



The

# Gathered View

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

## Prader-Willi Awareness Day

*Nationwide Event Planned by Chapter Leaders for Spring of '95*

by Don Goranson

*Vice President, PWSA (USA), and Chair, Public Relations Committee*

**T**he first-ever National Prader-Willi Syndrome Awareness Day has been set for April 29, 1995, and by now each chapter should be actively engaged in planning its own statewide or regional observance to take place during the week of April 29 through May 6.

Participants in the annual Chapter Presidents' Day meeting July 20 in Atlanta heard a presentation on Awareness Day, gave their feedback, and then shared in spirited discussions, brainstorming, and decision making. The presentation was made by Gay Harrington of Massachusetts, a sibling and member of the New England Chapter, who had been recruited by the board's Public Relations Committee to head the "conceptual" phase of National Awareness Day planning.

Countless ideas were hatched at the round-table discussion by chapter presidents, so many that time ran out and chapter heads decided to reconvene at 7 a.m. the next day over breakfast (perhaps a first for an annual conference) to continue their initial Awareness Day planning.

The concept of *awareness* has been clearly defined (along with

support and education) as one of the three major purposes of our national organization and, by extension, our 32 state and regional chapters. What we have not enjoyed to this point, however, is an opportunity to build unity — one time during the year when we all pull together in a joint effort.

By a vote of our chapter presidents, the official theme of the first National Awareness Day will be "A New Horizon for Prader-Willi Syndrome." This was one of a dozen themes proposed, but the "New Horizon" concept won by a nearly unanimous vote because of the tie-in with our new national logo and colors.

In addition to the dates and theme, chapter presidents reached consensus on a number of related issues. Among them are the following:

- That chapters attempt to plan a fund-raising event as a part of their Awareness Day activities. The national organization will provide — at no cost — copies of the new Prader-Willi brochure for use during these activities. National also will work to make premium items (tee shirts, key chains, refrigerator magnets, hats, etc.) available at cost. And it was

decided that there would be a 50-50 sharing of fund-raising proceeds between PWSA (USA) and individual chapters.

- That an Awareness Day "idea bank" be available at our national office in St. Louis. Dozens of ideas were suggested in July during the Atlanta conference, and these are being forwarded to St. Louis. Many of them are pledge-driven, meaning givers need not be present at the actual event.

- That PWSA (USA) work to get an official Congressional designation for PWS Awareness Day. While we did not feel this could be accomplished in time for our first observance next April, a respected Washington lobbyist with a connection to our organization has offered to help, and we hope to have further details about this in the near future.

- That we write to President and Mrs. Clinton next April during our first Awareness Day event and ask for their support. This suggestion proved to be so popular during the Atlanta conference that a letter was drafted on-site and hundreds of conference

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## Out of the Office

*by Russ Myler, Executive Director*

"I'm sorry, I never heard of that" is usually the response I get when I tell people for whom I work. I suppose we all can relate to the experience of having always to explain the syndrome. Sometimes it feels like we educate the

American public one person at a time. Well, all that will change if the national board achieves its plan.

Public awareness of the syndrome and PWSA (USA) is a high priority for the Association. PWSA (USA) has had successes in reaching many local media and two national appearances (on the Maury Povich and Jerry Springer shows). These successes came from the hard work of either chapters, national, and/or individual members across the country. Nationally we had not prepared for a systematic program to regularly increase our visibility. All that changed at the annual meeting of chapter presidents in Atlanta this year. Presidents had an excellent training session by a professional in the field on how to work with the media. They also began planning for the first nationally coordinated "PWS Awareness Day" to occur next spring.

Thanks to corporate sponsor BellSouth's development of an excellent press kit we will use for Awareness Day, we now have a tool we need. Prior to the conference we saturated the local media, using the kit, with information about the conference. As a result, we can now add Leeza Gibbons to the talk show list and add international coverage through the CNN program of "On The Menu." The staff at the national office is now frequently talking to reporters from all types of the "mainstream" press. As a regular part of our press contacts, we refer to local members or leaders for further information.

Won't it be nice to have someone say, "Oh yeah, I just read about that" ... or "I just saw something about that on TV." That day is coming ... SOON!

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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.

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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.

## Organization News

### New Voices

The alto and soprano voices you hear when you call the national office are owned by Cathie Stricker and Maureen Brown, respectively. Cathie began as half-time office assistant in August. Maureen began September 19 as office manager following Myra Miller's departure to assume her duties as executive director for the Ozark Area Camp Fire Association. We are sorry to lose Myra and wish her all the best in her new position.

Maureen and Cathie are siblings of a man with PWS who lives in southeast Missouri. Maureen has worked as an office manager of a major transportation firm located in St. Louis and is returning to work following a period as full-time wife and mother. Cathie began as a volunteer in the office and accepted the half-time position authorized by the board in July. Cathie has completed her degree in engineering and is looking for full-time professional employment. She assures me that her volunteerism with the Association will continue after she secures her professional position. Both Maureen and Cathie are involved in the Missouri Chapter.

Next time you contact the office, be sure to welcome them to the staff. But PLEASE don't ask them to sing!!!!

