



# THE GATHERED VIEW

Newsletter of PRADER-WILLI SYNDROME ASSOCIATION

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## PRESIDENT'S MESSAGE

With the inception of Social Security in 1936, this past half century has seen the progression of a trend, "It's too complex a problem for our local group to solve, get it done in Washington." About that time it was not uncommon in the newspaper of my hometown to read advertisements soliciting customers to one service or another that ended with "Let George Do It!" This became so common an expression that its meaning shifted to be applied to any task that someone did not wish to make the effort to do, at that particular time. Washington and George, how our founding father has suffered role reversal. From the rugged individualism that caused people to leave oppression in their homeland to a society that would rather let George do it in Washington, has been the social revolution occurring since the depression. National guilt for the less gifted has produced some strange developments. But, we do seem to learn. In an article from the Department of Education sponsored National Information Center for Handicapped Children and Youth, a completely opposite trend in responsibility is identified as growing in importance for planners for the disabled. Initially, reference is made to a statement from the book MEGATRENDS by John Naisbitt, "Decentralization empowers you to tackle problems and create change at a local level. Because political power is decentralized, you can make a difference locally..... Decentralization is the great facilitator of social change". Another pertinent quote is, "Leadership is coming from the bottom up, not the top down."

The effect of decentralization to produce greater local power will only occur when locals exert responsibility and do the work necessary to identify the problem, develop the solutions and demonstrate cost effective resolution. Local means not only geographically local, but more importantly close to the affected persons. In our area of interest, the cost effectiveness of dedicated facilities for support has been clearly proven to be the viable solution as compared to alternatives when the Prader-Willi family home can no longer provide for the needs of the Prader-Willi person. Please, let us not confound the situation by getting into the specific arguments of when that point is reached. It is critical to know that for some families the point of no support is reached and an alternative is essential. We are now at my point. For some there is a need for residential placement supportive to the specific needs of Prader-Willi persons. The current and assumed future needs will not be met by sending the problem to Washington to let George do it.

There is an alternative that can work but it will work only if persons concerned about Prader-Willi syndrome accept the responsibility and make the effort. Who are those people that bear this great responsibility? As Pogo so often pointed out, "It is us". The only people on the old Blue Marble that give a tinker's dam about Prader-Willi syndrome are the people who read THE GATHERED VIEW and if you have read this far, you belong to a very exclusive club, "The World's Experts on Prader-Willi Syndrome".

PRESIDENT'S MESSAGE cont.

At the June, 1984 meeting of your board of directors, the motion was made and carried that funds be designated to explore the possibility of creating The Prader-Willi Syndrome Association National Developmental Center. People have been working on this concept. You have received a product of this initial effort, the complete description of the proposal, as a separate mailing from our executive director, Marge Wett. On December 1, 1984 your board of directors and officers will meet again, at their own expense of close to \$10,000 to determine if it is worth the additional effort on their part to make this alternative happen. George will not do it. Only you, you and YOU can make it happen.

First, you must search in your mind to determine if you believe that there might be a time in your life when this project would be your only alternative to a very difficult decision, even the fortunes of the endowed can be hit. Our responsibilities far exceed our rights, and if each of us fails in our responsibility, there will be no rights for anyone.

When you are clear in your mind that this program is a necessary and viable alternative that may touch your life, then you must study and inquire about the specifics of the proposal's approach to the problem, weighing again any realistically possible alternatives. If you arrive at the conclusion that many have, there is no alternative, then you must join the effort. Write a letter to the office in Edina indicating that you favor the concept. Without that kind of support, the idea will die in December. You do it! George doesn't give a rip!

*Delfin J. Beltran, MD*

LACK OF SALIVA

A recent intensification of research into Sjogren's syndrome has produced some articles regarding the lack of saliva associated with this condition. Jane Brody recently wrote, "Saliva, a complex substance with many important roles, contains digestive enzymes that begin the process of breaking down foods for absorption farther along the digestive tract. Without saliva to wet food and sensory tissue in the mouth, food may have little taste. Saliva contains immunologically active substances that help protect the teeth and soft mouth tissues against attack by bacteria and fungi. Without the natural antibiotic functions of saliva, the teeth are highly vulnerable to decay-causing bacteria and the gums are subject to invasion by microorganisms that cause periodontal disease, which undermines the bone that supports the teeth. Saliva also bathes the teeth with such minerals as calcium, phosphorus, and fluoride, which strengthens them."

