

THE GATHERED VIEW

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Federal Way, Washington 98003

Newsletter of PRADER-WILLI SYNDROME PARENTS AND FRIENDS

Post Office Box 124
Harvard, Massachusetts 01451
Gene Deterling, Director

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PRADER WILLI SYNDROME PARENTS AND FRIENDS

Prader-Willi syndrome is a rare birth defect that results in initial hypotonia (lack of muscle tone) and later a compulsive tendency toward obesity as well as other complications. Because of the rarity of the affliction, there is much to be learned about its treatment and perhaps eventual prevention. The non-profit organization, PRADER-WILLI SYNDROME PARENTS AND FRIENDS, has been formed to provide a vehicle of communication for persons who have had experience with the syndrome and those who need to benefit by others' experience.

A one-year membership may be obtained by sending \$5.00 along with your name, address and telephone number to PRADER-WILLI SYNDROME PARENTS AND FRIENDS, Box 124, Harvard, Massachusetts 01451.

MESSAGE FROM THE DIRECTOR

We are beginning to see signs that allow us to be cautiously confident that there is progress being made toward a better understanding of Prader-Willi syndrome and the methods of coping with it. A year ago most of us were not aware of anyone who had any dedication to resolving the many problems associated with the syndrome. Today we are happy to report that we have a number of practitioners and other professional people who are deeply interested in working toward solutions to these problems. Perhaps the recent publicity in Newsweek and Today's Health have helped to inspire new interest, and perhaps our organization has helped. There were, however, a number of dedicated people working with Prader-Willi syndrome citizens prior to these articles and prior to the formation of our organization. We just didn't know about them. Now that we have a vehicle for communication we hope to be able to keep everyone apprised of what is happening.

Our organization now has over a dozen members associated with the medical profession. We receive correspondence every day from others. Those who reside in the California area should be pleased to know that we have just been informed of a program to explore the underlying disorder in patients with Prader-Willi syndrome at Harbor General Hospital in Torrance. Included in this newsletter is a letter from Dr. George Bray describing this program.

As a service to all our members we would be willing to supply the names of members in your local area who might be willing to work with you toward the formation of a local or state group. In some states we now have at least fifteen members.

What should also be of interest to all is that we are collaborating with Dr. Vanja Holm at the University of Washington on the preparation of a questionnaire that will be issued by Dr. Holm to our membership as well as other interested persons. The questionnaire should provide valuable information on various aspects of the syndrome, and through our newsletter we will report on the results.

One of our young college student members is preparing an independent study on Prader-Willi syndrome, including the social and psychological aspects. He has already compiled one hundred and thirty references. We hope to have more to report on this in future issues.

Let us keep working together.

Gene Deterling

REVENUE/EXPENSE STATEMENT - December 31, 1975 - PRADER-WILLI SYNDROME PARENTS AND FRIENDS

Membership Dues:		Expenses:	
53 One-year U.S. memberships @ \$5	\$265.00	Forwarded for publication. . . .	\$223.00
1 Three-year U.S. membership @ \$15	15.00	expenses	
1 Two-year overseas membership @ \$12	12.00	Postage.	38.00
Total membership dues	<u>\$292.00</u>	Office supplies.	37.66
Susan Leyshon Memorial Fund	\$428.00	Telephone.	18.68
		Miscellaneous.	9.50
		Total expenses	<u>\$326.84</u>
Total Revenue	\$720.00	Cash on hand	\$393.16

Gene Deterling, Director

REVENUE/EXPENSE STATEMENT - December 31, 1975 - THE GATHERED VIEW

Receipts:		Expenses:	
Received from membership dues	\$223.00	Mimeograph supplies.	\$ 11.34
Donation*	21.00	Postage.	15.50
Supplies sold*.	1.25	Printing	24.22
Total receipts	<u>\$245.25</u>	Bank charges	7.99
		Total expenses.	<u>\$ 59.05</u>

* The donation was money received in payment for taking my child to the University of Washington to participate in a research project. Since the amount more than covered our actual travel costs, we preferred to donate it to THE GATHERED VIEW. The supplies sold were mimeograph supplies, which are no longer needed since we changed to printing.