### President's Message

Jerry Park, our new president, had every intention of having his first article in this issue. However, living with a person with PWS does involve the unexpected. The unexpected happened and much time has been spent in the resolution of same. The President's Message will appear in subsequent issues of *The Gathered View*.

—Russ Myler

### Awareness Day

(continued from page 1)

attendees signed it. Chapter presidents were asked to gather the signatures of non-conference attendees — as many as possible — and forward them to our national office so that they may be included with the mailing.

It is hoped that this fall will have been used as a time for planning, spreading the word, and generating spirit. Chapter presidents had been asked to report to the national office on their initial progress by mid-October.

Awareness Day communications and coordination are to be assigned in a variety of ways, including the *Chapter Presidents' Quarterly*, the national office serving as a central clearinghouse, members of the board's Public Relations Committee and individual board members who have been asked to maintain close contact with those chapters to whom they have been assigned liaison responsibility.

Specific plans are to be reported by the end of January, and orders for special materials must be received no later than February 15.

## Calendar

**Oct. 28-29 - PW Florida Association** Joint Conference with Angelman Syndrome Foundation, Delta Orlando Resort, Orlando, Fla.  
Contact: Wauneta Lehman, (407) 773-9503.

**Nov. 5 - PWSA of New England** First Annual Fundraiser, wine and hors d'oeuvres party, 7 p.m., Bedford, Mass. \$15/person or \$25/couple.  
Contact: Meg Comeau, (617) 275-9586.

**Nov. 13 - PWSA of Maryland and Metropolitan Washington** general chapter meeting, Beltsville, Md. Speaker: Suzanne Hart, Ph.D., on oral/dental characteristics; dental exams available.  
Contact: Linda Keder, (301) 384-4955.

### 1995 Events

**Jan. 20-22 - PWSA Board of Directors** meeting, St. Louis, Mo.  
Contact: national PWSA office, 1-800-962-4797.

**April 27-29 - Third Joint Conference of New England and New York Alliance** chapters, Burlington Marriott, Burlington, Mass.  
Contact: Meg Comeau, (617) 275-9586.

**April 29-May 6 - First National PWS Awareness Day** events. Contact the national PWSA office or your local PWSA chapter.

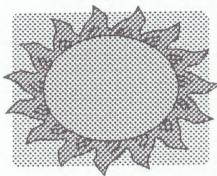
**June 15-18 - Second Prader-Willi Syndrome International Scientific Workshop and Conference**, Oslo, Norway. Send inquiries and requests for materials to the attention of Christian Aashamar, 2nd PWS International Conference, Frambu Helsesenter, 1404 Siggerud, Norway.

**July 20-23 - 17th Annual PWSA National Conference**, Seattle, Wash.  
Contact: Steve Lundh, (206) 285-7679.

These winter dates may seem to be far off in the future, but they are just around the corner. Planning must be completed by that time if we are to enjoy success. And our initial Awareness Day **MUST** be one that we will want to look back upon as having been lots of fun, financially rewarding, and successful in the spirit of enhancing greater awareness for the cause that has brought us together.

Watch for more details on PWS Awareness Day in future issues of *The Gathered View*.

## Chapter Notes



### Keep Hope Alive in '95

Greetings from beautiful, sunny (yes, *sunny*) Seattle, Washington. The Northwest Chapter of PWSA would like to take this opportunity to personally invite you to join us in July 1995 here in Seattle for the 17th Annual PWSA National Conference. As you can see by the title of this article (which also happens to be our vision for the '95 conference), we are very serious about making this an event you will not want to miss.

We have three main goals for Conference '95: to provide you with the latest information on all aspects of PWS, to make sure that there is ample opportunity for everyone to meet and share with old and new friends, and—last but not least—we want to make you laugh. Laughter has a medicinal purpose all its own and, besides that, it's fun!

Watch for Conference '95 updates in future issues of *The Gathered View* and plan now to join us in Seattle next July. If you have any questions or suggestions for the '95 conference, please feel free to phone me at (206) 285-7679 or fax to (206) 284-5260.

Steve Lundh

'95 PWSA Conference Chair

*...“As I reflect on my participation at the recent PWSA national conference in Atlanta, I was very impressed by the continuous improvements made in the quality of each year's national conference. The planning committee promised a program that every member of the family would find interesting, informative, and fun. From the pre-conference sessions to the closing remarks by the Georgia committee, the conference exceeded that promise! The Northwest group will have a challenge next year in Seattle to top this year's conference in Atlanta.”...*

Jim Koerber

President, Prader-Willi California Foundation  
PWCF News

### More Rave Reviews

“What a wonderful time and learning experience our Florida families had in Atlanta, Georgia, at the Prader-Willi Syndrome Association National Conference in July! One of my favorite seminars was ‘Estate Planning for Persons with Disabilities.’ The presenter was Richard W. Fee, M.A., M.Ed. He gave us 12 Basic Life Planning Steps ...”

