

THE GATHERED VIEW

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Newsletter of PRADER-WILLI SYNDROME PARENTS AND FRIENDS

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MESSAGE FROM THE DIRECTOR

From our viewpoint 1976 seems to have been a better year than any of the other previous years for many Prader-Willi people and their parents. Whereas previously so many of the letters we received had a tone of desperation and despair, we have recently noted a sign of hope and encouragement in the majority of our mail. We can't account for this change in the tone of our correspondence, but one thing of which we are sure is that there is a marked improvement in awareness in the professional field about Prader-Willi syndrome. There is a corresponding improvement in the treatment opportunities for those affected by the syndrome. Many of the people from whom we now hear have recently found the professional help for which they have so long searched.

There are still a large number of parents, however, desperately searching for the right help. It is our most sincere wish that 1977 will be a still better year and all our Prader-Willi friends will be started on the road to a happier life. We hope that in some way we can help.

We regret that our Post Office Box does not have great stability, but because of the U.S. Government decision to build a new Post Office, we are forced once again to change our box number. Our new Post Office Box No. is 392. Everything else is the same. Mail directed to our old number will still reach us.

Another article on Prader-Willi syndrome should be appearing soon in a national publication. A syndicated columnist from a New York firm has contacted us about information for an article she is writing for GOOD HOUSEKEEPING magazine. We have supplied her with information about our organization, but expect the article will deal mainly with the positive results of diet control for the Prader-Willi people. The article should be appearing early this year.

Gene Deterling

REVENUE/EXPENSE STATEMENT - 1976 - THE GATHERED VIEW

Receipts:

Received from membership dues . . . \$507.00
Bank balance, Jan. 1, 1976 . . . \$ 74.39
Total . . . \$684.39

Expenses:

Printing \$148.41
Postage \$116.40
Office Supplies . . . \$ 10.89
Bank Service Charge . \$ 10.34
Total Expenditures . . \$286.04
Bank Balance, Dec. 31 \$398.35

THIS MATTER OF INTELLIGENCE

Judging from letters received by THE GATHERED VIEW, questions about intelligence rank high on the list of parental concerns.

At a meeting of parents of children with Prader-Willi syndrome at the Child Development and Mental Retardation Center of the University of Washington, a team of professionals tried to clarify some of the questions and to show the work that is being done to try to help children with intellectual handicaps to use their intelligence to best advantage.

Beverly VanderVeer, Ph.D., psychologist, spoke on intelligence tests. She told parents that basically an intelligence test measures the quantity of knowledge a person possesses that he is able to organize and communicate.

Intelligence tests for younger children are age scale tests. The child is given questions to answer and tasks to perform, and his performance is compared to other children of various ages. The result tells us his mental age. If a child can answer questions and perform tasks at the level of a normal three-year-old, then his mental age is three, regardless of his chronological age.

Intelligence tests for older children compare the child to a large sample of children his own age and rate him on a scale. On this scale, a score of 100 is considered to be the mid-point of the range of intelligence. An I.Q. of eighty to 120 is average, and about two-thirds of the population fall within this range. One-third of the population are either above or below average in intelligence as measured by this method.

The whole idea behind intelligence tests originally was to predict how well a child would do in school with traditional teaching methods being used. As a result, the tests are not designed to pinpoint specific abilities and weaknesses an individual child may have.

The problem of trying to pinpoint these abilities and weaknesses is being addressed by Judith Warren, as she works toward her doctor's degree under the direction of Dr. Earl Hunt. Warren spoke about the work she is doing.

She told the parents that modern cognitive theory has isolated a number of skills needed by the human being to receive a sensory impression and to respond to that impression. First a person has to code the incoming information. The person who codes information faster gets more information in the course of his every day experiences. Second, the person has to hold this information in his mind while he does something about it. This is called short-term memory. A person with poor short-term memory has difficulty solving problems. Third comes a process called transformation, or deciding which information is relevant according to how the individual thinks he will need to use the information in the future. At this point, a person has problems if his coding system for long-term memory does not work very well.

Other factors involved in learning relate to individual differences in abilities involving simple recognition and recall. In simple recognition, a person can recognize something when he is given a cue, such as in a multiple choice test. In recall, the person must be able to remember things without any such clue. Learning is also affected by the attention factor, or how well a person is able to concentrate.

Warren's research has involved looking at how children think, trying to identify differences, and trying to determine what goes wrong in those children who have learning problems. Children with the same syndromes were compared in an attempt to learn what similarities, if any, exist in their patterns of learning. She wanted to know what their strengths and weaknesses are so that teaching strategies can be devised to utilize the strengths and hopefully correct the weaknesses. After using new teaching strategies, the children will be tested again. By this time it should be evident whether the weaknesses can be corrected, or whether it is necessary to simply work around them.