Shirley Neason, Editor

LETTER FROM DR. BRAY

Prader-Willi Syndrome Parents and Friends
 P. O. Box 124
 Harvard, Massachusetts 01451

Dear Mr. Deterling:

We have just discharged the sixth patient with the Prader-Willi syndrome from our Ward for Clinical Investigation. In interviewing this patient's relatives, we heard of your organization and were interested in learning more about it.

As a background, let me tell you about the program for evaluating patients with the Prader-Willi syndrome at the Harbor General Hospital. My major interest over the past ten years has been the problem of obesity in its many forms. Upon moving from the New England Medical Center to Los Angeles, one of my pediatric colleagues and I began to develop a program to explore the underlying disorder in patients with Prader-Willi syndrome. We have approached the problem from a number of directions, including metabolic studies, endocrine studies, psychological studies, studies of eating control and developmental studies. We would be most interested in helping members of your group who may reside in California as well as making your existence known to the patients in this area.

Yours sincerely,
 George A. Bray, M.D.
 Professor of Medicine, UCLA School of Medicine
 Director, Clinical Research Center,
 Harbor General Hospital

This is the story of Christopher, written by his mother, Ann Watson, of Mansfield, Nottingham, England. Mrs. Watson wrote this portion of her story in 1972. In 1975 she wrote another chapter to update her story. The 1975 chapter will be printed next issue.

Christopher was born on a wet Wednesday evening in February, 1967. It was quite a difficult birth, but baby seemed all right after his first struggle for life. Christopher was my first child, so I was like all new mums, not quite sure what progress baby should make. I took him to the clinic every Wednesday afternoon to have him weighed and chat with the other mums about our children's progress.

My first little fear began when Christopher was six months old and wasn't making any attempt to sit up. I was told not to worry; he would sit up in his own time. As he was rather a plump baby, I accepted this. He had two teeth by the time he was five months old, which was quite normal, so I tried not to worry so much. Time went by with me having my little doubts, but trying to convince myself that Christopher was all right.

Then came the nightmare day when Christopher was one year and two weeks old. Quite a normal day to begin with, but come lunchtime he didn't want any dinner which was very unusual. But he seemed all right, so we went to visit a friend. There he sat on the floor and looked round, which now I can see was not right. On the way, I thought he had fallen asleep, but on arriving home, I could see something was wrong. He was drained of all colour. I took him out of his pushchair, and called in a neighbour whilst I went for the doctor. By this time he looked awful, and was trying to be sick, and his eyes rolled in a peculiar way. Doctor could not come, so the receptionist told me to take him down. I had to wait until 4:40 when my husband came home to take us. When we arrived, we were told to wait; Doctor wouldn't be long. Christopher seemed to be getting worse. He just lay in my arms like a waxen figure until the Doctor saw him. The doctor sent him to the hospital immediately.

It was virus pneumonia. I was told later by the welfare visitor I was very lucky not to have lost him, and that he must have a good resistance. He was in the hospital one week, and after he came home he stayed in one room and no one else was allowed in the house.

After this we settled down to what I thought would be life as it was before, but this wasn't to be. Christopher would scream, and wouldn't go into his cot; he didn't seem to want much sleep. I couldn't bathe him; he would go stiff and wouldn't let me put him in. This went on for over a year. I would get up several times in the night; I would even bring him down and let him play. The doctor gave me the usual syrups to make him sleep, but they never did. I was also told not to let him cry, but not why; I later learned it was because he might have a fit.

Christopher wasn't making any attempt to either crawl or walk. When I went out, people asked me if he was walking yet, and when the answer was "no," they would say, "Never mind; there is plenty of time." Yet I could see they didn't think he would. I stayed in more and more until eventually we didn't go out much at all.

Since coming out of the hospital Christopher had been having what I called the jumps. It was very frightening at the time, but I gradually got used to them. I found that if I comforted him he would go back to sleep. This occurred quite a few times during the evening and night. Also he was having very slight attacks during the day which only lasted seconds. I seemed to be the only one who saw them, so again I was told I was worrying too much. Finally I just couldn't take any more and persuaded my husband to go to the doctor with me, and we were told these attacks were petit mal, a mild form of epilepsy. Apparently the doctor had known this, but didn't want to worry me. If only he had realised I had worried more through not knowing. Bathing was still an awful experience. In the end I gave up and gave him an all over wash. Gradually I started to take him in the bathroom with me until he grew more confident and would get in the bath with me. Then after a while I started to get out and leave him for a few seconds until he went in again by himself. All this took a very long time.