—Sandy Stone, Florida Chapter

*Editor's note: For information on this firm's nationwide estate planning services, contact Richard W. Fee, Executive Director, Estate Planning for Persons with Disabilities (EPPD), 1200 Corporate Dr., Suite 330, Birmingham, AL 35242. Telephone 1-800-448-1071 or (205) 803-6800.*

## Wisconsin Research Update

As of the end of September, Dr. David Allen, the pediatric endocrinologist attempting to launch a growth hormone study in Wisconsin, has not been able to secure sponsorship from a pharmaceutical company. This means that the study won't get started as early as we had hoped. The doctors still hope to find a sponsor for at least a two-year study, if not the full five-year study they wanted. When the funding and approvals have gone through, or if the study is officially canceled, all individuals who signed up as study participants will be notified. I know this is disappointing news, but we will have to be patient and keep our fingers crossed.



PWSA of Wisconsin, Inc., would like to thank Russ Myler and the national PWSA organization for their recent support of our one-day workshop on “Addressing the Residential Needs of People with PWS.” More than 100 residential providers, case managers and parents attended this workshop. We were very pleased with the first conference that has been sponsored by our organization. It was a start at getting providers across the state, as well as from other states to address this much needed topic.

Barb Dorn

President

PWSA of Wisconsin

## New Chapters

Welcome to our two newest affiliated chapters: the **New Jersey** state chapter and the regional chapter of **Maryland and Metropolitan Washington, D.C.**

Call the national office to locate your nearest PWSA chapter and phone numbers for local chapter officials.

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## From the Home Front

### Someone Else Out There Like Us

For the first time as a Prader-Willi parent, I was able to feel that there is someone else out there like us. The article in the June Gathered View about Steve Oakley could be a replica of our Joanne. However, Joanne is 26 years old and still being caught searching for food.

I have locks on the refrigerator. I have to keep the cellar door locked because all other edible items are downstairs. Joanne doesn't eat toothpaste, but if she gets into the freezer, she could consume raw bacon, chicken, etc. There are times when I think we have everything under control, [then] I'll find empty containers of leftovers under her bed. She's smart enough to find the keys.

I'll find my purse looked through during the night as she searches for the keys. It's like a contest at times as to who could fox who. Her temper tantrums are worse when other members of the family are around; because we don't want to get everyone upset, we give in to her.

She weighs 277 lbs. at 4'8". It's very difficult to find clothes for her. Joanne is the youngest of seven children and lives at home with my husband and me. Thanks for a real good true story.

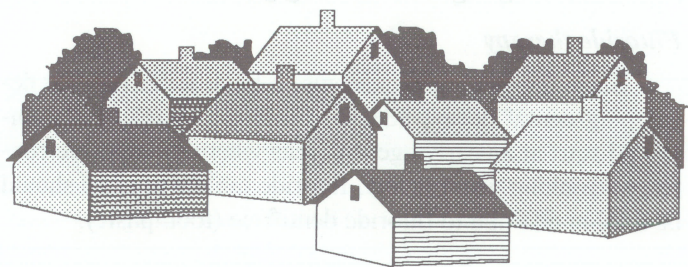
*Louise Vacanelli  
Waterbury, Connecticut*

### The Right Group Home

I just wanted to let you know our daughter Michele has finally found the right group home—finally, after five years of battles. She was placed when she was 16 [and went] from one to another ... She's been in Gainesville, Florida, since March of this year, has lost 36 lbs ... It's one of the best programs going. She reads labels and measures her food. It's hard to believe how healthy she looks. It makes a parent happy knowing your child is doing so well.

The staff makes a difference too; it's hard when group homes change staff a lot. Most of the staff have been there at least three years. ARC has showed us things to be happy about again. Hope this letter can be shared.

*Dave and Judy Kennedy  
Largo, Florida*



### From Russia with Questions

*(The following letter is translated from the Russian.)*

Thank you so much for ... information on PWS. ... We hope your association will certainly become ours from now on! ... We've got rather incomplete command of English, hence we had to have recourse to translators ...

... our baby is 3 years and 10 months old. He walks already independently (started one year ago) but he speaks nothing, just trying to pronounce some syllables and sounds such as "ma," "va," etc. Is that characteristic to the PWS? Can we hope he will start speaking one day? The problem of his speech is exacerbated [because] there are just three teeth left in his mouth ... one of which is very bad and is going to be extracted soon, too! He's got enamel dysplasia, teeth are strongly grinded off, dental abscesses start, and we have to have them extracted. He has already had his 17 teeth extracted ... We had our boy treated at the Genetics Department of the Pediatrics and Children's Surgery Institute in the City of Moscow in June 1993. Their doctor told us this phenomenon is entailed with the low-level immunity. Baby of ours had the whooping cough disease in October 1993. According to advice of the local doctors we gave him three injections of the human immunoglobulin. After this his immune system weakened at all. He is constantly ill with cold. The least contact with sick people makes him ill. ...

... All this hampers his development very much, do not let us carry out all the recommendations of the doctor, since the treatment has to be interrupted. Are the problems of immune system that typical for the PWS? ...

... Our little boy does not still ask for toilet. Maybe it is connected with the general hypotonia! Is that still typical for the children with PWS? Are there any special children's preschool establishments in your country or these children attend some public children's institution?