When Warren tested the children, the first test she gave was a simple recognition test. The children were shown a series of pictures. Some pictures were repeated at intervals. The child was asked whether or not he had seen each picture before in that test period. The test was repeated the following day, with some pictures being new, other pictures the same as the previous day. The child had to remember which pictures he had seen previously during that day's test period. The results for children with Prader-Willi syndrome was very revealing. They scored ten points lower on the percentile scale compared to children who tested in the same I.Q. range, and who were similarly placed in school. This indicates that children with Prader-Willi syndrome have a poorer short term memory than the other children tested.

The second test was a pairing test. The children were shown a series of slides. Each slide contained a number with a picture of objects representing the number. For example the first slide might have one cat and the second might have two cars. The next slide would have just a number without a picture. The child would have to remember what object went with that number. The next two slides would have the same numbers, but one of the objects would be changed. For example, they might show one cat and two airplanes. It was discovered that the children had little difficulty remembering the object if the blank slide showed the same number as the one just before it. However, if there was a gap, performance went down. This corroborated the results of the first test that children with Prader-Willi syndrome tend to have poor short term memories.

The pairing test was repeated on the day following the original test. On this second test, the children were asked to continually repeat the numbers and objects on the set of slides they were being shown. This time there was a significant increase in the number of correct answers, showing that the children remember better if they can verbalize the information they are trying to learn.

The third test given was a coding test. The children were given pairs of letters and asked if the two letters were the same letter of the alphabet. Letters might be physically identical (AA) or just name-identical (Aa), or different (AB). Typically it takes people longer to decide that the name-identical letters have the same name than it does to decide that the physically identical letters have the same name. (Of course, both decisions take only seconds.) Children with Prader-Willi syndrome followed the typical pattern on this test, performing equally as well as other children of equivalent mental age.

As a result of these tests, Warren has concluded that children with Prader-Willi syndrome also must have some learning strengths that other children with intelligence quotients in the same range do not have. This would account for the fact that they are able to do as well in I.Q. tests when they are obviously weaker in some learning skills. In future testing, she hopes to find out what these strengths are.

In conclusion Warren gave parents two things to keep in mind in order to help their children improve their learning ability. First, give the child the information he is learning a little slower than you would give it to another child. Give him time to assimilate and digest information and skills before going on to new ones. Second, give him opportunity to talk about or recite the material he is learning.

Although there is still much to be learned about intelligence in children with Prader-Willi syndrome, parents are encouraged by the fact that research is still being carried on that will lead to new discoveries that will enable parents to turn their concern into action that will benefit their children.

TO REDUCE THE NUMBER OF CALORIES IN GROUND BEEF: Brown the beef in a pan without added fat. Drain off the accumulated fat. Then squeeze the browned meat in paper towels to "squeeze out the calories."

A NOTE TO THE TEACHER

Each year I have had a conference with my child's teacher at the beginning of the year to explain his problems. Because of the difficulty of remembering everything I wanted to say, I decided this year to write out a list of problems and ways of handling them. Now the teacher can have it for reference throughout the year. Another Prader-Willi mother read my note and asked for a copy for her child's teacher. As a result, I decided to print it in case other parents might have some use for it. Some parts may not apply to your child, but if you care to use the information, you can delete the parts that do not apply.

CHARACTERISTICS OF CHILDREN WITH PRADER-WILLI SYNDROME THAT COULD AFFECT THEIR SCHOOL PERFORMANCE

1. Tendency to obesity. The child gains weight on what would be a normal diet for the average child. Therefore, his food intake must be closely monitored to see that he gets no food not allowed for in his diet.
2. Compulsion to eat. To complicate the first problem, the child has an overriding compulsion to eat, especially sweets. His relationship to food is similar to that of the alcoholic to alcohol. He cannot help himself. If the food is where he can get it, he eats it. He controls himself somewhat better away from home, but supervision is the only reliable control. Because of this, it is not wise to use food as either reward or punishment.
3. Temper tantrums. Children with Prader-Willi syndrome seem to have a short fuse. Although they are congenial people most of the time, they can throw violent temper tantrums. Our way of handling tantrums has been to isolate him until he calms down. We also firmly let him know that he may not be disruptive or destructive during a tantrum. Again, he seems to have more control away from home.
4. Lack of muscle tone. Prader-Willi syndrome causes weak muscles, which make it difficult for him to participate in physical activities on a level with his peers. However, he does need exercise, and should be encouraged to be active during recess.
5. Brittle bones. The bones are brittle and easily broken. However, his lack of activity reduces the risk. Parents should be notified of falls or injuries.
6. Poor short term memory. The child has poor short-term memory, and may forget what he has been told just moments before. However, once a fact or skill has been impressed upon his mind, he remembers it well. Repetition, drill, and verbalizing material will help him learn faster.
7. Distractability. He is easily distracted by what those around him are doing. When he is distracted, he has difficulty getting back to what he was doing. Sometimes guidance is necessary to get him back to his task.
8. He finds self-correction difficult. Once he gets on the wrong track, he often cannot correct himself without guidance. When this happens, he needs to be stopped at the point where he is making the error and shown the proper way.
9. Lacks social skills. The child has the same problem with self-correction in social skills. He has difficulty approaching and relating to people in an acceptable manner. If he is rejected because of his inappropriate behavior, he simply tries the same thing again, but tries harder. It is necessary to take him aside and let him know that he needs to change his approach. Even then he may argue about it. The psychologist has told us that as a general rule a child with this kind of difficulty finds it easier to try a familiar behavior even though it doesn't work for him, than to take the risk of trying something new. Role playing sometimes helps, with the child taking the role of the other person and someone else playing his role.