Christopher was two years and two months old when he took his first faltering steps. I really began to think then that things were not as bad as I had first thought. Then four months later he broke the femur of his left leg, for which he spent three weeks in traction. The nurses were very kind to him, although if they got too close he would pull their hair or scratch them. He had been doing this for some time, particularly to other children. I was told this was frustration because he couldn't get about and do what others were doing.

A VIEW OF WHO'S WHO, Continued

The day he came off traction was Christmas Eve. His legs were taken down, and he was taken to the plaster room to have his leg put into plaster so he could come home for Christmas. He cried all the time, and the Sister said he wanted a good spanking; there was nothing for him to cry for. He wasn't a mardy child. He didn't cry when he broke his leg, but his back must have been sore when his legs were taken down, and he was frightened. I was so upset; I told the Sister on the ward when we got back. She said it would do her good to work on the children's ward for a while. I hope this never happens as she would probably make the children unhappy.

Just before this, Christopher had started Playschool. The welfare visitor thought it would help him mix with other children, as he would sit in a corner when children came to play with him. It was Easter before he was able to go again. He was still in nappies when he went out because he wouldn't tell me when he wanted to wee. The supervisor at playschool told me to take him in pants, and she would see if he would copy the other children and go to the toilet. This wasn't successful for many months, but eventually he did tell her sometimes, which was a great improvement. He still wets himself once or twice a day and wets the bed.

When he first went to Playschool he painted the other children if he had the chance, and knocked things over. By the time he was five he was just starting to try to do the same as other children, although he was still quite limited with hands and legs. There were a lot of things he couldn't do. The Supervisor and her helpers spent a lot of time and patience trying to teach him.

All this time Christopher was visiting the doctor for the usual children's ailments. At one of these visits I asked about his little tail which seemed very small. I also couldn't find his testicles; apparently he has one, but the other hasn't come down. Again I was told there was nothing to worry about; his tail just looked small due to his having a lot of fat.

Then came time to start school. He went Wednesday afternoons for three weeks. I told the headmistress a little about Christopher. I was told not to worry; I think she got the impression I didn't want him to go to school. The first afternoon I was told he was perfectly all right; he had enjoyed himself.

He started full time after Christmas just before he was five. One day I was told by the headmistress that the health authorities had been informed that they wouldn't be able to keep him. He was wetting himself, painting other children, and pulling everything to pieces. She asked why I hadn't said anything about this as I must have known what he was like and shouldn't have put his name down for ordinary school. This really hurt me as I had tried to tell everyone for so long that there was something wrong.

Things really moved after this. The health visitor came to find out all the things Christopher could and could not do. An appointment was made for us to see the specialist at the local hospital. She told us at once what was wrong. Apparently his condition is very difficult to detect, as so many different things go together and each case is different, but the doctor had worked under the professor who discovered this. We were lucky to meet her, or we would probably still not know what was wrong.

Christopher was then assessed to see which school would be most suitable for him. He was assessed as having a mental age of two, which I thought was wrong as a lot of the things he was asked to do, he wouldn't do for the doctor, but could do quite well on his own. It was decided he would go to a new school just opened for educationally sub-normal children.

He was taken on trial for six months. If no improvement was made, he would have to be moved to another school.

We have reached the end of the six months, and although we haven't been told officially, the teacher does not think he will be able to stay, as he hasn't made any progress academically, although socially he has improved a lot. Although we expected this, we feel the change will upset Christopher. After only one and one half days at the infant school, he wanted to know why he couldn't go with his little friends any more; children he had grown up with and started school on the same day.

Christopher now has a brother ten months old. He is living parts of the babyhood he missed through Neil. He crawls with him, and plays with his rattles, which he never did when he was a baby himself. We now have to wait and see just how much Christopher progresses. His complaint is so rare no one seems to be able to tell us what to expect in the future.

