

THE GATHERED VIEW

Shirley Neason, Editor

Newsletter of PRADER-WILLI SYNDROME ASSOCIATION

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Box 392
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PRESIDENT'S MESSAGE

We are always very sad to hear about the loss of any of our Prader-Willi people, but it is something that we must expect will occur with somewhat greater frequency than among those of us with better well being. We recently learned of the death of David Mears, the son of Mr. and Mrs. Raymond Mears of Livingston, New Jersey. David was only sixteen years old, and until his appendicitis attack on February 14th, was in good health. He succumbed on March 9th to bilateral pneumonia as a follow-on from massive post-operative infections which resulted from a ruptured appendix. Mr. and Mrs. Mears have been most considerate, especially during this difficult period, to provide us with information related to their son's death that may alert all of us to some special concerns we should have for our Prader-Willi friends. We believe their letter should be of interest to all and have included it in this issue of THE GATHERED VIEW.

I am sure the Mears' tragic loss will touch all of us deeply. We must be thankful, however, that David's parents not only loved him intensely during his lifetime, but that they still care enough to want to help those of us who have so much left to learn about the syndrome. Perhaps through David's passing our own children won't have to view the early twenties as middle age.

Needless to say, we are also most grateful to the Mears for designating the Prader-Willi Syndrome Association to receive donations in David's memory.

Diets are always a fertile subject for any gathering of people concerned about Prader-Willi syndrome. Now some people are not only talking about diets in terms of weight loss, but also in terms of behavioral impact. Because of the Feingold Diet's apparent success in controlling the hyperactive child and numerous other "hyper" and "hypo" conditions, many of us are wondering whether there might be something in it for Prader-Willis. In the Feingold Diet certain food additives, particularly food colorings, are eliminated entirely from the diet. Some of our members are experimenting with it, and it has sparked our interest. We certainly would like to hear from any of you who have had any experience with the diet.

The really good news is that things are looking up at the WISER Acres

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PRESIDENT'S MESSAGE, Continued

group home. We were delightfully surprised to receive our first major donation to help support the special needs of the home. One of our very generous new members, whose son will begin residency there this summer, has donated one thousand dollars to the WISER Acres cause. Additionally, more state aid for the special diets has been approved. Although still not sufficient to cover the special needs spelled out in the last GATHERED VIEW, it is a large encouraging step toward establishing the permanency of this first-of-a-kind group home. This, along with many other smaller donations to the Association, is "making it all worthwhile."

Gane Deterling

ANNUAL BOARD OF DIRECTORS MEETING

The annual meeting of the Board of Directors of PRADER-WILLI SYNDROME ASSOCIATION will be held June 10, 1978, 9:00 a.m., in Room 666 of Moffitt Hospital of the University of California San Francisco. The meeting will include reports from the officers and publications, planning for next year, and reports from Dr. Vanya Holmand and Andrea Netten. Budget and by-laws will be discussed, and board members for the following year will be elected. Members who have opinions or ideas they wish the board to consider, please make them known to the President.

THE MEDIA VIEW

Several orders have come for the parents' handbook, and I know you are anxious for your copies to arrive. The printer called me in today to approve the final paste-up, so it should be soon. I won't give any date, however, as I have found that I had much to learn about how long it takes to get a booklet printed! Please send orders for the booklet to THE GATHERED VIEW, and enclose a minimum donation of \$1.00 per copy. We estimate our cost for printing and postage will be \$2.00 per copy. Approximately half of that has been donated by generous members. If you can send more than the \$1.00 per copy, we assure you the money will be used to help others who have need of our services. We have already received a request for materials in the Spanish language, so here is already a need that will require further funds so we can get some of our basic information translated.

THE GATHERED EXCHANGE, Continued

A new approach was made by a mother in New York:

When my daughter was diagnosed they put her on a 1200 calorie diet, since reduced to 1000. As I had been investigating additives and artificial coloring, etc., with regard to allergies, I used the 1000 calories according to the Feingold diet. I have since noticed that her scratching has been greatly reduced. Also she was restricted on sugar, and when she got hold of a chocolate bar, she came down with a headache which made her feel she had a temperature. Then she got hold of some lifesavers and broke out in hives. Since she has had little sugar, temper tantrums have practically ceased. It could be taking out the additive containing foods plus the limiting of sugar. Since I am an adult onset diabetic (but brittle), I know when my sugar is high or low; my appetite is triggered by the imbalance, and I also get the itchy feeling in my skin.

