diagnosis of PWS hit our family like a locomotive. I'm on my way to attend my first national conference. On the plane the mind begins to play those tricks for which the human mind is (in)famous -- fear, anxiety, dread, apprehension, and just about any other synonym my thesaurus can come up with. The fear, etc., is tempered with a feeling that I'm doing a very positive thing by attending this conference. Maybe some of the fear can be assuaged through the learning process. *Maybe I'll find answers -- if not answers, maybe I can begin to better understand my questions!*

The plane lands. *Reality is just a short ride away.* Arriving at the hotel I enter the lobby. In the lobby are several people with PWS. Some are young children; some are adolescents and adults. Some are obese; others have obviously been able to control their weight. *Am I looking at Emily's future?*

One thing about PW kids -- and adults -- they are friendly and will usually strike up a conversation with you right off the bat. "Are you a Prader-Willi?" one youngster asks me (I'm a bit overweight, so maybe he was fooled!). "No," is my reply, "but I have a daughter who is." "How old is she," I'm asked. "She's six," I answer. What's her name, my new friend asks. "Emily," is about all I can offer as the tears begin to ooze, even though I try hard to be brave. "Oh," is his answer. "See you later," he concludes our conversation with a sincere handshake and moves on to another adventure. *Not a bad-looking kid. Maybe Em will grow up to be like him. Wouldn't that be nice!*

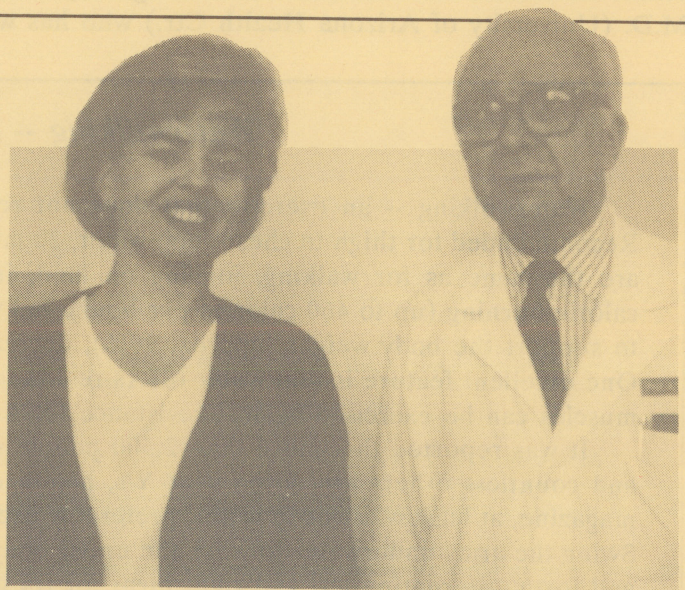
The ice being broken, the initial emotions having been encountered and shelved for the time being, I can concentrate on getting down to business, getting to know my new "extended family," getting on with my life as a father of a child with Prader-Willi Syndrome. As the hours and days unfold, I reaffirm my belief that attending the conference was perhaps the most positive action I could possibly take. There's a perspective out there, and maybe I'll pull it into focus as the week goes on.

I'll not clutter up my reminiscences with the facts of the conference -- they're in my mind for keeps and will surely be recounted by others. There was much more going on in Calgary that week than the superb meetings, lectures and workshops. What I hope to convey to you, readers of this newsletter, most likely parents of PW people, is that by attending the national conference, I realized for the very first time that I am not alone. Not by any means. There is a family out there waiting to share with you, waiting to help you handle the problems, the pain, even the pleasures, eager to get in touch and stay in touch. Next year we meet again, and I will be there. In fact, I wouldn't miss it for anything. I hope more of you will be there, too. I won't be a "rookie" anymore, so maybe I can be a comfort to you just as so many fine people I met were a comfort to me. -- *Sheldon L. Tarakan, Long Island, NY*

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### *Dear Gathered View . . .*

"Just thought I would share with you a picture from our recent trip to Switzerland. We stayed three days in Zurich where the Children's Hospital is located. This is the hospital our famous Dr. Prader is located. We ventured out one morning and with a little luck and the grace of God we were able to meet him. We had a nice talk and he asked of our daughter, Beth. When we told him that she was 14 yrs. old, 56" tall and 85 lbs., he looked at us and with all sincerity he said, 'you are to be congratulated'. Dr. Prader is a very humble and gracious man, giving much credit to his partners Drs. Labhart and Willi. We thanked him for all that he has done and he stated, 'I did very little--there is so very much more to be learned and discovered.' Meeting Dr. Prader was one of the highlights of our trip, and we felt it a great honor to talk with him.



