The Gathered View

Newsletter of the Prader-Willi Syndrome Association

MARCH-APRIL 1989 VOLUME XV, NUMBER 2

PRESIDENT'S MESSAGE

As I sat down to write this letter for the Gathered View the computer automatically printed the date across the top. It was February 12, known to every school aged person as the birthday of Abraham Lincoln. During my growing up years that day was always special for another reason. Lincoln was lucky enough to be born on my Mother's birthday. There is always someone special in every life. As parents of persons with Prader-Willi syndrome we are ever aware of that someone special. But in the lives of these persons with PWS there is usually someone special.

In the life of my daughter, Sarah, there was a special person who came into our lives during the first year that we were introduced to the names for Sarah's problems. One of the first things that we learned about PWS was that she would need a special kind of person to become her teacher. I remember taking Sarah for our interview and to observe the classroom activities at the speech and language school. She was too young to start in public school and our pediatrician recommended that we start her with Donna's class. For almost ten years, even after Sarah left her classroom, Donna's guiding hand was felt on Sarah's educational progress. Probably one of the most impressive concepts to me was the way Donna created a sense of security for her students, even to the degree that behavior not suitable to the group's activity at the time was managed by directing the person to a safe corner to gather themselves together until they could come back to assume their responsible role in the classroom group. The last time that I had a chance to talk to Donna was at one of our annual conferences, between sessions. It gave me a chance to give her an update on Sarah's progress. Towards the end of the conversation we discussed how she had so impressed Sarah with the teacher role that Sarah had now assumed that role in her quiet play times and had expressed her strong desire to be a teacher when she finished college. My eyes moistened more than men like to admit when she responded that such a goal was not impossible. That was several years ago and now my daughter is in a high school special education class. The other day while we were shopping, walking hand in hand through the mall, she turned to me and said in that high pitched happy voice, "Dad, I want more than anything to be a teacher. I know it is going to be a lot of hard work. But, if someone wants something bad enough. I can do that hard work and I'll be a teacher." Anyone know what a salty smile is?

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6490 Excelsior Blvd., E-102, St. Louis Park, MN 55426

PWSA
6490 Excelsior Blvd., E-102
St. Louis Pk., MN 55426
(612) 926-1947
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President's Message (continued)

Yes, there is at least one like that in most of our lives. It's impossible to live with persons who have disabilities without crossing paths with some caregiver that gives one hundred and fifty percent doing their job. If you have read this far I'll wager you are now sorting through your mind all of the good people that have helped you and yours to survive the problems that have been a challenge. I do in fact have an ulterior motive for letting you peek at Donna's role in Sarah's life.

Yes, there have been others. Some were physicians, social workers, other teachers and in some cases they were groups of people who made a support group or school or clinic. Each of these persons responded to an individual with PWS. Each learned something from that encounter and similarly each took something away with them that enriched their lives and knowledge, something that may be lying dormant waiting to enrich the life of another person with PWS or the family that give support.

While the names of the persons and their faces float through your mind and bring smiles inside of you, get their names onto paper, attach an address and telephone number. Now send that information to the PWSA office so those names can be added to our Directory of Services for future reference by another PW support person looking for help. Almost forgot. A little information regarding each person's talent, such as occupational therapist, speech teacher, school psychologist and the organization that they work for. Thank you. -- *Delfin J. Beltran, M.D.*

Information Sought

PWSA is aware that we have members whose children have the characteristics of PWS caused by accidents, tumors, etc. and have a membership because some of their problems are similar. One such parent would like a listing of these parents in order to share information. If you would like your name shared, please contact the National office.

Vocational Placement

Facing life following the completion of a child's schooling, can be difficult for parents. While our young people do not need the repetitious training needed by some retarded people while still in school, preparations must be made before the child graduates. Frequently there is only one workshop in an area and the waiting list may be "years" long.