SINCERE SYMPATHY

Our deepest sympathy is extended to members Raymond and Kathleen Mears of Livingston, New Jersey, in the loss of their sixteen year old son, David, on March 9, 1978.

Mr. Mears wrote the following letter concerning his son in the hope that the information therein would benefit others with the syndrome:

"On March 9, 1978, we suffered the tragic loss of our sixteen year old son, David, who had Prader-Willi syndrome. We hope that the knowledge of the complications which led to his passing, and donations to the Association in his memorium will benefit others stricken with the strange syndrome.

"David underwent surgery for a ruptured appendix on February 14. His apparent weak defense mechanisms, a most probable part of the syndrome, could not overcome the peritonitis. Massive post-operative infections spread through the abdomen, liver, chest cavity, and eventually bilateral pneumonia took his life. An important observation throughout the tragic ordeal was David's extremely high mental threshold for or insensitivity to pain. On only a few occasions did David say he had pain. He usually responded, "Fine", to the question, "How do you feel?" Yet the pain appeared to take its physical toll as he lost his aggressiveness, cheerful mental attitudes, and finally his appetite, but never his love. It appears that this strange reaction to pain is part of the syndrome, and that the mental insensitivity to it increased with age. Certainly it contributed to the tragic ending by overshadowing the extent of serious medical complications, not only to his family, but to the medical profession. Earlier appendicitis detection could have prevented rupture and saved his life. Also it was not until autopsy time that the massive extent of infection through apparent lack of defense mechanisms was discovered. To us an antibiotic is a wonder drug in fighting peritonitis, but not to the unfortunate Prader-Willi syndrome stricken child. Perhaps routine appendix removal should be considered as a preventive measure and treatment for all Prader-Willi children.

We requested that donations be made, in David's memorium, to Prader-Willi Syndrome Association in lieu of flowers. We appreciate the fine efforts of the Association and will continue to cooperate with these efforts for such an important cause."

More than \$300.00 has been received by the Prader-Willi Syndrome Association in memory of David Mears. We express our appreciation to David's family and friends for these funds.

NEW PRADER-WILLI CLINIC STARTED

The Mental Retardation Institute of the New York Medical College in Valhalla, New York, has started a clinic for Prader-Willi clients.

HEY! THAT'S GREAT!

Normally this column is devoted to outstanding achievements by people with Prader-Willi syndrome. However, we recently received news of some outstanding work being done by the mother of a young man with Prader-Willi syndrome, and we wanted to pass the news along. The information is based on an article by Diantha McNerney in the October 24, 1977, issue of the Marathon, N.Y. Independent Villager.

Ruby Stephens of Cortland, N.Y., is president of the board of directors of Seven Valley Shepherds, which sponsors a workshop where young handicapped persons can work in a relaxed atmosphere where they can experience the satisfaction of accomplishment.

The Seven Valley Shepherds began after Mrs. Stephens and other Cortland parents attended a workshop conducted by Lambs, Inc., of Libertyville, Illinois. The Lambs began as a pet shop, and was so successful that it relocated to a farm, where it expanded to include a restaurant, bakery, gift shop, and farmers' market.

While The Lambs has a residency program in addition to its day workers, Seven Valley Shepherds has not yet grown to that extent. However, the parents hope that eventually there will be a residency program, with a special wing for residents with Prader-Willi syndrome and other dietary problems.



Mrs. Ruby Stephens consults with a worker at Seven Valley Shepherd in Cortland, N. Y.

Before the workshop began, Mrs. Stephens had been working in ceramics with her son, Billy. Someone donated a kiln for his use, so with this equipment and background, it was only natural that the Seven Valley Shepherds should begin with a ceramic shop, which soon branched out into the related craft of macrame. In addition to selling the wares crafted in the shop, Seven Valley Shepherds also offers classes in ceramics and macrame to the general public.

"We believe that the mentally retarded can develop beyond their current functioning level if they are assigned tasks beyond the routine," says Mrs. Stephens. "The assumed responsibility, the public contact, and the work variety all demand and develop the latent potential of the mentally handicapped. Individuals can achieve a composed outlook on life, devoid of fear and hostility, which is indeed mentally and emotionally healthy."