THE PROFESSIONAL VIEWDental Care

Do children with Prader-Willi syndrome generally have poor tooth enamel? In a recent talk to the parent group in the Seattle area, Dr. Richard Rolla, dentist on the Prader-Willi team at the University of Washington Hospital's Child Development and Mental Retardation Center, challenged this widely held belief.

Dr. Rolla reports that he has found good solid enamel in the teeth of most people with Prader-Willi syndrome that he has examined. The problem that does exist is that, in spite of the good enamel, the teeth show a consistent pattern of wear greater than that of the average population.

Why this wear pattern? Dr. Rolla stated that he would like to examine a large number of people with Prader-Willi syndrome to see if he can find the answer to this question. He would like to examine these individuals at different ages to measure their growth pattern, and to see if he can find a consistent pattern of problems such as the lower jaw being too far forward or an unusual shape of the mouth.

Dr. Rolla emphasized that when decay does occur in the teeth of children with Prader-Willi syndrome, chances are it is the usual garden variety of tooth decay. This good news for parents was followed by his expert advice on how to care for their children's teeth.

The first step, Dr. Rolla advises, is to select a good dentist and begin taking the child to see him as early as three years of age. Select a dentist who not only has good professional qualifications, but who will make dental care as pleasant as possible for your child. If you find that a mistake has been made in your choice of a dentist, don't hesitate to seek out a different one.

Most important to cavity-free teeth is removal of dental plaque. The parent needs to perform this chore until the child is old enough to be taught to do an adequate job for himself.

Select a soft toothbrush. Place the toothbrush at a 45° angle to the gumline, and brush with a vibratory motion. Turn the brush on end to clean the inside of the front teeth. Regularly at least once a week--use a food stain on the teeth to reveal plaque that has been missed in brushing. Brush away this revealed plaque until there is no longer any stain. If your child is not old enough to use the tablets sold for this purpose, ordinary food coloring applied with a cotton swab will do just as well.

Daily flossing is also required in order to remove the plaque that brushing cannot reach. Use a fairly long (at least 24-inch) piece of floss, wrap it around the middle fingers of your two hands, and hold it taut between your thumbs and forefingers. Slide it gently between the teeth, and floss with gentle, shoe-shiny strokes. Allow the floss to reach into the gum areas.

There may be some bleeding when these cleaning procedures are begun, but the gums will soon become toughened, and the bleeding should stop.

The low-sugar diet on which most children with Prader-Willi syndrome are placed gives parents a head start toward building healthy teeth and gums. If Dr. Rolla's advice on dental care is followed, parents can be confident that they are doing all they can to give their child the best care they can.

ILLUSTRATED BOOKLET AVAILABLE

We have available a number of copies of a booklet that explains through precise directions and illustrations the best method of toothbrushing and flossing. The booklets are free of charge. If you want one, all you have to do is send your name and address to THE GATHERED VIEW, and ask for the booklet, "Good Oral Health Can Be Forever."

THE BOOK VIEW

Understand Those Feelings, by Eugene T. McDonald, Pittsburgh, Stanwix House, Inc., c1962.

This book was recommended to all us Moms in our Prader-Willi parent group by a social worker who attended our last meeting. Dr. McDonald has written the book on comments and feelings parents have expressed to him over the years. It's very easy to read.

The parents all have a child born with some kind of disability. The feelings are so real I have seen myself or my husband many times in the book. I recommend the book to any parents; it will help you understand lots of feelings!

Reviewed by Sue Iverson, Kent, Washington

THE GATHERED EXCHANGE

Recipe: 5-Hour Stew:

3 lbs. lean stew meat
2 medium onions
2 cups celery, cut up
2 cups carrots, cut up
5 medium potatoes, cut in chunks

Put all ingredients in a roaster with a cover. Salt and pepper to taste. Add 1 large can of tomatoes or 2 cups tomato juice. Cover and place in 300° oven for five hours. Do not lift lid during that time. Very good!

Sue Iverson

An idea on social development:

Our children should interact with other children their age for normal social and speech development. I believe the more they are stimulated by their surroundings, the "eating" drive becomes less. As they find satisfying experiences, they will learn to direct themselves better.

Virginia Cararella, Madison, Wisconsin

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