We have found no doctor here who would be interested in our baby and his disease. These doctors-geneticists do not treat such children and scantily know about the PWS. That's why it is very important for us to keep connection with your association, contact with other similar families! ...

*Tatyana and Sergey Baranovs  
Republic of Moldova (ex U.S.S.R.)*

*(Editor's note: The Baranovs' letter also included questions about their son's adverse reactions to inoculations and about treatment for underdeveloped genitals and undescended testes. In addition to providing this family information, PWSA referred them to a PWS organization in Poland.)*

*After Dr. Kari Storhaug's informative presentation in Atlanta, many conference attendees asked if she would put her findings and recommendations in writing. In response, Dr. Storhaug sent us the following article.*

# Dental Health Problems in Prader-Willi Syndrome

by Kari Storhaug, D.D.S., Ph.D.

Resource Centre for Oral Health in Rare Medical Conditions

University of Oslo, Norway

**N**ot much research has been carried out on oral/dental characteristics of Prader-Willi syndrome. However, I will in the following try to present the essence of what has been published and what I have found after having examined around 50 individuals with the syndrome. I will also outline recommendations for the prevention of avoidable dental health problems in PWS.

## Oro-facial Findings

The specific facial features of PWS are well known, and the **muscular hypotonia** frequently causes a dysfunction of the oro-facial muscles. This may result in a high arched palate, shorter dental arches, and crowding of the lower front teeth.

A survey was carried out on 30 PWS children in Norway. There were 16 girls and 14 boys participating, and the mean age was 12 years, the range 7-16 years. Two kinds of x-rays—orthopantomograms and lateral cephalograms—were taken of all but three children who would not cooperate for the cephalograms. Impressions of the upper and lower jaws were obtained from 23 of the children. Measurements were made both on available x-rays and plaster casts made from the impressions. Comparisons were made with established standards for girls and boys of different ages.

The results showed a pronounced deviation in lip-morphology in the PWS children, with retrusive lips and a short upper lip in 24 of the children. Twenty-six had increased posterior palatal height (back part of palate was higher than normal). Twenty-five children had a shorter lower jaw and 22 a shorter upper jaw than we normally see. All girls except two and all boys except four had crowding (lack of space) of the lower front teeth.

The sparse literature on this issue also reports a high frequency of dental decay, poor oral hygiene, gum bleeding, hypomineralized (weak) tooth enamel, delayed tooth eruption, small teeth (microdontia), dry mouth (hyposalivation), and tooth grinding (bruxism). Recently rumination (keeping food in mouth) and vomiting, resulting in enamel erosion, have also been reported.

**Concerning the dry mouth problem**, we tried to measure the salivary flow in 20 patients, with success only in 15. We did two different measurements: **stimulated saliva**, where the patient chews on wax and spits out after a set time; and **resting saliva**, where the patient sits with head bent forward and mouth open so that the saliva drips into a glass tube instead of being swallowed.

Of the 15 who managed to chew wax and spit, 11 had a low flow rate (less than 0.7 ml/minute). The saliva's ability to neutralize acid was normal only in four out of 14 tested; for the other 10 it took much longer than normal for the saliva to neutralize acid. This means that there will be an acid attack on the enamel for a prolonged period after each intake of food or sugar-containing beverages.

The resting saliva test was difficult, but seven managed to cooperate and all had very little resting saliva. The amount of resting saliva in a mouth is considered the most important concerning prevention of dental disease: very low resting saliva—very high risk of dental disease. We can therefore say that many PWS patients are at risk for developing dental disease due to the composition of their saliva.

## Preventing Oral Health Problems

### Oral motor training

For infants and young children with poorly developed sucking, swallowing, and chewing reflexes, oral motor training will often prove beneficial. This training mainly comprises stimulating massage of facial muscles, lips, gums, and tongue combined with sucking, blowing, and chewing exercises (encouraging use of pacifiers, drinking from straws of varying lengths and widths, blowing bubbles and wind instruments, using sugarless chewing gum, etc.).

### Fluoride therapy

Since the PWS group seems to be at great risk for developing dental decay, adequate fluoride supplementation should start at an early age. In areas where the fluoride content in drinking water is low, fluoride tablets or rinse should be used in addition to fluoride dentifrice (toothpaste).

### **Diet control**

The most serious challenge for dental health in PWS is diet control. Frequent intake of sugar-containing food and beverages is the most common reason for development of dental decay. Thus diet control enhances both general and dental health. Outside of regular meals, sugarless drinks and foods should be the rule.

### **Oral hygiene**

Regular toothbrushing twice daily may start as soon as the first teeth appear. Advice on suitable brushes and techniques can be obtained from dental health personnel. Thick brush handles and special brushes (e.g., Collis curve, Action 2) may be a help when the PWS person is going to carry out toothbrushing herself. Chemical toothcleaning with chlorhexidine rinse and use of sugar-free chewing gum to stimulate salivary flow may also be useful.

Increasing salivary secretion by medication is recommended by some doctors. The effect of such medication and of other ways to stimulate salivary flow should be investigated further.

