THE GATHERERED VIEW

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Newsletter of PRADER-WILLI SYNDROME PARENTS AND FRIENDS
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MESSAGE FROM THE DIRECTOR

In 1956 Drs. A. Prader, A. Labhart, and H. Willi first described the syndrome we now know as Prader-Willi syndrome. To many of us it seems that not much has transpired in the last twenty-one years to give us a better insight into the cause and treatment of the syndrome. For one thing, there was very little mention made in any easily accessible literature for the first twelve years after it was described. To our knowledge it was nearly twenty years before the first mention was made in any general interest publication (NEWSWEEK, October 13, 1975- "The Glutton Child.")

A lot has transpired in the last twenty-one years, however, that has not yet been publicized. The number of people working on solutions to the Prader-Willi problems has doubled, I'm sure, if not increased ten-fold in the past ten years, and more and more lay people and professionals are becoming involved each year. I have no absolute numbers, but from the communication we receive, I have reason to believe the number of people involved will double again in the next two years.

What I have been trying to describe is the "rolling snowball effect." It often takes time to get one started, but once it starts rolling, it picks up speed and size at an accelerated rate. The Prader-Willi syndrome effort, in my estimation, just got started about four years ago, and it is just about ready to start rolling. It would be presumptuous to try to predict the result of this effort, but one thing is for certain, something better is bound to result from it.

When I consider how long it has taken to get the overall Prader-Willi effort started, I don't feel quite as frustrated about our Parents and Friends organizational activities not being as advanced as I hoped. I am, however, disappointed that some of the more tangible items that we promised in previous newsletters would be available by now, have not yet come to pass. I can only assure you that we have merely been delayed. We will soon have some additional publications, a good bibliography, state-by-state membership lists and other worthwhile material.

Many of you have already received a copy of the Prader-Willi Syndrome Questionnaire sent out by Dr. Vanja Holm's office at the Child Development and Mental Retardation Center. We encourage everyone to respond quickly to the questionnaire as it can only serve to benefit us all. If you did not receive a copy of this questionnaire, please either contact CDMRC or let us know here at our home office, and we will try to arrange for one to be sent to you.

For those of you who are new members and anyone else who has a need, we have extra copies of all of our back issues of THE GATHERED VIEW, and will forward them to you free of charge as long as the supply holds out.

(Continued on page two)

MESSAGE FROM THE DIRECTOR, Continued

The promised article in GOOD HOUSEKEEPING magazine has just come out in the May issue. It is entitled "The Eating Disease". The author expects to wri a future piece with a mention of our organization.

Gene Dabeling

A VIEW OF WHO'S WHO

This column receives the most material from our subscribers. If you se your story, and it has not yet appeared, be patient. Someday there will surely be room for it.

Our first story this month is from a Texas mother.

I had written asking if you knew of anyone in the Dallas-Fort Worth are who knew about Prader-Willi syndrome. Since you did not, I made phone calls, e found that Children's Medical Center in Dallas was the best place. Dr. Peter S Sherrod, Assistant Director of the Birth Defects Center, confirmed our suspicio that Joy does, in deed, have this syndrome. Because approximately 30% of patie with Prader-Willi syndrome develop diabetes in their teens, Joy was given a glu cose tolerance test and found that her curve is compatible with a pre-diabetic curve, and may be related only to her gross overweight--at that time (July, 197 130 pounds. There were students as well as doctors asking questions and doing various tests, x-rays, etc. along with the dietician playing around with her food, until finally she was on 500 calories. All of this, along with the fasti for the glucose tolerance changed Joy from a lovable sweetheart they had been having so much fun with into a screaming, fighting, wild person of whom they we afraid. I was terrified to find that even in a hospital there is nothing that be done during these times, and that I am the only one who could do anything wi her until it passed.

After we returned home, Joy started taking ritalin. She started with 1 daily and worked up to 40 mg. It has curbed her appetite, and at times seems t open new channels of reasoning.

I felt that the 500 calorie diet Joy was on in the hospital was not som thing we could work with effectively at home, so we joined Weight Watchers agai I knew it would work because Joy had lost sixty pounds before on the program, b because we did not know the origin of her problems, she gained it back. Since person with Prader-Willi gains weight on what a normal person loses on, Joy can only have half of the food allowed on the youth program. I discovered this the first time she was on the program, before we heard of Prader-Willi. She has lo 35 pounds. We are so thankful and proud of her. It is so hard for a child, especially one with so many other problems. We have a reward system to ease th burden, because she deserves every help possible.

I am interested in corresponding with someone from Texas. Surely we ha people in this area with Prader-Willi syndrome who would be willing to organize as a group so our children can benefit from educational and recreational progra that children in California and Washington enjoy. I want to thank other parent for sharing with me through this newsletter their most private affairs and feel of their children. It has helped me understand Joy better.