Lack of saliva, thicker saliva with crusty formations in the corners of the mouth, strange food preferences, lack of tears, all have been reported by parents -- you can't help but wonder if fluid-secreting glands are also affected in our children.

Some of the suggestions for treating dry mouth included lots of fluids between, as well as with, meals, avoiding citrus juices, limiting caffeine, using a mouthwash with fluoride rather than alcohol, or purchasing artificial saliva products available in pharmacies, if problems seem more acute.

### ADULT STUDY

Louise Greenswag, R.N. Ph.D, reported at the June conference some of the information she obtained through a study of adults (16 and over) with PWS. Her report will be included with the 1984 conference papers. If anyone has a child that will be 16 this month or missed responding to her study previously, she would appreciate having additional people in her study. You may obtain the study directly from Louise at 6401 Utica Ridge Rd., #25, Davenport, IA 52807.

This study is not to be confused with the on-going questionnaires being sent to you by the PWSA national office. This national study is also still in progress as you will note in another article in this issue.

### SUCCESSFUL PLACEMENT

A member writes, I wanted to tell you how much the GV has given me insight on the syndrome. I own an adult foster care home and have operated it for 25 years. Last year I got a young adult with PW. At first I was ready to give up, but then I started reading about the syndrome and it became a challenge. My resident has gone from 215 lbs. to 150, I no longer lock cupboards but the teaching has to be consistent. I give her high protein meals with lots of fresh, low-calorie vegetables.

One thing I have observed, when she has even a small amount of sugar she did not lose and she would have temper tantrums and lose control of her emotions. I feel the more we can learn about the syndrome, the more comfortable we feel about their care.

Her placement in an excellent learning center has gone well, we work together for consistency.

### NEW RULES

Combined Federal Campaign (Solicitation of Federal Civilian and Uniformed Services Personnel) has new rules which will allow contributors to designate any tax-exempt charity they choose.

PWSA does qualify (we are a 501C-3 corporation). We thank member Lou Levisque for designating us for his payroll deductions and sharing this news with us.

### DENTAL INFORMATION

An English member shared the following information with us. Their son has developed damage to his teeth, which at first appeared to be from grinding, but can also be seen on surfaces not in a physical proximity to be caused by grinding. It is a consultant orthodontist's opinion that this damage is a result of the acidic properties of citrus fruits and carbonated drinks.

Since we have two dentists in Minnesota that are treating the fifteen residents of Oakwood, we asked for their comments. Dr. Brandstetter and Dr. Olson wrote:

"Upon initial examination the patients had poor to average oral hygiene. There were higher than average incidents of unfilled and decayed teeth. Their oral hygiene habits were poor, and as a consequence there was a higher rate of gingival disease, gingivitis and periodontitis.

This observation is consistent with other population groups with slight to moderate mental

DENTAL, cont.

retardation. Through extensive oral hygiene instruction and excellent reinforcement and support from the nursing staff at Oakwood Residence, the oral hygiene improved dramatically. A substantial drop was noted in subsequent recall visits in the gingivitis and decay rates, and now is within the normal limits of the general population. The dental disease seem to be related to oral hygiene and diet. There did not seem to be a systemic connection between dental disease and the Prader-Willi syndrome. Dietary changes such as decrease in caloric intake and decrease in the frequency of snacking is believed to be a contributing factor in the normalization of the gingivitis and decay rates.

One note of caution -- one PW patient exhibited above normal demineralization of enamel that may be correlated to an excess of citrus fruits.