PW parents need to be very involved when vocational plans are being made. A school that has worked with a child for years will still recommend light office work, janitorial work, or restaurant work without giving one thought to the restrictions of food control and understanding of behavior. They will only look at the mild retardation and say, certainly this person is capable of community based employment. (As always, it must be mentioned we are speaking of individuals, and individual needs differ but the "average" person with PW falls in the mild retardation range of I.Q. and functioning.)

Accepting the other limitations of the syndrome is very difficult for professionals. Even most sheltered workshops have a state mandate to work their clients into less restrictive jobs. Is it being fair to our young people to repeatedly be placed in the community just to face another failure? Certainly there are circumstances, when properly arranged, that can meet the needs of our young adults without the gaining of weight being a trade-off for more independence.

We would like to share more actual stories of vocational placement, how about it?

PW Family Joys

My family has learned to eat better and the importance of daily exercise. Our family rule is we must all exercise on a regular basis -- exercise can be biking, walking, swimming, organized sports, or any exercise that improves our health. I am very proud of my children's physical condition -- my 12-year-old is a black belt in Taw Won do, and a winner of a triathalon. My daughter (our child with PW), who is 10, is a good swimmer and was the only female in the State of Oklahoma entered in the 50 meter events at the Special Olympics last year. My 5-yr-old is also a good swimmer and biker.

In answer to a previous request, I believe we must see what we can do and not dwell on what we can't do. I believe much of the "doom and gloom" stems from the work we have to do to achieve a place in this world for our children. It is terrifying to be the only parent in a whole city with our syndrome. Many professionals are completely in the dark about our needs. I propose that we all work toward a PW Awareness Day. If all of the members joined together in this project we could be impressive.

SPECIAL REQUEST

A consultant psychiatrist of the CPRI has asked for information from parents regarding Methylphenidate (Ritalin) success in treating attention deficits and classroom concentra tion, as well as reduction in appetite. They are also interested in seeing children with PWS who might be living in their local area of SW Ontario. The contact is: L. Jerome, M.B.,Ch.B.,M.Sc.MRC, CPRI, Ministry of Community and Social Services, Sanatorium Rd, PO Box 2460, London, Ontario N6A 4G6, Canada, (519)471-2540.

Notes from Nutrition Book

The wisest way to diet is to continue to eat the full variety of foods in the Basic Four, including bread and cereal products, but cut down on portion size. Persons on extended weight loss programs, those with major cutbacks in calories, should take a daily multivitamin and mineral supplement.

If your diet contains fresh fruits, vegetables, good nutritious foods and Vitamin C, that supplement should not be necessary.

Certain drugs interfere with the action of certain vitamins - and - certain vitamins taken to excess can interfere with the action of certain drugs. Your physician should be informed what your child is taking.

Women taking oral contraceptives are often found to have reduced blood levels of thiamin, riboflavin, B-6, B-12, folacin and Vitamin C. Deficiency of B-6 has been associated with mental depression. Some readers might wonder why oral contraceptive are mentioned in connection with PWS, but this can be a requirement of a group home and is sometimes prescribed for estrogen therapy.

Natural forms of Vitamin E are slightly different in chemical structure and more effective but there is no difference, except to the cost of natural vitamins vs the synthesized ones.

Rights and Responsibilities

As the next conference approaches, it is time for the membership to give some thought to the running of the organization. Members have rights and responsibilities in the election of the Board of Directors and can also initiate change if they so choose. In the next issue of the GV we will run a profile of the board members who will be coming up for re-election. If you, as a member, would like to nominate someone for one of these positions, a nomination now would give us time to profile that candidate as well as the incumbents. Nominations made at the conference will not have this exposure. You may also make a motion to be considered by the members in attendance at the July meeting by submitting that motion in writing to PWSA at least 90 days prior to the meeting. Now's your chance if you want it.

Our New Look!

Frequently members ask what they can do to help PWSA, and unfortunately most of the time little can be done away from the national office. One of our members, who just happens to run a PR firm, offered to edit and format our newsletter. We have been making changes slowly the past few months as a direct publishing program was being learned, but with this issue we have taken a giant step forward by having this issue "professionally" done. This offer, which is most generous, frees the office to spend additional time on other projects. We hope you like the "new look".