### **Regular dental visits**

Early contact (around 1-1/2 years of age) and frequent visits, four times per year, will help the PWS person become familiar with the dental office and the staff. Examination, preventive treatment with fluoride varnish, and early intervention if dental disease develops will hopefully eliminate the need for extensive treatment and use of general anesthesia in the future.

### **Orthodontic treatment**

Many PWS persons will need orthodontic treatment, and all children with the syndrome should be assessed by an orthodontist at 10-12 years of age or earlier.

### **Growth hormones**

If the PWS child is going to start growth hormone treatment, profile x-rays and jaw measurements should be taken prior to treatment, and the growth pattern of the lower jaw should be followed during treatment. This is because there are reports on an accelerated growth of the lower jaw during such treatment.

### **Conclusion**

PWS persons have various dental health problems which are related to the syndrome. More research is needed both in the study of salivary flow and composition and in the area of facial development.

However, we have enough knowledge today to prevent dental disease in Prader-Willi syndrome.

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*Editor's note: Dr. Storhaug can be contacted at the following address:*

*TAKO-Centre  
Resource Centre for Oral Health in Rare Medical  
Conditions  
P.O. Box 1109 Blindern  
0317 Oslo, Norway*

## **Toothbrush Tips**

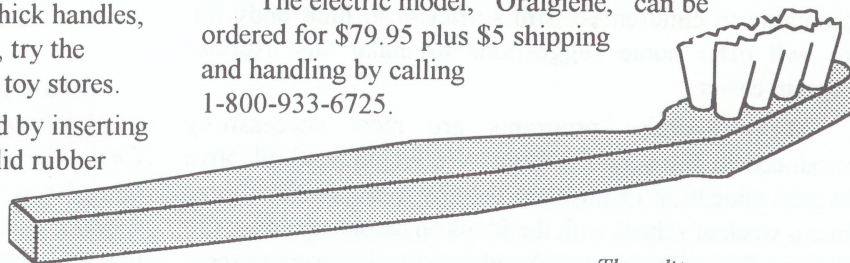
**A** thick handle makes a toothbrush easier to grip and maneuver. Some commercial brushes can be purchased with a thick handle; the other option is to build up the handle on a standard toothbrush. For example:

- "Reach" and "Crest Complete" are two brands that offer children's toothbrushes with thick handles decorated with rubber for a better grip.
- Electric toothbrushes of all types have thick handles, but they tend to be expensive. For a child, try the battery-operated type commonly found in toy stores.
- A standard brush handle can be enlarged by inserting it in a bicycle handle grip or through a solid rubber ball, or by adding special grips commonly sold through adaptive equipment/occupational therapy catalogs.

We have not yet identified the source of the brushes Dr. Storhaug recommends, but we've found ads for two U.S. brushes — one manual and one electric — that sound similar. They have multiple brush heads, enabling a person to brush all surfaces of a tooth in the same motion.

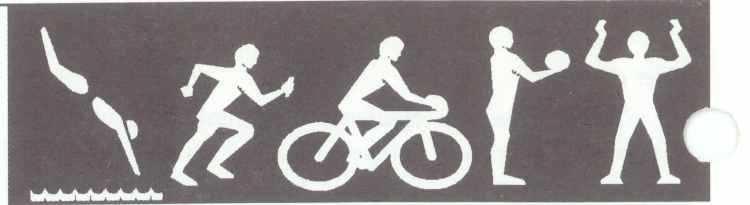
The manual model, "Dentrust," might be found in local stores; if not, call 1-800-872-0099 to order it. (Note: It has a standard toothbrush handle.)

The electric model, "Oralgene," can be ordered for \$79.95 plus \$5 shipping and handling by calling 1-800-933-6725.



— The editors

# An Exercise Program for the Child with Prader-Willi Syndrome



by Randah Whitley, P.T., and Adrian Sandler, M.D.

**W**eight control is a critical issue for families of children with PWS. Most families are counseled early about nutrition and diet. Although exercise has probably been mentioned, it is usually harder to implement an exercise program into a family's busy routine.

Some studies of normal children have shown that limiting caloric intake and increasing the level of physical activity early in life will prevent progressive obesity. Other studies have shown that interventions early in life can decrease the number of fat cells as well as control long-term weight gain. Exercise may prevent future obesity by the same mechanism. Young rats in a swimming program in the first month of life were noted to have lower fat cell counts than either dieting or control animals.

The ideal form of exercise for a child would be aerobic, involving high caloric output, no special skills or equipment, little risk for injury or discomfort, and opportunity for socialization. Endurance activities such as walking, biking, running, and swimming come closest to meeting these goals. However, dancing to rock music or jumping rope with a friend or sibling is just as acceptable. Weight lifting and high-resistance exercise do not burn fat but do raise body metabolism by increasing muscle work. Resistance exercises performed with a high number of repetitions may be more useful in burning calories. This may be especially important for hypotonic children.

Getting inactive children to exercise is not easy. Recommendations to "eat less" and "exercise more" will have little impact because children and their families need specific recommendations. Obese and/or inactive children lose interest in exercise, so weight control becomes a family problem. The objective of a balanced therapy program is to introduce dietary manipulations, aerobic and resistive exercise, and behavior modification modest enough to be acceptable to children yet still sufficient to limit body fat. We will offer some suggestions to make this exercise program easier.