WEIGHT REDUCTION PROGRAM FOR CALIFORNIANS

The Clinical Research Unit of the Camarillo-Neuropsychiatric Institute Research Program, Camarillo, California, has developed a successful weight reduction program for adolescent and adult patient/residents diagnosed as Prader-Willi syndrome. They would like to obtain information concerning patients/residents who might be appropriate candidates for such treatment. These patient/residents would have to be eligible for admission to Camarillo State Hospital through the Short-Doyle Program. If you can provide any information concerning such patients/residents, contact B. D. Marshall, Jr., M.D., Supervisor, Clinical Research Unit, Camarillo-Neuropsychiatric Institute Research Program, Box 'A', Camarillo, CA 93010.

GATHERED REPORTSSeattle, Washington

Seattle parents met in April on the regular clinic date at the Child development and Mental Retardation Center of the University of Washington. Center staff members Steve Sulzbacher, Ph. D., Jude Leconte, M.S.W., Alice Lobenstein, R.N., and Nancy Couhig, R.D., were present to answer questions.

One parent brought up the need for social training in the school to help the children learn appropriate social behavior. Dr. Sulzbacher will write a memo to teachers to let them know what kinds of things they can do.

Another parent expressed concern that siblings often have difficulty ignoring inappropriate behavior. A suggestion was made to reinforce mature behavior. One parent expressed frustration that there were so many problems she didn't know where to start, and it was difficult to sustain the effort to follow through on behavior modification. Judi suggested that families begin by working on behavior that most irritates the most members of the family and all work together to replace it with desirable behavior. She volunteered to write a plan for helping the family decide what behaviors they want to work on (see THE PROFESSIONAL VIEW.)

Several mothers indicated that their children seem to have a problem knowing how to appropriately open a conversation with acquaintances or strangers. Some suggestions to deal with the problem were to help the child memorize a few appropriate phrases to use in opening conversations, to give him cue cards to carry in his pocket, and to role play with him. It was emphasized that we need to stroke appropriate behavior initially in order to encourage it. It was also suggested that the child be given feedback from friends so he understands how others view his behavior (this applies especially to positive feedback.)

Dr. Sulzbacher commented, "Remember that progress becomes the next problem." He went on to cite examples of how the child may learn to initiate conversations, only to run into new problems of knowing how to listen or when to stop talking. Parents need to be prepared to deal with these new problems, remembering that they represent progress.

Judi made the suggestion that family members and others who are working on behavior problems need to be rewarded themselves at intervals for the efforts they are putting forth.

Another parent brought up the problem of sores created when the child picks at his skin. Substitute activities were suggested, such as squeezing a ball (which is good exercise for the hands in addition) and playing with string.

The matter of organizing formally as a parent group was brought up, and Jane Schmelzer volunteered to look into the legal requirements and bring a recommendation at a later meeting. The consensus was that organization would help us to more effectively support the work of WISER Acres and Double R R Ranch.

Nancy gave parents some low-calorie recipes to take home. (These recipes will be published in THE GATHERED VIEW at a later date.)

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* 392, Long Lake, Minnesota, 55356. Editorial material may be sent to THE *
* GATHERED VIEW, 147 South 294th Place, Federal Way, Washington 98003. *

THE PROFESSIONAL VIEW

We have two articles in the PROFESSIONAL VIEW column this issue: an article on behavior management by Judi Leconte, M.S.W., and an interview with Peggy Pipes, M.P.H. Both are on the staff of the Child Development and Mental Retardation Center of the University of Washington, Judi as a social worker and Peggy as a nutritionist.

Working with Behaviors in a Family Setting, by Judi Leconte, M.S.W.

Behavior management in a home setting is extremely difficult due to 1) the amount and variety of activities acquired by various family members (particularly parents), 2) the lack of predictability in individual schedules, and 3) individual variance in what "bugs" you most. These are just three of many factors which make home behavior management so exhausting. One way to eliminate some barriers to behavioral change is to hold a family meeting to discuss problems and find workable solutions. This is not an original or new idea. Several family therapists have supported and written about this idea (i.e. Family Council, Rudolf Dreikurs, Shirley Gould, and Raymond Corsimi, 1974, Henry Regnery Co.; Families, Gerald Patterson, 1971, Chapters 7-9.) Prior to the family meeting, it will be helpful to go over the following questions to clarify your definition of the problem, how to change it, and who will be involved.

I. THE BEHAVIOR

1. What is it? Be specific!. (i.e. "He looks at me funny" is not specific. "He stares at me for five minutes or more" is specific.)
2. How many times does it happen in a given day and for how long?
3. Is it public or private?
4. Who is present when it occurs?
5. Who does it bother most? Least?

