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 Newsletter of PRADER WILLI SYNDROME PARENTS AND FRIENDS

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 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 \$6.00 along with your name, address, and telephone number to PRADER WILLI SYNDROME PARENTS AND FRIENDS, Box 252, Long Lake, Minnesota 55356.

NEW! NEW! NEW!

PRADER WILLI SYNDROME PARENTS AND FRIENDS has a new address. Please send further communications to the following address:

PRADER WILLI SYNDROME PARENTS AND FRIENDS
 Box 252
 Long Lake, Minnesota 55356.

If you already have communications in the mail, don't worry about them. The post office has the forwarding address and the mail will reach Mr. Deterling at his new address.

THE GATHERED VIEW, however, still has the same address in Federal Way, Washington. If you have letters for the editor or material you want published in the newsletter, please send them directly to THE GATHERED VIEW. However, membership renewals should be sent to PRADER WILLI SYNDROME PARENTS AND FRIENDS at the Minnesota address. Every piece of mail directed incorrectly means money spent to forward the mail to the correct person. The more money we can save, the more we have to spend directly helping our children and friends who have Prader Willi syndrome.

RECIPE FOR EGGNOG

¼ cup skim milk powder
 ¼ teaspoon vanilla
 1 egg
 ¼ cup water
 Artificial sweetener to
 equal 1 tablespoon
 sugar
 Dash nutmeg

Mix all ingredients except nutmeg in blend.
 Sprinkle nutmeg on top. Any flavoring can be used in place of vanilla. I keep my flavorings and spices in alphabetical order on the shelf and my child loves to go through the alphabet, choosing a different flavor each time he has eggnog.

MESSAGE FROM THE DIRECTOR

When we first conceived of forming this organization a little over a year ago our prime intent was to establish a vehicle for communication between parents and friends of persons with Prader Willi syndrome. In the ensuing period, it has become evident from the numerous requests we have received that much more is expected of us. We daily receive requests for assistance in locating professional help and treatment centers, dietary information, general information about the syndrome, bibliographies, statistical data, names of nearby members, and a variety of other very specific requests. We try to answer all of these requests. During the past several months, however, we have fallen behind in our efforts to keep up with our replies to all of these requests and this deserves some explanation.

Since March we have been considering a relocation to the Minneapolis area and are now in the process of making that move. Those who have been through such a move can appreciate how time-consuming the process can be and should be able to understand why some of our organizational activities are temporarily delayed. By August we will be relocated in the suburban Minneapolis area, and because of its more central location and excellent medical reputation, we hopefully will be able to strengthen our organizational activities. We look forward to making better progress in achieving our goal of more tangible benefits other than just a vehicle for communication.

Please note that our new address will be:

PRADER WILLI SYNDROME PARENTS AND FRIENDS
Box 252
Long Lake, Minnesota 55356

We will also retain our Harvard post office address for a period of six months during this transition period.

Many of you have requested back issues of THE GATHERED VIEW and not all of you have received them. Our initial supply of back issues was depleted by the deluge of requests for information from persons interested in joining our organization. We have, however, reprinted most of our back issues and can now provide them to those who wish to have them. If you requested copies in the past, but did not receive them, we ask that you please resubmit your request to our new Minnesota address. If there are other requests that you have made and are not fully satisfied with our response, please do not hesitate to contact us.

Now that we are into our second year of actual operation, membership renewal will be required for those who have been with us for a full year. Since we cannot expect to operate without additional funds, we are increasing our membership fee to \$6.00 effective 1 July. Overseas membership will increase to \$7.00 per year. We trust that this will not place a substantial burden on anyone. Reminders for membership renewal will be mailed, but we would welcome your independent action to renew your membership.

Eugene Deterling

IT'S COMING

The group of parents in the Seattle area who have been working on a booklet for parents are still carrying on. The manuscript has been typed, and a meeting is planned for late July to work on revisions. We are still searching for funding to get it published, but hope to have this problem solved by the time the final revision is complete.

A VIEW OF WHO'S WHO

In our WHO'S WHO column, we have previously published stories pretty much in the order in which they came in. However, this story received the past month, has so many good ideas, we have decided to publish it immediately.

Darren was born 18 May 1969 after a long labor. My husband was stationed with the Air Force in Thailand at the time, and I was staying in Mason City, Iowa, with my in-laws. At birth Darren was hypotonic and could not suck well. When I left the hospital he could take only 1/2 oz. formula, then was too tired. He didn't cry properly, but only "squeaked" a little. The doctors didn't know what was wrong, but felt he was retarded. They suspected Downs syndrome.