Mrs. Vernie Gardner, 1216 Olen Lane, Bedford, Texas 76021

A VIEW OF WHO'S WHO, Continued

Another story is from a mother in England.

My son, Joseph, was born 6th August 1970. He was overdue, but had signs of prematurity. He was unable to suck or maintain normal temperature. The first week he was kept in an incubator and fed through his nose. He got an infection, and was on anti-biotics. His breathing was shallow. He recovered and came home at six weeks. He could have come home sooner, but I had two small children at home, and the hospital did not think I would be able to cope.

I fed him with a flexible bottle and squeezed milk into him; he swallowed well. He did not suck until eight months. At two and a half months I started mixed feeding, because he was not putting on weight. He took to this well, and had no difficulty chewing. Even at this age he preferred sweet food rather than savoury. He could not hold his head up until he was ten months, and he sat up at twelve months. At fourteen months he was shuffling on his bottom. When he was eighteen months my health visitor took me to a physiotherapist who advised me to teach him to crawl. I did not realise how important this stage is. Within two weeks he was crawling and at twenty-three months he was walking. He has been talking in his own way for years, but it is only in the last year that we have been able to understand him. Speech therapy has helped. When my three-year-old, Michael, started talking, so did Joseph. He is still improving.

He could understand what was said to him from an early age, could finger-feed himself from twelve months, and at that age could unscrew things.

He has a small penis and no testes. He is overweight, weighs 4 1/4 stone. (A stone is a British unit equal to fourteen pounds--Ed.) He was underweight until age three, then put on weight rapidly. He was diagnosed at age four.

He is about average height. His hands and feet are small, but his feet have grown since he has become more active. He would not walk any distance when he was three, but due to the efforts of his teachers at Special School, who take the class for regular walks, he now walks well. I feel that because of the lack of muscle tone, exercise is important. Joseph's legs were very small, but since he has been walking more, his legs have grown and developed. He now climbs, jumps, and walks on his toes. Joseph enjoys swimming. He goes with the school, and we take him in summer to the local swimming baths and the sea.

I have four children, three of which are normal. Eileen is eight, Teresa seven, Joseph six, and Michael three. I find it difficult to control Joseph's eating habits. He has, however, managed to maintain the same weight for a year, although he is growing rapidly. This is due to advise from my health visitor and doctors at the hospital where he goes for yearly checks. Obesity produces clothing problems. I cannot buy trousers to fit him, so I make them. I could have them made, but this would be more expensive.

He has been attending Special School since he was three. In this country there are the normal schools, the E.S.N. (educationally sub-normal), and the Special Schools. Joseph was top of his class when he was five and was transferred to the E.S.N. school. He found it more demanding and did not fit in with the other children well, so he was transferred back to Special School, where he is happier. His speech improved and he mastered dressing and undressing. He tends to be lazy and can often do more than he actually does.

He has strabismus in one eye, but only squints when tired or excited. He has reasonable control of his eye, so does not need treatment at the moment. He visits the eye hospital for yearly checks. My husband's brother had an operation for a squint when he was young, and we thought it was just a family weakness.

Joseph has sensitive skin. We have to be careful of sunburn in summer. I have also noticed that he will not get into his bath unless the water is lukewarm. He was so pale as a baby that the hospital tested his blood, but he was not anemic. He has blue eyes and fair hair as well as fair skin. He is generally a happy child. Recently he has been having more tantrums than usual, but gets over them fairly quickly.

GOOD NEWS ABOUT THE BOOKLET!

It appears that money is now available for the printing of the proposed booklet for parents of children with Prader-Willi syndrome. One of our members, W. J. Russell, whose grandson has Prader-Willi syndrome, has offered to loan $500.00 interest free, to be repaid as the booklets are sold. In addition, we have the promise of a donation of $113.00 from a memorial fund.

Several have read the manuscript and made suggestions. I am currently re-writing according to these suggestions. I will try to get this done in time to get the manuscript to the printer in July. Hopefully, by the time you receive your September GATHERED VIEW, your copy will be ready and waiting for you to send for it.

GATHERED NEWS

This begins a new column to help us keep up with what the news media reports, not only about Prader-Willi syndrome, but other areas that might be of interest and aid to parents and others. Sources for the column are a clipping service provided by the President's Committee on Mental Retardation and articles forwarded by members. Special thanks this month to Martha Leyshon who sent several items.

A respite care program has been initiated in a three-county area in Oregon. Care providers are trained, who then care for handicapped children for short periods of time in either the child's or their own home so parents can go on vacation or just get a period of relief from the problems of day-to-day care of a handicapped child. from the Beaverton, OR, Valley Times

In Pennsylvania, a horticultural training center has been opened for mentally handicapped adults. The students operate a retail garden shop where they sell flower arrangements, hanging baskets, and Christmas trees. In addition they receive basic educational skills and instruction in the activities of daily living. from the King of Prussia, PA, Today's Post

The United Nations General Assembly has proclaimed 1981 as the International Year for Disabled Persons. from International Rehabilitation Review

A strain of mice with a disorder similar to Prader-Willi syndrome has been developed at Chicago Medical School, and it is hoped that studies on these animals will lead to discovery of a drug that might help control appetite. from the Chicago Daily News

DID YOU MISS AN ISSUE?