Many exercise programs are most successfully introduced in the school setting. We recommend adaptive physical education or physical therapy a minimum of three times a week at school with the focus on aerobic exercise (20 minutes of non-stop exercise) and some resistive exercises.

The home exercise program can be approached in two ways. Most children need adult supervision to exercise for

20 minutes without stopping. This usually means that exercise for the child becomes exercise for the family. Twenty-minute walks around the block, biking around the neighborhood, or a game of basketball in the driveway are ways that families can exercise together. A sticker chart to reward the child can be useful for those children who are resistant to exercise. The child gets a sticker for each day that he exercises, and after he reaches a specified number of stickers he gets a "reward."

**T**he other approach to family exercise is to increase lifestyle activities. This gentle approach decreases the negative attitude of "working out" but will still increase calories burned. Walking instead of riding, not parking in the nearest space at the mall, pacing while talking on the phone or waiting for the bus, using the stairs instead of the elevator, helping mom carry the groceries, helping to push heavy objects, or dancing to music can be encouraged by parents. It may take some creativity on the parents' part to increase a child's activity without calling it exercise.

*In summary*, exercise will burn calories, increase muscle bulk, and reduce the number of fat cells if introduced early in life. Exercise for the child with Prader-Willi syndrome must be viewed as a family problem, requiring cooperation and support by all family members. Exercise programs for children should be conducted with adult supervision, involving activities sufficiently intense to increase caloric output but avoiding bodily stress. It is important to discuss the child's exercise program with a physical therapist or a physical education teacher to ensure that he is getting an appropriate program at school.

*Reference:* Rowland, Thomas W. (1990). "Obesity and Physical Activity." *Exercise and Children's Health*, p. 129-159.

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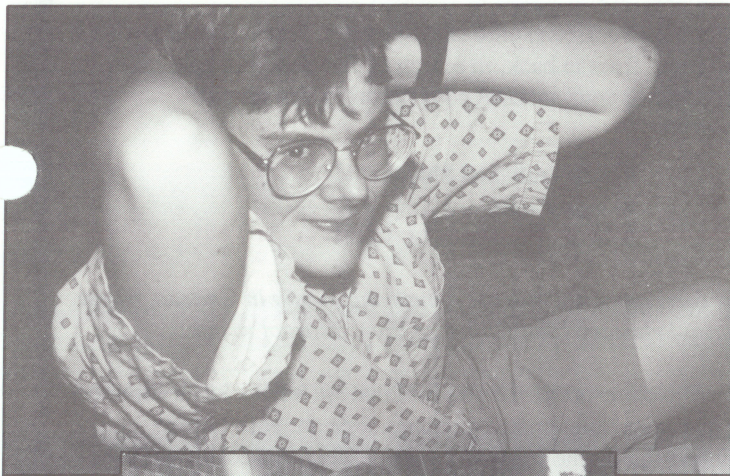
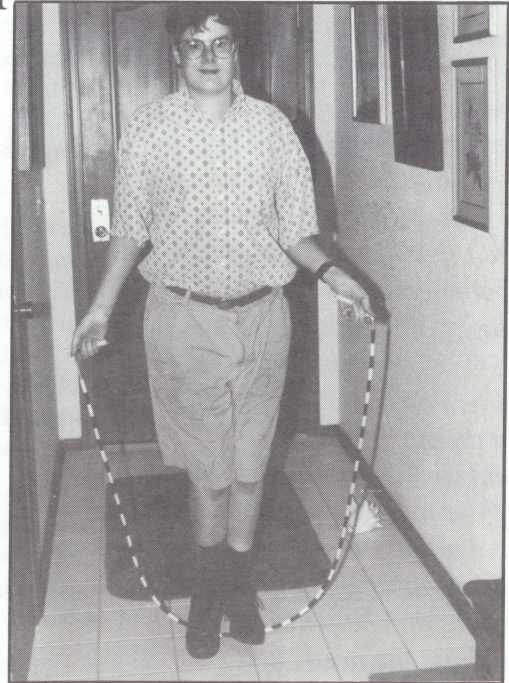
*Editor's note: This article is reprinted from the newsletter of the Prader-Willi Syndrome Association of North Carolina. Randah Whitley is a senior physical therapist at the Neurodevelopmental Nutrition Program (NDNP), Center for Development and Learning, University of North Carolina-Chapel Hill. Dr. Adrian Sandler, former program manager at NDNP, is now at the Thomas Rehabilitation Center in Asheville, N.C.*

## Jon Knows: Persistence Pays Off

by Marian Shacklett, Tulsa, Oklahoma

When our son Jon was about 13 years old, we realized that exercise was needing to become a key issue in our lifestyle. We thought about and tried out many forms of exercise — walking, jogging, swimming. Somewhere along the line, we got hold of a jump rope and Jon tried it out. At first he couldn't even jump at the precise time to get the rope under his feet. I wondered whether this form of exercise would even be a possibility because of his lack of hand/feet coordination. But we decided to give it another try and another and another. One, two — he got the rope under his feet and over his head without getting all tied up in knots. What a success!