Thanks for Your Response

Dorothy Thompson would like to thank the many members who called and sent cards wishing her a fast recovery from her recent fall. Recovery is slow but she is coming along fine. Your response shows how very much we appreciate all this lovely lady has done for us.

Foreword from *Management of Prader-Willi Syndrome*

I have had a major interest in Prader-Willi syndrome (PWS) for over 30 years and, having dealt with many patients, have reached the conclusion that PWS is one of the two most grave ailments I have encountered -- the other being Huntington's Disease (HD). Anyone who has witnessed the mood swings and the relentless, progressive, intellectual, and physical deterioration associated with HD would agree beyond a doubt that it is a devastating condition. PWS is an equally devastating birth defect that characteristically presents major problems from birth. The enormous difficulties associated with the first phase of PWS cause frustration and guilt in mothers who perceive themselves as inept at feeding and nurturing their affected child. This guilt intensifies during the second phase, as PWS children constantly appear plagued by a relentless hunger that dominated their lives. The presence of this insatiable urge to eat, which is beyond the control of the patient, his family, or physician, becomes the primary focus for the child and inhibits all other activities and interests. In addition to the issue of satiety, a PWS child faces a life of sexual incompetence. Reactions to these problems are further aggravated by hypothalamic dysregulation, which seems to affect temperament. It is not surprising, therefore, that emotional incontinence increases in severity and frequency as the PWS child grows older. Parents, usually unable to manage diets, food-seeking activities, and bizarre behavior, become distraught and emotionally drained. Family systems deteriorate and life becomes hell for all concerned. In my experience, parents of PWS children come to the physician's office in great distress and total despair more often than parents of children with any other birth defect. They speak of their helplessness, the sacrifices of other family members, their love for their affected child, and their concerns about the future. One common theme is the reluctance of many parents to consider alternative living arrangements. This may reflect attempts at denial that a problem exists with which they cannot cope. Fortunately, effective management, even for the most severe cases, is a realistic possibility, particularly after parents are finally able to accept the fact that the family home is not the best place for the affected adolescent or adult.

It has taken years to recognize that solutions to the multifaceted problems of PWS require the expertise of many specialists -- pediatricians, neurologists, endocrinologists, nutritionists, psychologists, nurses, special education consultants, speech therapists, physiotherapists, and occupational therapists -- all play important, collaborative roles in the lives of PWS individuals and their families.

This book is the result of the realization of its editors that a cooperative effort is needed to ensure appropriate interventions. They were able to assemble an array of experts, each of whom presents suggestions for how a specific discipline can best help. The book gives useful directions to all of those involved in the care of children and adults with PWS, not least to the parents. It may not only help to provide a sophisticated treatment program for PWS, but it may also encourage specialists to collaborate to help the PWS individuals and their families to carry their lot.

Hans Zellweger, M.D.

Opioids

Alan Levine, Ph.D., Assoc. Director of Research VA Medical Center, has focused his research on how endogenous opioids influence eating. He mentioned abnormalities of eating are prominent features of several developmental disabilities including PWS. He stated the possible role of opiate drugs and opioid receptor mechanisms in the brain in food intake has long interested pharmacologists and food scientists.

Naloxone, an opiate antagonist, has been studied with regards to appetite control. Levine and his colleagues were one of the earlier groups to note that animals might become addicted to its endogenous opioids.

A study was conducted by Levine and James Mitchell, Dept of Psychiatry, University of Mn and found no difference in weight loss between the treated group and control group, indeed a disappointing finding. However, several investigators have shown that naloxone and naltrexone can be effective in reducing short term feeding.

While Levine believes opioids are involved in feeding in an important way, he does not think it is realistic to believe that obesity can be controlled by regulating feeding through antagonism of the opioid receptor alone. Body weight is not regulated by food intake alone; energy expenditure is also a part of the equation. There is some evidence that opioids might also affect energy expenditure, but this area of research is in its infancy. (Excerpts from MCRDD, Institute of Disabilities)

Chapter Updates

Many chapters meet regularly in many parts of the country. Meeting local goals and meeting the needs of all of their members is a great challenge for these groups. Recent new presidents are: Tim Inwood, CT; Glyndia Williams, KY; Kathy Wyka, Midlantic; and Paul Alterman, GA. Congratulations. We know the chapters are in good hands.