He was our third child, and I knew things were not right. He would just lie in his crib sleeping, or lie contentedly not caring about anything. I would make him notice things and play games with him even when he wasn't interested.

At four months he gave up his bottle; sucking was too tedious. He just pushed it out with his tongue. He was eating very little baby food. I poured milk down him with a childrens' cup with a lip on.

He was contented and happy and slept a lot. When he was three months old we moved to Omaha, Nebraska, where we spent the next five years. We were lucky to have a dedicated Air Force pediatrician, Dr. John Flom. He examined him every month and was very interested in him. Darren was hospitalized several times for tests, including chromosome, endocrine, brain wave, and muscle biopsy.

He sat up at one year and this was when he finally started to eat well. I was so happy I fed him continuously, and he became very chubby.

At fourteen months Dr. Flom referred us to the University of Minnesota for more tests. He had interned there and was acquainted with their facilities.

After the evaluation we were told it was Prader Willi syndrome. Upon our return to Omaha, Darren was placed on a diet and was referred to a pediatric neurologist. He followed Darren every month till he was three. His first steps were around this age and he only spoke about a dozen words.

We decided to get him into a program where he could be around other children. He was still a very contented and social child.

He was entered into an ENCOR school (East Nebraska Council of Retardation), where he was able to have speech and many other things on a one-to-one basis. In addition, I worked with him at home, especially on motor skills, fine and gross. My father-in-law built a special stairway consisting of three steps up, three steps down, each only three inches high, and a platform in the center. There was a rail along one side. I placed this in the living room and encouraged Darren to walk up and down. At first I had to hold the other hand, but eventually he did it with only one hand on the rail.

I started him swimming because no balance was necessary for this. By age five he was swimming like a fish, with natural ability to float and no fear.

He continued in the ENCOR school till he was four and then was placed into an ENCOR preschool program. ENCOR teachers, each with two children, were integrated into a normal preschool. The feeling was that non-delayed children would stimulate them to do more things.

They set up an obstacle course for Darren to run through, climb, etc. for his coordination, while his one-to-one relationship with his teacher continued for speech and a few other things.

Every two months I had a meeting with the teachers to discuss different problems or gains.

They served a hot lunch, but I packed Darren's lunch to insure he kept to his diet. Food stealing was now in full swing, but if he stole food, food was withheld.

He was still not toilet trained during the day, but if he stayed dry, his teacher gave him a star; his goal was a row full of stars each week.

He left the pre-school when he was five, as we moved to Germany. Retesting showed him in the low-normal range, whereas before schooling he tested in the retarded range.

Currently we live in a little village in Germany and Darren goes by bus to kindergarten. This is his second year of kindergarten, and possibly next year he will go one-half day to first grade and one-half day to special education.

He started stealing food at night, and I wouldn't find out till morning, so we now lock the kitchen at night. During the day he is never alone in the kitchen. Food is kept out of his reach, and as he has difficulty climbing, he doesn't try to get it.

His weight is fantastic. He has only gained five pounds a year since he was three; this is normal growing. He is just a little chubby around the tummy. He will be seven in a couple of weeks, and weighs only 51 pounds.

We work on exercise as much as we can. He didn't want to at first, but we insisted (he found out it was a lot easier to sit and color, etc.) We go for walks when weather permits, and swimming one or two times a week. I got him a springboard "spring-a-lene" to bounce on, and we make a game of counting how many jumps he can do. His father "jogs" every night with him. He can't run very well but he feels so special with just him and Dad. When we go shopping I park a long way off so we can walk a little extra. Sometimes his slowness can be a drag, but I persevere and try to stay patient.

We praise him all the time and tell him how lucky we are to have such a nice boy, and he is so well adjusted and self-confident. We are very pleased. He is rarely aggressive and has a very outgoing personality.

The biggest problem now is that the German neighbours give cakes and cookies because they love children. The language barrier prevents explaining his problem. At first he used to gobble it up, then tell me about it later. Rather than scold him, I explained to him about his diet and said he should bring it home and save it for dessert. Now he brings it home (after a nibble here and there), and I tell him how proud I am of him and how grown-up he's getting. When he goes back to play, I cut it in half, and so far he never notices. I have to allow him to keep it because if I took it away, he would eat it before he brought it home and I might not even know. At his meal I then have to take something away. When I explain he can't have his bread, or meat, or whatever because he's having cake, he never minds. He is starting to be aware of what he can or can't have on his diet.

It's amazing that when he steals, he can eat such huge quantities so fast, i.e. seven bananas in ten minutes. He visits the dietician once a month now. This means a lot to him to be weighed and he tries harder.