There were no other significant dental differences noted with these PWS patients. Dental disease appears to be able to be controlled by eliminating snacking between meals as much as possible and control of the amount of refined sugar in their diets. Excellent oral hygiene habits should be established on a one-to-one basis with each patient. Thorough flossing and brushing according to the Bass technique was used, and appeared on the small group to be able to control the dental disease."

In conversation with Dr. Brandstetter, he cautioned continual snacking, for example giving one section of an orange each hour would cause the acidity to remain in the mouth continuously and be extremely hard on the teeth. This also contributes to their attitude of frequency eating. It is much better to allow one or two snacks per day at set times, and no other food inbetween meals. Oakwood residents are allowed two snacks and three cans of diet pop per day.

A MOTHER WRITES:

As a new member of PWSA, I am very excited and relieved to be receiving information that is such a comfort to me. For so long I have felt so alone in raising a PWS child. Our daughter, Alitha is nine. She is now in a self-contained classroom with EMR children. She is very happy there and the teachers are so willing to work with me on diet and behavior problems.

I have solved her bed wetting problem with an alarm clock that goes off three hours after bedtime. She gets up, turns off the alarm, uses the bathroom and returns to bed by herself. We have a slip up once or twice a month but I can live with that.

A big problem for over a year now is her urge to write in books. She loves to copy chapter headings, connect all the dots, circle certain words. Even though we hide all the pens and pencils, she manages to find one. My question is are PWS children capable of learning right from wrong? Does this concept ever come? Also do they ever learn to share with siblings? Am I wrong to try and make her? Seems like the harder I try the harder she tries. We are much happier if we let her have her own territory most of the time.

Alitha has been a source of frustrations and yet such joys. She really is so loving and been very special in our lives.

One answer: It is my opinion that your child's school placement is appropriate. A lot of parents get arguments from teachers and special education personnel that this placement is too restrictive and they are the "top of the pile" there, but I don't believe they understand the unique needs of our children. I believe you will find the two teacher s' presentations at the 1984 conference interesting. Teachers have to be willing to learn about

cont.

the syndrome and not compare our children with other disabilities that function differently.

Regarding the compulsions, I find they manifest themselves in many other areas beside food. I certainly do not advocate spoiling these children just because I feel their compulsions are a part of their syndrome, but I do feel you have to decide just how much of a problem the compulsive behavior is and then decide whether it is worth the effort to try to conquer it. I believe with the sharing, it is far easier to try to get your other children to accept Alitha's differences. Other children learn that sharing has benefits, this type of abstract thinking is very hard for young people with PW.

When trying to correct a behavior, don't scold or lecture, just set down the rules. It will save you many frustrations and hours. Catching them in the act of doing something wrong does not produce guilt; scolding or lecturing will just produce frustrations for you because they don't respond. Make a rule -- when you do this -- I do that, and stick to it. Be sure the denial is something you can carry out. Breaking rules doesn't mean they can't tell the difference between right and wrong, they just look at it differently than other people. Parents find this hard to accept. It is against our "rules" to make exceptions for this child, we feel all our children should be treated the same. I can't stress how important I feel it is to be frank with your other children about her differences. Explaining to them that Alitha probably has an injury in part of her brain that changes her whole message system may be one approach to use. When you want to move your leg to walk, the brain sends that message and if everything goes as it should, the leg moves. Alitha's brain sends different messages, therefore she does not function the same.

Whether we are working with doctors, educators, group home staff, workshop personnel or whomever, I feel it is very important that you share some of the written materials that PWSA has available. When they accept there are unique things about our young people, they are more willing to develop programs to meet their needs.

### QUESTIONNAIRE

Part four of the National PWSA survey will be in the mail sometime in November. Over 500 parents have responded with the return of at least one or more of these questionnaires. We hope you will continue to participate in this study. After all of this data is accumulated, a wealth of information will be available to be shared with parents and professionals.

The National PWSA office has recently become involved with charges of child abuse against a sibling. The case is based solely on the testimony of the young adult with PWS. Those of us that have been privileged to having attended some of the National Conferences have found through conversation with other parents our children have many traits in common that are not always found in the printed literature. One of these traits includes truthfulness or the ability of determining fact from fantasy. Accumulating information on this syndrome from 500 cases will develop a very accurate picture of this syndrome. This is very important to this family right now, and some day may also be very helpful to you.