A NOTE TO THE TEACHER, Continued

10. Needs firm guidance. The child functions best when he has well-defined limits and an adult in charge who keeps firm control of those limits. He has to be led in independence more gradually than the normal child.
11. Sleepiness. He seems to have a tendency to fall asleep at times when his mind and body are not actively engaged. If he falls asleep in class, it is best to awaken him and get him started back on the business at hand.
12. Self-image. Because he does more poorly than his peers in almost everything he does, he needs help to find acceptance and love from the people around him.

A VIEW OF WHO'S WHO

Here is news to bring us up-to-date on the story of Christopher, from England, whose story appeared in the March and May, 1976, issues.

We are so thrilled with Christopher. He has lost ten pounds since April. I took Peggy Pipes' letters to my dietician at the local hospital and she worked out a diet for me from these. It has worked! We can't believe it; we have tried so many diets before, and he has still continued to put on weight. He has just had his first pair of trousers bought ready made from a shop, and he is very pleased with them.

Christopher walks easier now. Ten pounds doesn't sound a lot, but it certainly has made a difference to him. He is still very stubborn and wets himself during the day. Sometimes he is worse than others. He rarely wets at school; mostly at home. If anyone has any ideas about trying to stop this, I would be very grateful. Perhaps some other parents have the same problem. I look forward to the newsletters; it helps a lot to hear about other peoples' experiences.

Ann Watson

THE GATHERED EXCHANGE

From a mother on bed-wetting:

My son is twenty-four years of age, and bed-wetting is a problem. I will be very interested to read about others who have to deal with the same problem and how it is being handled.

From another mother on the same subject:

We used a bedwetting alarm device successfully with our child during his early school years. This device causes a buzzer to go off when the person starts to wet, waking him up. We got it from Sears, but perhaps it is also sold in drugstores.

More on relief from the scab problem:

We did find one thing to help the scab picking. Our doctor suggested lanolin rubbed into the scabs two to three times a day, and it seemed to help Kathy. If she just remembers to use it, she can get them all healed up. Simple treatment, but it works for her.

Mrs. Harold L. Olson

Another mother has this to say on the same subject:

Our pediatrician gave us some advice on the problem of dry skin and picking. He said:
1. Keep the fingernails short (this probably should have been obvious, but it had never occurred to me to keep them extra short.)

THE GATHERED EXCHANGE, Continued

2. Use a good moisturizing cream on dry areas and massage it in well. Two he suggested trying are Pond's Dry Skin Cream and Keri-Lotion.
3. Use an over-the-counter anti-biotic ointment on scabs around the nose and other areas where infection might easily occur.
4. If dryness or rash on the feet is a problem, try using cotton or wool socks rather than those made of synthetic fibers.

Mrs. Olson also has a question:

Do you know if there are different degrees of severity in Prader-Willi syndrome? Kathy has such a terrific drive for food as compared with case histories I've read.

And here is a comment on her question:

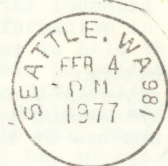
From information gleaned through contact with other parents in our parent group, I would say yes. Some children seem to have no control at all over their eating desire, while others have developed varying degrees of self-control. Even in our own child we see variation at different times in his ability to control his desire for food not allowed for in his diet.

RECIPE: Basic Oil and Vinegar Dressing:

1 tablespoon cornstarch
Pinch of pepper
1 cup water
 $\frac{1}{4}$ cup cider or wine vinegar
2 tablespoons vegetable oil
1 clove garlic, minced

Combine cornstarch, salt, pepper, water, vinegar, and oil in small saucepan. Cook over medium heat, stirring constantly until mixture comes to a boil. Simmer one minute. Stir in garlic; cool. Refrigerate in covered container. Makes $1\frac{1}{4}$ cups.
3 tablespoons = one fat exchange
14 calories per tablespoonful

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