II. SOLUTIONS

1. How many ways can you think of to change the behavior? (Again, be specific.)
2. For each way you think of, answer the following:
 - a. Who in the family has to be involved in the program to make it effective?
 - b. How much time will it take from each person? For how long?
 - c. Who will get "less done" from those family members involved in the program? How much less?

The family meeting needs organization and this is usually supplied by the member presenting the problem (usually parents). All of the above should be worked out in advance of the meeting.

At the meeting, be open to new solutions based on various family members' responses to the following questions:

1. Who is interested/concerned about this problem? How concerned? A lot? A little?
2. How many family members are willing to be involved in a program of change? What, specifically, is required of each person?
3. Who (how many individuals) will get less attention from other family members, and how will they feel about getting less? (i.e., if the behavior occurs, attending to it will have priority over someone else's need.) What would they be willing to give up? For how long?
4. Find ways to work through someone feeling left out by setting up agreements for time use.
5. Agree on a time frame to try the new program and evaluate it.

THE PROFESSIONAL VIEW is continued on Page Seven

THE PROFESSIONAL VIEW, Continued

Peggy Pipes, M.P.H., responded to questions readers have asked.

QUESTION: What is the Feingold diet?

PEGGY: The Feingold diet was developed in response to the hypothesis that food additives are the cause of hyperactivity in children. Since then, well-developed studies have shown a limited response to the diet. Those children whose response to certain additives is pharmaceutical will always respond within three hours after eating a food to which they are sensitive.

QUESTION: Does sugar in the diet cause behavior problems in children with Prader-Willi syndrome?

PEGGY: The experience in our clinic is that reasonable intake of sugar appears to cause no problem. An excess of sugar, independent reports indicate, causes deterioration of behavior.

QUESTION: How much sugar, then, is appropriate in the diet of a child with Prader-Willi syndrome?

PEGGY: Sugar is so much a part of our culture that I would not recommend its total elimination. However, it must be remembered that the nutritional needs of a person with Prader-Willi syndrome are the same as those of other people. If the quantity of sugar is not limited, he will inevitably become obese, or his diet will not be adequate because he will not consume enough of the high nutrition foods.

QUESTION: Have children with Prader-Willi syndrome been checked for hypoglycemia?

PEGGY: The children have been checked for hypoglycemia and found to be normal.

QUESTION: Does diabetes occur frequently in people with Prader-Willi syndrome?

PEGGY: Diabetes is common in people with Prader-Willi syndrome who are obese, but has occurred less in those whose weight is under control. When it does occur, it usually appears during or after adolescence.

THE GATHERED EXCHANGE

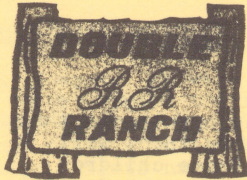
A comment on "food stealing":

Our son is five and looks forward to his meals, but does not steal food. I keep things off the counters, but do not have to lock things up. He lost 27 pounds from October, 1976, to August, 1977, and has maintained his weight well. He is a very handsome boy, I might add. I do give him sweets occasionally by working them into his diet. Possibly that gives him a less desire to steal--I don't know. Possibly I will have more of a problem as he gets older and is away from us more.

A question about speech problems:

I would like to see more in your newsletter about the speech problem Prader-Willi kids have. Our boy has been very delayed in speech and therapy is helping. Do other parents have this problem?

Continued on page two



Double R R Ranch was established specifically to meet the special needs of individuals with Prader-Willi syndrome who require a controlled environment and limited caloric intake if weight reduction is to occur and be maintained.

Double R R Ranch serves children and adolescents 6-18 years of age. Please contact the Director for information regarding respite for persons over 18. Individuals with

other developmental disabilities will be served when obesity is a problem, on a space available basis.

Double R R Ranch also provides a residential component for children, adolescents, and young adults. Individuals under 21 attend public schools. Individuals over 21 attend a vocational evaluation and training program in the community.

Diets are designed based on the caloric needs of each individual and in conjunction with the interdisciplinary Prader-Willi team at the University of Washington, Seattle.

Parental respite services include consultation with a professional multi-disciplinary Prader-Willi team, arranged with adequate advance notice. Vacation and tour arrangements are available.

For further information, write to: Director, Double R R Ranch, P. O. Box 578, Bothell, Washington 98011.

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