If you are not participating at the present time, it is not too late to request the first three parts from the national office. If you are late returning one of the parts, just mail them in at your convenience. All of this information will be shared with the membership after it is completed.

## SUGAR-FREE DIET

A parent wrote: Eight years ago, when Scott was six, I placed him on the Feingold diet, which eliminates artificial colors and flavors. The purpose of this diet is to alleviate hyperactivity in children. I used it to see if I could eliminate Scott's temper tantrums, which were occurring 3-4 per day, lasting 10-15 minutes each. Improvement after a week, reduced these outbursts to one every other day. Scott hated the diet and I found it very difficult to implement; the school found it nearly impossible because of birthday parties and snacks.

After six months, I stopped the diet and Scott seemed to be better able to control his temper on his own. Ever so often I threatened him with that diet when he would have too many "bad days".

At age 12, Scott had his first major tantrum and it lasted 3 hours. Since then he has had two more plus his daily smaller outbursts. We were considering medication or an alternative placement to relieve the tension in our home. Two months ago I read "Allergies and Your Hyperactive Child" by pediatric-allergist Doris M. Rapp. Her theory, rather simply stated, is that many unacceptable behaviors (aggression, tantrums, talkativeness, short temper, hyperactivity) are caused by something just eaten, smelled or touched. She has recorded dramatic results of allergic reactions which are similar to my own son's actions.

By following Dr. Rapp's suggested elimination diet for one week and then reintroducing those eliminated food, one at a time, any adverse reactions can be fairly easily noted. After 5 days on the diet, Scott's disposition improved. On the day Scott went back to eating granulated sugar, he developed a very bad headache within 15 minutes of ingestion, 4 hours later he was fighting with sisters, yelling and slamming doors. On the day artificial coloring and flavorings were reintroduced, he was stamping his feet and throwing things. When eggs, chocolate, wheat, corn, and milk were reintroduced, everything went smoothly. For Scott it appears sugar and artificial flavorings and colors are the culprits.

If our family goes to a restaurant or Scott is going to have restricted food, he takes an Alka-Seltzer without aspirin 20 minutes before eating or as soon as possible after eating and the reaction can be relieved or even eliminated. For 1 month Scott has had restricted food only twice without the bicarbonate preparation and he became argumentative, cried easily and was generally unhappy. The rest of the time Scott has been like a new person, he is happier and has not had a bad day at school in spite of the fact he just began junior high.

We eliminate sugar (brown, granulated and powdered), corn syrup and dextrose but allow fructose, honey, pure maple syrup and sweet & low. I purchase hot dogs, soups, granola bars, cereal, ketchup, mayonnaise and salad dressings, ice cream, yogurt, bread, candy, fructose and cookies in the health food store. From the supermarket he can have most juices (not juice drinks), shredded wheat cereal, Health Valley granola bars, certain breads, Mott's natural applesauce, Hunt's tomato paste, white cheeses, some Progressa soups, sugar-free peanut butter (usually found in the refrigerated sections). After about three weeks of label reading, shopping is becoming easier again. I have found by shopping very carefully at health food stores, Scott can have treats (ice cream, cookies etc.) sweetened by honey or fructose. As long as he can have his occasional "junk food", Scott is tolerant of his new diet. He feels happy with himself and his ability to control his behavior and our family is basking in a new-found peace. Surprisingly too, Scott has lost 3 lbs. in the last month even though we haven't strived for that. It appears without sugar, he can eat more calories and still maintain his weight. While the diet is difficult, the results are fantastically encouraging to us.

DIET, cont.

When using recipes that call for sugar, I substitute fructose. Once baked, the cookies or cake do taste slightly different but not offensive. Some recipes I use:

**BANANA SMOOTHY**

1½ c. skim milk    1 lg. banana    1 T. honey    ¼ t. vanilla  
Combine in blender and mix well. Serves 2 or more.