THE GATHERED VIEW is a volunteer operation, run from the editor's den, which is also used for many other activities. Consequently, it is quite possible that errors can occur which result in some subscribers not getting their copies. If you miss receiving any issue, let me know, and I will get it to you. THE GATHERED VIEW has been published every two months since July, 1975.

GATHERED REPORTS

Seattle, Washington

Seattle area parents will meet on Thursday, May 26, at 7:30 p.m. at the Child Development and Mental Retardation Center of the University of Washington. Margo Thornley, Director of the Double R.R. Ranch, a recreational and respite residence for children with Prader-Willi syndrome, will be the speaker.

THE BOOK VIEW

No book review this month, but comments on two recent articles.

"The Eating Disease," by Helen Bottel. Good Housekeeping, May, 1977.

This article is factual and well-written. There is less exaggeration than has appeared in some popular magazines, although the familiar anecdotes seem to become a little larger with each telling. Best of all, the article has an optimistic note, telling what can be accomplished by diet and other therapy.

"Metabolic Aspects of a Protein-Sparing Modified Fast in the Dietary Management of Prader-Willi Obesity," by Bruce R. Bistrian, M.D., Ph.D., George L. Blackburn, M.D., Ph.D., and John B. Stanbury, M.D. The New England Journal of Medicine, April 7, 1977.

This article will be of interest to nutritionists and physicians working with patients with Prader-Willi syndrome. Parents who wish to read the article need to understand that this diet was undertaken only under strict medical supervision and should not be attempted otherwise.

THE GATHERED EXCHANGE

Information concerning DuPont Institute:

We took Caroline to the Dupont Institute last fall to have Dr. Dean MacEwen operate on her dislocated hip. The surgery was successful and I just cannot say enough nice things about the Institute. It was inconvenient hauling her cross-country in a body cast on a plane, but the results were worth it. They were very helpful to me in the dietary aspects, too, since she had done her previous gaining in body casts. I would recommend them to anyone who lives in that area, and Dr. MacEwen deserves every bit of his international reputation.

A. P., Texas

Can someone share information with this mother?

We are fighting a losing battle with our daughter's (age 25) weight problem. She recently left a community living facility to return to the state hospital for retarded because they could not provide staff to handle her eating problems. Even at the hospital she is the only Prader-Willi, and they do not operate a cottage with enough food security for her. As she has continued to gain they have encouraged us to consider the gastric bypass. In trying to learn more about this procedure, I have heard glowing reports, also discouraging reports. I wonder if through the newsletter I could reach any parent whose child has had or will be having this surgery. I called Dr. Holm at the U. of Washington, and she said she was aware of maybe two that had this surgery, but did not have names or addresses. I would certainly appreciate any first-hand experiences that might help us decide. Thank you.

Mrs. H. L. Olson

Comment from the editor:

In the May issue of Good Housekeeping's regular medical feature, "Family Doctor", the pros and cons of this surgery are discussed. The syndicated newspaper column on nutrition by Drs. Jean Mayer and Johanna Dwyer also recently discussed this surgery. They recommended that the person who is contemplating bypass surgery consult an internist and a psychiatrist in addition to a surgeon.

THE GATHERED EXCHANGE, Continued

Another parent is concerned about her daughter's future:

> Joy will be 16 in August, and we, like the rest of your families, are concerned for her future. Do you feel the same as we do that there is no place that meets the needs of people with Prader-Willi? We have found that the so-called "controlled environment" that the state schools afford is not what we feel we would want for Joy in case something happened to us. Please inform us if any of you have successfully found any place that we could look into for our needs.
>
> Mrs. Vernie Gardner

I would like to add to Mrs. Gardner's request:

> I have heard from enough parents concerning this problem to know that it is a concern shared by many. Who can suggest concrete steps we can take to get something started to meet this need? If you have suggestions or information on the subject, please send it to THE GATHERED VIEW.

Here are some exercises that will help improve coordination:

> Suspend a rubber ball from a ceiling or doorway. Have the child practice hitting it with an old broom handle. Mark the broom handle with colored bands to be used as targets.
>
> Provide balls of different textures, shapes, sizes, and weights for the child to play with. Make yarn balls in addition to the ones you buy.
>
> Place a rope flat on the floor, and let the child walk along it. After he has gained skill, arrange the rope into shapes, numbers and letters.

Here is how to squeeze as many calories as possible out of ground meat: After browning ground meat (with no fat added), pour off the fat that cooked out of the meat, and squeeze the meat in paper towels to get rid of as much fat as possible.

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