Each day we would practice — each day he would improve — four jumps, then six, eight right in a row. We set a goal of 50, stayed with that for a while, next 100, then 250. We saw how he was progressing and jumping with ease and smoothness. We praised him while he enjoyed his successes too. Any resistance he may have had fell away. He knew this was going to be a part of his routine before dinner.



Many years (12) have passed since those early days of struggle to learn to jump rope — and to do sit-ups. Today Jon does 425 jump ropes and 50 sit-ups daily five times a week. If, on occasion, he walks about a mile or so, that substitutes for his jumping. Either my husband or I count for Jon. At times when we can't free ourselves up to count we set the timer for 12 minutes, but this really isn't as effective because Jon seems to rest oftener between jumps or lean over to stare at the timer to see how much time is left.

Finding something that works for Jon and us has been the best thing to keep our son fit.

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*Editor's note: While Jon's exercise program is impressive, we learned at the Atlanta conference that he has other remarkable talents and a generous spirit as well. In addition to his fishing hobby, Jon makes beautiful latch-hook rugs and brought a selection to the conference to sell. The \$400 he made from their sale Jon contributed to PWSA!*



**He gets the fish jumping, too! ...** Jon's fishing hobby may bring him a stringer of catfish from a nearby creek or the "catch of the day" — an 8½ pounder at a ranch in eastern Oklahoma.

## Parent Requests

*While much of the literature about Prader-Willi syndrome focuses on the day-to-day challenges associated with Stage II (insatiable hunger, excess weight, difficult behaviors, etc.), parents need support and information in other stages of dealing with PWS, as evidenced by the letters on these two pages. In the spirit of "sharing and caring," we invite all of our members—parents and professionals alike—to respond to these letters and help us to address the needs and issues raised here. Please send your letters or articles to The Gathered View editors, in care of the PWSA national office.*

### ***Birth to Five***

*In a series of letters to former PWSA President Janalee Tomaseski-Heinemann, a mother from Israel stresses the need for more research and guidance on early intervention strategies and the importance of finding other parents who can share the frustrations and hopes of the early years.*

Dear Janalee,

I am the mother of Guy, who was born on February 21, 1993, and six months later diagnosed by DNA testing as having Prader-Willi syndrome. That short sentence is devoid of all the heartache and trauma involved in those first months of Guy's birth and the endless running around to get a final diagnosis for Guy, who was born severely floppy but with no need for any external aid in feeding.

As Israel has such a small population compared to the U.S.A. and Canada, we don't really have a support group. There is a fledgling PWS Association whose membership is small and the age group of the children is very wide. Also the medical profession are not that well informed, and I rely on my membership in PWSA for aid and information. I am particularly interested in anything to do with early-childhood food conditioning. Are there any studies that are ongoing or published? What I do with Guy is more or less based on common sense: no sweet things, low-fat, low-carbohydrate, even though at this stage he shows no signs of food obsession. I wish that I had more qualified information in that field.

Guy progresses continually, and although I know he will crawl and walk I find his slow progress frustrating, and I am in constant battle with myself trying not to compare him with peers. I have found very few parents of PWS children who even remember their feelings at this stage, and worry that the second stage will be so devastating that this will, on looking back, seem like a pleasant dream.

I envy your ability to organize yourselves into state chapters, and avidly read The Gathered View from back to front, front to back, in search of something to hold onto. Perhaps one year I'll be able to get to your national convention. Keep up your wonderful work. I admire you all.

Yours Truly and Shalom  
Margot Finks  
Kibbutz Ein Hashofet, Israel

*In a subsequent letter, after corresponding with the Armstrong family in Canada, whose letter appeared in the January-February 1994 Gathered View, Margot writes:*

... [We] have started up a terrific correspondence and find that we have a lot in common when it comes to our sons, questions, dilemmas, and hopes. We are both terribly frustrated by the lack of information on the very early years. I feel that covering the topics of early feeding problems, floppiness, and saying that they are rather angelic babies is not enough. I guess what we want is a more definitive plan of action. Perhaps we are just not ready to accept the inevitability of all we read about PWS. I feel that there has to be more advantage in early diagnosis than what is known and would be interested to know if there is any research going on in this area. I know that my hope of prevention BEFORE management may seem overly optimistic or even a foolish dream, but as I have yet to find any information to the contrary, instinct is the only tool I have to go by.

My son Guy is now 16 months old and other than physiotherapy (from 4 months) and speech therapy (starting at one year) we are also building on his love for water and have recently started hydrotherapy. We are actively trying to encourage early food-related techniques, such as regular meal schedules, no snacks or sweets, no food for reward or as a pacifier, and carefully monitoring his weight and height. He is a gorgeous child who started to crawl two months ago and has been making great progress ever since. He now sits, stands, walks when holding on, has a vocabulary of six or seven words, follows simple instructions, has a clear sense of humor and an infectious laugh! ...