**FROZEN FRUIT SLUSH**

1 can fruit (packed in own juice, frozen)

Defrost slightly by holding can under hot running water for 1-2 minutes. Open can, pour contents into blender. Blend to slush consistency and serve in glass with a straw.

**SNACKS**

Add honey, sliced banana, grated carrots and raisins, cut-up dates, carob chips to sugar-free peanut butter. Spread on sugar-free bread, celery sticks, apple slices, sugar-free graham crackers, or cucumbers.

NOTE: The medical profession has been very critical of Dr. Feingold's diet because of his statistics and reports. The University of Washington did a controlled study of sugar ingestion on nine patients in 1979 or 1980, and found sucrose did not have an adverse or a favorable effect on the motor behavior or error performance of the children but authors Peggy Otto and Dr. Sulzbacher did state, "Certain individuals might show a relationship between sugar ingestion and behavior change, but more extensive research is needed in this area." (National Conference Papers, 1980). Other experts have commented that the extra attention which accompanies a special diet may contribute to behavioral changes, but the PWSA office has received a fair number of letters over the past couple of years such as the letter quoted above. It certainly seems reasonable to give any healthy suggestion a try. Improved behavior and a happier child seems worth the efforts to try some elimination techniques.

Any rebuttal from the medical profession or other parents?

ANOTHER PARENT WRITES:

Our daughter is now residing in a State Institution. In the past year she has lost 88 pounds. We had made application for our daughter for a PW group home in our state but our county refused to pay out of county funding -- instead she was placed in the state institution at triple the cost.

I am angry about this. They tell me the state funds this but we are the states money source through our taxes. We had kept our daughter at home for 22 years but she had become so heavy she could hardly walk and this was our only alternative.

1984 NATIONAL CONFERENCE PAPERS

We have followed our usual procedure in taping and transcribing the presentations made at the June Conference in Minneapolis. These papers will be ready for purchase by the time this newsletter arrives. Each set will be \$6.00 to cover the cost, order from the PWSA national office.

## SLIDE PRESENTATION

The professionally done slide presentation that includes 60 slides and an audio tape, runs for 12 minutes, has proven to be very helpful to present the PWS to groups either for educational or fund raising purposes.

This can be borrowed from the National PWSA office or purchased for the \$60.00 cost to reproduce it. Many individuals and chapters have taken advantage of its use already.

Bud Bush writes from California that he recently subbed for Lee Shadell and made a presentation at the bi-monthly meeting of the Association of Regional Center Agencies in LA. "I used the audio/visual slide presentation for the approximately 60 persons, which included delegates from each of the 21 regional center agencies in CA. It was a great opportunity to reach the entire regional center system. From comments received afterward, I would say it was a success. The slides with audio were an effective way to give an overview of the syndrome. Following the presentation, I made my pitch which covered these specific requests:

- 1) Request for regional center assistance to help us identify more PW persons.
- 2) Request that regional centers use the PW CA Foundation as a resource for information on PW - refer inquiries to our group.
- 3) Include PW persons and required programs in their funding needs. Help us establish additional community living facilities in CA for PW persons.

We intend to follow up our presentation with additional contacts at each regional center. We feel we have opened another door for informing more about PW in CA."

Marge Wett also used this presentation while visiting the Colorado Chapter in October. Included with the chapter members was Frank Traylor, Head of Hospitals and Institutions for the State of CO and a representative from a group home located in Montrose.

## WELCOME

PWSA is happy to welcome the 12th Chapter to our organization. The Prader-Willi Syndrome of Michigan has established an active organization in the State of Michigan and officially became our newest chapter in October.

Linda Van Houten has been elected President of the group with Clarissa Miller as Vice-President, Rose Nanzig, Secretary. Their membership already includes 25 persons and couples and they have a very active group working in their state.

We are happy to have another state chapter working on "sharing and caring" campaign for PWS.