Several groups are working on designated group homes. The new Kansas Chapter has just started a newsletter, "The View from Kansas".

If you are not an active member in your local chapter please consider the advantages of making this effort. If you wish to know the nearest chapter to you, please ask national for a referral.

Annual Conference Announced for July 19 to 22, 1989

Despite the cold, summer is coming . . .

. . . and with summer, we have to give thought to the upcoming conference. It will be the 11th Annual National Conference, and the 1st International Conference. We are looking forward to having a terrific meeting in Calgary, Alberta, Canada.

Preparations having been ongoing for a year already, and the preliminary agenda looks very informative. Much thought is going into making the Youth Activity Program very inviting while attempting to making it accommodate the needs of all of those attending. Anyone who has worked on this program knows it is indeed a great deal of work but knowing the response of those who attend, we know it is well worth the efforts.

The Pre-registration packets will be prepared and bulk mailed (U.S.) in March. The Calgary Host Committee will be directly mailing the Canadian packets. If you have moved, and this edition was forwarded to you, you must get your new address to us in order to receive this packet. Bulk mail is not forwarded.

Get that vacation time scheduled now and we'll see you in Calgary in July. *Don't forget -- those in financial need may make an application for our Conference Grant. For information, contact the National office.*

PRADER-WILLI SYNDROME ASSOCIATION
Annual Conference
CALGARY, ALBERTA, CANADA
July 19-22, 1989

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Genetics

Mario R. Capecchi, Professor of Biology, The University of Utah, recently reported a technique which allows a new gene to be targeted so that it will search out and replace another gene exactly. That means that it will insert itself into the same location on the same chromosome as the original gene. Naturally, years of research will follow and there are some other researchers that hailed the research but state there are many important questions remaining to be answered.

Among the big-budget, big-science proposals facing the new Congress is the $3 billion human genome project proposed by the National Academy of Sciences. It is predicted they would need to spend $200 million a year for 10-15 years on this project, and it is hopeful that it will be approved.

Weight Management

Obesity is a medical term defined as the storage of excess fat in the body. Fat cells are essential to the body as they serve to cushion our major organs. Fat cells only become a problem when more energy is going into our bodies resulting in storage of fat than is going out. It is a fact:

. . . diets and programs designed to lose weight fast will not keep you thin and may actually be harmful.

. . . weight lost in the early part of a strict dietary program is not fat but water. Your body quickly adjusts to reduced food intake by lowering your metabolic rate which inturn resists burning off fat.

. . . obesity is not only caused by too much food, but also by too little exercise.

. . . exercise will not only retain but will actually increase active muscle mass. Fat deposits are mobilized in your body to be burned as energy by the body, weight loss through exercise is nearly all fat loss.

. . . it takes 3500 calories to lose or gain one pound (for the normal person). Walking 3- miles in 1 hour will burn 300 calories. This does not seem like much but if we continue this for a whole year, we are talking of 30 lbs. not 1 lb.

A Parent Shares

Our son was not diagnosed until 2-years ago when he was 11 years old. At 13 he weighs 70 lbs. (a loss of 40 lbs.), exercises 5 days a week and is fairly well managed emotionally. We face each day at a time and keep perspective. After all we have two other beautiful children and a wonderful life together. It's not all bad. Self-pity and depression feeds on itself creating a disability far worse than PWS.

Michigan Parents Share

Our son was born a term baby by dates but turned out to be a intrauterine growth retarded baby of 35 weeks with severe hypoglycemia and hypotonia. The first few weeks were a struggle with feedings and with the grief of loss of our normal expected child. We had had amniocentesis and were assured of a chromosomally normal child. Subsequent testing has determined that he has the expected abnormality of his chromosome 15. Our greatest frustrations have been the extreme prejudice of the neighbours, not all but some. It really has been a remarkable learning experience for us to find the extreme lack of compassion and general understanding of handicapped children.