Our other two children had to learn never to leave food on the table when they were done as he'd grab it. He's even taken something off their plates when they looked the other way. One day they tried the trick on him. He didn't like their stealing his food, and that cured that.

Darren noticed other children got cookies and candy and started telling me, "Well, so and so can have a cookie." I told him that they don't have a weight problem like his, and he accepts that. I remind him of the low-calorie "treats" I make for him.

He can't keep up with other children, but he's content to play on his own. He was especially proud at the pool to show off to other 5-year-olds one year that he could swim and they couldn't. They were amazed at him, and he showed off all the more.

All in all he's doing beautifully. We have to work hard at it day by day but it's paying off. The only noticeably different thing about him to strangers is his speech. We are so lucky to have him, and so proud of his accomplishments. He has taught us a lot.

A LOW-CALORIE NIBBLER

Drain a can of mushrooms. Spread the mushrooms out on a cookie sheet and heat in a slow oven until dry. Has a nutlike flavor and only 17 calories per 90 grams of drained mushrooms (weighed before roasting.)

THE PROFESSIONAL VIEW

THE QUESTION, as submitted by a mother:

How many calories should a four-year-old girl with Prader Willi syndrome have each day?

THE ANSWER, as furnished by Peggy Pipes, Nutritionist:

The caloric (energy) needs of any 4-year-old child depends primarily on the basal metabolic rate, energy expended in physical activity, and the energy requirement for growth. Therefore, among any group of four-year-old children, the range of caloric requirements is large. To ascertain specifically the number of calories children with Prader Willi syndrome should receive, we have asked mothers to weigh and record all food that their children consume over a ninety-day period. By estimating the calorie cost of any weight children have lost or gained, it has then been possible to establish the caloric requirements for the specific children. The data that has been collected indicates that most children with Prader Willi syndrome maintain their weight in growth channel consuming ten to eleven calories per centimeter of height. To effect a weight loss of two pounds per month, it has been necessary to reduce the caloric intake to 8.5 calories per centimeter of height. It is important for parents to recognize that the figures reported are averages. In any group of children there will be those who should consume greater numbers of calories than average; also children who should receive less.

For those reasons, it is not possible to answer the question specifically. It is important for parents of children with Prader Willi syndrome to find an individual who can help them establish a reasonable level of calorie intake for their individual children.

Peggy Pipes, Nutritionist
Clinical Training Unit
Child Development and Mental Retardation Center
University of Washington, Seattle, WA 98195

THE BOOK VIEW

Are you racking your brain for things to keep your child and his sisters and brothers occupied throughout the long summer days? Two books that can come to your rescue are Home Play for the Preschool Child and 838 Ways to Amuse a Child, both by June Johnson. There are enough activities described in these two books to keep a child busy every summer from ages two to twelve.

The author has a rich background as a teacher and mother of three, and presents her ideas in easy-to-follow directions, illustrated with pictures and diagrams.

Each child is an individual, and has his own individual interests, but the broad range of June Johnson's material is sure to provide something for every child in your family.

Home Play for the Preschool Child, by June Johnson, New York, Harper & Brothers, c1957.

838 Ways to Amuse a Child, by June Johnson, New York, Gramercy Publishing Company, c1960.

Reviewed by Shirley Neason

THE GATHERED EXCHANGE

School Experiences:

I have found a most harmful experience in the educational system for children with Prader Willi syndrome is using food for behavior modification. Parents should be fore-warned of the implementation of this tool. I believe the child has the ability to turn this around and manipulate situations with it. It would be much simpler to forbid the use of food, than to turn it around later. This would have to be stated very clearly and understood before schooling begins. Hugs, kisses, pats on the back, a story, listening to a favorite record, a rah! rah! are a good way of rewarding and really help any child to feel secure and self-confident. He will also in the long-term learn to reward himself appropriately as an adult.

Virginia Cardarella, Madison, Wisconsin

Bedtime Snack Suggestion:

Our boy, now 24, always wanted a bedtime snack. He was 22 before we had a specific diet for him, but I came up with this solution. It is about 150 calories, which can be taken off one of the meals.

110 calories - 1 oz. spoon size shredded wheat
½ cup skim milk

This satisfies his urge for food and stopped his getting up in the night for food.

Ruby Stephens, Cortland, N.Y.

Physical Development:

In sharing with one another we should be able to find aids that are beneficial in physical development. Swimming is excellent and can be programmed at a young age. Small motor development aids are puzzles, Lego blocks, dressing and undressing dolls, keys and locks, and keyboard instruments, such as inexpensive electric organs.

Virginia Cardarella

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