## MARK YOUR CALENDAR NOW

It seems the last annual conference just ended but as we move to the end of the year, we know it won't be that long before next year's conference is here again. Mark your calendar now -- JUNE 20, 21, 22, 1985 -- in Hartford, Connecticut. Ask anyone who has attended before, it is an experience that cannot be duplicated.

## SUGGESTION FOR DIETING

A mother writes, "I enjoy reading the GV, I learn a little more and gain more understanding each issue. It can be frightening and somewhat depressing sometimes reading what's ahead, but I want to know everything possible about the syndrome and what lies ahead for our 4-year old Lisa.

Lisa attends school 5 days a week. She truly has received every therapy possible in the 3 years since she was diagnosed. I would like to share some of the things we do:

We eat lots of baked apples. Core, sprinkle insides with cinnamon, nutmeg, and a touch of brown sugar or sugar substitute. Put unsweetened thawed strawberries inside, spoon juice over apple, bake 45-60 mins in a regular oven or a few mins. in a micro-wave.

I keep Lisa's diet high in fibre. These muffins are 39 calories each and are from a recipe in a TOPS cookbook.

1 c. flour	$\frac{1}{2}$ tsp. salt	$\frac{1}{2}$ tsp. baking soda	$\frac{1}{2}$ tsp. baking powder
2 c. bran	2 Tbsp. molasses	$1\frac{1}{2}$ c. water	1 Tbsp. butter

Optional: artificial sweetener or raisins.

Sift flour, add salt, baking soda and powder, bran. Combine with molasses, sweetener, water, melted butter. Add to dry ingredients, stir with a few swift strokes. Fill muffin tins  $\frac{3}{4}$  full, bake 375° 15-20 mins. Makes 24 2" muffins.

Sometimes for a treat I bake: **PINEAPPLE COOKIES**

Mix 1 cup well drained unsweetened crushed pineapple, 1-1/3 c. dry skim milk powder,  $\frac{1}{2}$  tsp. vanilla, sweetened to taste. Drop by tsps. onto teflon cookie sheet. (Optional additions, 6 walnut halves or 1 Tbsp. raisins). Bake 350° 15-20 mins. Refrigerate -- tastes better 2nd day.

We enjoy munching on cucumbers that have been sliced lengthwise, seeds removed, sliced into thinner "fingers" and marinated in a tall glass of vinegar, water, and sweetener.

A lunch suggestion: Hot dog, salad bowl, homemade broth or yogurt. I slice the hot dog lengthwise and actually only use half. Before placing in bun, I break away part of the bun on both sides. (I do the same for hamburgers).

A micro-wave has been a great addition to our house. I no longer start cooking supper at 2:30 PM and very often prepare the meals the night before and just reheat in the micro-wave. When I do not spend as much time in the kitchen, Lisa isn't thinking about food.

Other suggestions: Mix fruit with water or fill the glasses with ice.

Use smaller plates so it looks like they have more.

I find, with Lisa, if I put a lot on the plate with little calories, she is satisfied. I use celery, carrots, salad bowls, soup broth, low calorie breads and cheeses, and yogurt. When these are served as "courses", she even sometimes leaves a little.

Instead of low-calorie jams, I buy overripe fruits in season (peaches, cherries, pears). Stew a bit, add a sweetener, freeze in small plastic containers and serve in place of purchased jams.

## ANESTHESIA REPORT

We have been accumulating statistics about PWS and anesthesia from the questionnaire. To date we have received a return of 173 forms; 18 reporting no surgery, and 155 with information.

Preliminary figures have been compiled with many persons reporting multiple operations and anesthesia. There were few problems or complications. The two most common complications were respiratory infections and sleepiness postoperatively for hours to days.

The most common operations were:

- 1) Tonsillectomy and adenoidectomy.
- 2) Orchiopexy for undescended testes. (Ages ranging from 21 mos. to 27)
- 3) Eye muscle surgery for strabismus. (Ages from 2-14)
- 4) Dental repair and teeth extractions.
- 5) Orthopedic surgery. (Scoliosis roding and fracture repairs, ages 2-32)

We will have more complete information in the future when more data has been accumulated.

*R. J. Wett, MD*

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