Also, July 1st, our newly formed North Carolina chapter of PWSA was honored with the presence of Dr. Jeane Hanchett from the Rehabilitation Institute of Pittsburgh. She presented a very informative talk on PWS. As always, we very much enjoyed being with Dr. Hanchett--she is at the top of the list of dedicated professionals and a wonderful human being." -- *Penny and Bob Townsend*



### *For Your Information*

Louise Greenswag, Ph.D. and Randell Alexander, M.D.

*Question:* I read about precocious puberty in the Dec.'88 issue of the GV. My 7-yr. old started to develop pubic hair, oily scalp hair and blackheads around her nose. She turned 8 in 4-89 and I have noticed that the amount of her pubic hair has not increased, there has been no breast development or appearance of underarm hair. I have many concerns but I am primarily interested in knowing if she will remain shorter than the average PWS female because of the onset of this "puberty" so young. What has happened to similar cases? We will appreciate any information and will be glad to share our daughter's history to help others. Mrs. K. Costa

*Answer:* To better understand what you are observing in your daughter, it is important to define two terms: "true" precocious puberty and "pseudo-precocious" puberty. True precocious puberty involves an activation of a part of the brain known as the hypothalamic-pituitary-gonadal axis. When activated, this part of the brain begins cyclical release of brain hormones which in turn triggers the release of sex hormones from the ovaries or, in the case of males, the testes. This "true" precocious puberty does not fully occur in individuals with PWS.

Pseudo-precocious puberty involves physiological mechanisms other than triggering the activity of usual sex hormones. The vast majority of cases occur when the adrenal glands (which sit on top of the kidneys) release more than the average amount of a particular hormone. Curiously enough, this type of male hormone is usually produced in small quantities by both males and females. Children with pseudo-precocious puberty develop sparse pubic hair and sign of acne but do not develop further--exactly what you describe for your daughter.

No doubt you can tell that this topic can get very complicated. Pseudo-precocious puberty has been reported in individuals with PWS. Breast growth has been described which, on examination has revealed fatty tissue rather than gland development. Some instances of "menses-like" spotting has also been observed but not regular menstrual flow based on cyclical sex hormone function. Pseudo-precocious puberty does not cause abnormally advanced bone growth leading to short stature and we are not aware of any reports of a relationship between short stature and precocious puberty in association with PWS. Please keep in mind that females with PWS only average 4'8" in height because of the syndrome itself.

We suggest that your doctor check to be sure that everything is as it appears. We would be interested in following your daughter's progress over a period of time and comparing her growth and evidence of further sexual maturation to other females of her age. If you have other questions, you might contact Suzanne Cassidy, M.D. (University of Arizona Health Ctr.) who has written extensively about hormones in the PWS.

### *Waterwalking -- A New Possibility*

Waterwalking -- an exercise that does not require special equipment or even swimming skills. Recommended for thigh to chest-deep water, 20 minutes at least three times a week. Potential benefits are the same as for walking, jogging on land, improved muscle tone, heart health, flexibility and calorie-burning (up to 460 calories per hour), but without the sweating and risk of injury. Water helps to support the body weight (approx. 90%), decreasing the trauma to the weight bearing joints. One excellent feature is that water offers resistance to movement in all directions. This means all the muscles can be exercised against resistance for a more uniform workout.

It was reported that more than 2,500 people waterwalked at the Cleveland YMCA, Norman, OK, and countless others are doing it at Y's, health clubs and in private pools. According to "Walking" magazine, at least waist-deep water, preferably chest-deep is recommended for increasing calorie burn. Swing the arms underwater to work the upper body. A long straight pool is ideal but you can walk back and forth, alternating left and right turns, or try walking backwards, sideways alternating short, quick steps with long steps or even try some fancy step kicking or knee-touching with opposite hand or elbows. Sounds good, especially for those who are overweight and exercise can threaten joint injuries.