June 15, 1994

Dear Friends,

Enclosed please find my check for my annual dues and an additional contribution in memory of my late son Adam. I want to take this opportunity to thank the association for being there. Sometimes, it was nice to know that we weren't alone in our struggle to raise a family and husband sufficient energy to keep going in the face of all the additional work necessary to survive the impact of Prader-Willi syndrome. Not that I am in agreement with those who pose everything as bright and sunshiny, but optimism is certainly a necessary component of survival when dealing with the impact of the disorder and the system which often is unsympathetic to the needs of the individual.

Now that Adam has been dead for almost a year, we are beginning to be able to put his life in perspective, and I am sufficiently far enough along in my own grieving process to be able to commit to paper some of my observations that hopefully might have some value to other parents.

We spent the first 17 years of Adam's life obsessed with food. Every pound gained was a catastrophe. We locked the food cabinets and refrigerator and they remained locked until Adam's death. However, because of Adam's intelligence and ability to move about the community independently, there was no way to stop him from eating and gaining weight. The choice came down to institutionalization or eventual morbid obesity. Adam helped us with that choice. He stated that he would rather live a short life but be able to do what he wanted to do rather than go to some institution and be thin and locked up. He didn't want to be obese, but given the limited alternatives, he didn't want to be locked away. After much soul-searching, we decided to do it his way. He spent his last five years free to come and go as he pleased. He had two minor skirmishes with the police involving tantrumming behavior, but there are plenty of so-called normal people who perpetrate far worse acts in the community.

Adam qualified for SSI and had this independent income that gave him both mobility and the ability to interact with the community as a normal person. He was a collector, and, as I need not tell you, collected obsessively. Adam's legacy includes thousands each of books, comic books, cassettes, and CDs. The value of his collection is such that it will ultimately provide college tuition for his niece who was not yet born when he died last year.

Adam planned his own itinerary. He became an expert at using regional public transportation to get anywhere at any time. As an adult, he would take the casino buses to Atlantic City—not to gamble but to walk the boards and, of course, eat. He tried gambling once, lost a few dollars, and said that it was a waste of money. If you lose it, you can't spend it! He loved wrestling and attended every professional match anywhere in the region. He was insatiable in this interest. Some of the wrestlers took an interest in him, and he was taken backstage and even called by some of them.

Despite rapid and horrendous weight gain during his last few years, Adam was medically healthy enough to have never been to a doctor for other than the flu after the age of two, when he had mild pneumonia. The last day of his life was spent on a trip downtown and an outing to the swimming pool. The fact that it was the hottest day in the history of Philadelphia was the direct cause of his death, although morbid obesity was the underlying cause. He spent 12 hours outside in 105-degree heat and, although he seemed fine when he came home, died peacefully in his sleep two hours later. Again, the compulsion for "more" was insatiable.

The wise and compassionate clergyman who delivered his eulogy told us that we should in our mourning celebrate Adam's life for the fact that he was a man who exceeded his potential. He lived his life to its fullest capacity, and it was his time because he had accomplished all he could in this life.

Today would have been Adam's twenty-third birthday. The check I am sending is the one we would have given him as one of his birthday presents. I believe he would have wanted us to do this—as long as he couldn't be here to spend it! Above all, he was a kind and generous person who cared about others—at least when the disorder that drove his life was not interfering with who he tried to be.

(Continued on page 12)

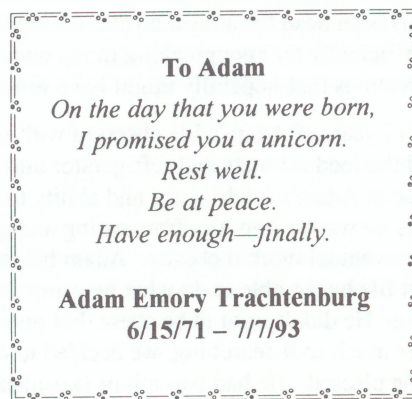
## *Life After PWS*

*In a letter to PWSA, a father whose son died at age 22 asks for recognition and support for the parents who have outlived their children with PWS.*

(Continued from page 11)

There is one small favor that I ask of you. Now that we are healing both from the life and death of our son, I wonder if it is the proper place of PWSA to deal with this parental life-stage since PWS often leaves parents outliving children. This after-life is not like a parent who lost a "normal" child because on top of the grief (which each family handles in its own manner), besides the horror of losing a child, there are the scars of having fought a monumental battle just so that your child could exist in a world hostile to individual differences. Re-attaining a normal life is difficult, if not impossible, because so much of our lives have transpired during the previous stage of life. It was like going to sleep at age 32 and awakening at 54. Perhaps The Gathered View might be open to printing an occasional column about "life after PWS." An increasing number of families will be facing this problem as time passes. Thanks for thinking about this option.

Thank you for allowing me to share my feelings at this moment in my life when I need a listener. I want to close this letter with a brief tribute to my late son:



Sincerely yours,  
Milton Trachtenburg  
Philadelphia, Pa.

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(September)

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*Many of our members contribute to PWSA (USA) through the United Way or other community fund collections. The distribution policies of these funds vary widely. If you are interested in making such a contribution, check with your local fund drive officials as to whether and how you can designate a contribution for PWSA (USA).*

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