The pre-primary learning program with the intermediate school district in our community has been working with our son and I think it has been an experience that has benefitted him. Presently he is 18 mos. and is beginning to pull himself up and starting to bear weight. He is vocalizing well and cognitively is functioning at his age. We of course know that this most likely will change as he grows older. However, he has not been the "disaster" the neonatalogist predicted. He is in fact a very loving and beautiful young man. I was most encouraged by a recent newsletter. There were positive reports by parents! We have a tough road to follow in the future but it is with determination and love that we hope to succeed. I am grateful to you and all the organizers who continue to diligently work on our son's and our behalf.

The Necessities of Operating

Annually PWSA has held a spring fund drive (for various purposes). As we are in the process of readying our contacts for the drive this year, we would appreciate our members sharing the complete names and addresses of friends and relatives that they feel may be willing to make a donation. In the past few years we have contacted over 1000 "outside" sources for donations and have added considerably to our donated total.

It only takes a bit of effort to drag out that address book or Christmas card list, and it can be a considerable help to our efforts.

In doing this drive we mail not only to these friends and relatives but we also include our membership. It would be tremendously difficult and time consuming for us to remove the names of our systematic and generous donors, so please know your donations during the year are appreciated and this funding effort is aimed at increasing the number of donors from our membership.

PWSA has also conducted a Conference Fund Raising Raffle each year for the past several years in order to offset higher registration costs. As with most fund raisers, the idea wears out and it is time to start something new. It has been decided to conduct a raffle through the purchase of a pocket calendar. We have ordered these calendars for the year 1990 and will begin selling them in July. More information will be shared with you in a later edition.

One Response to Temper

One of our members asked membership response to the problem of lack of control. One letter mentioned food and artificial colors and flavors allergies. This is a subject that is treated with disagreement in the medical community but has been reported enough to at least be considered. This mother suggests that even though it is a great deal more work, the mother should consider eliminating all convenience foods from her child's diet and make everything from "scratch" eliminating sugars, colors and flavors. If this seems to diminish the number of control problems, certain foods could be added back, one at a time, to see if any increase is noted.

MacArthur House Group Home

The MacArthur home, a designated PW home operated by Lutheran Social Services, is seeking a fourth resident. An appropriate referral would be an adult male between the ages of 18 and 30, with a functional level in the EMR-TMR range. They would be seeking someone who plans to make this a long-term placement. For further information, call Program Supervisor Randy Schubert at (608) 221-8786.

Rehabilitation Summer Camp

The Rehabilitation Institute of Pittsburgh will again hold their summer programs for those with PWS. Due to this facility's ability, they are able to offer a camping experience that qualifies for insurance reimbursement. Their first program, from June 19 to July 14, will be for teens 14 to 20 years old. The second program, from July 24 to August 25 will be for children 7 to 13 years old. Over the years they have added new activities; however, their major emphasis remains on improving adaptive behavior, improving muscle tone and physical fitness, and nutrition awareness and weight loss. For further information, contact Program Coordinator Bea Maier, Ph.D. at (412) 521-9000.

A Canadian Mother Shares

When our son with PW is away and I am home alone, it is very easy to plan to do many things because "it is my time". This is particularly true for working mothers who are not home while the child is in school or mothers whose child does not attend school. It has taken me a long time to learn that the best use of my "alone" time is to fully relax, to recoup my resources in order to be best able to handle whatever the next day may bring.

Coming Attractions!

New Brochures: Committees are working on the text, formatting will be done and hopefully will be approved in July. We feel these will be a great addition to the information now available.

Handbook in Spanish: Thanks to a grant and the suggestion of Dr. Cassidy in Arizona, our present handbook is being translated into Spanish and hopefully will be available in the future.

Handbook for Younger Parents: Sue Inwood, CT, is nearing the completion of her efforts to develop a handbook for younger parents. We hope to have this available to our membership in the next few months.