The Gathered View
National Newsletter of the Prader-Willi Syndrome Association (USA)

USA
PRADER-WILLI SYNDROME ASSOCIATION
Still hungry for a cure.

# Obesity Still Kills

***By Janalee Heinemann, MSW, PWSA (USA) Director of Research & Medical Affairs***

As I write this, I am working on getting money to pay toward the funeral of a nine-year-old girl with PWS, reminding me once again that PWS is still the deadly appetite. Although currently many of our children and adults who have Prader-Willi syndrome (PWS) are slim due to early diagnosis, education of parents and other care providers, plus growth hormone, I do not want us to get too complacent.

**While the holiday season usually requires extra vigilance, careful monitoring day in and day out all year is essential.** Recent calls for help that I have received in our crisis program include: girl, age 4, 120 pounds; girl, age 5, 117 pounds; boy, age 9, 126 pounds; girl, age 13, 254 pounds; boy, age 11, 220 pounds; teen, 326 pounds; male, age 24, 550 pounds–and the list goes on.

In studying the 163 PWS deaths that are in our **PWSA (USA) Study of Death** database, Dr. Stevenson wrote: "*Regarding the obesity-related deaths, in removing all pediatric cases and cases where cause of death was unknown or clearly not obesity related (accidents, choking, etc. – although some of them may have some obesity component), the average weight at death for these individuals was 257 pounds.*" Besides people with PWS being at greater risk of obesity, this obesity puts them at risk to die of obesity-related complications. In separate international reports of deaths in PWS, a large proportion (half or more) were related to respiratory or cardio-respiratory disease.

Dr. Linda Gourash (Pittsburgh Partnership) reports in her poster presentation, **Clinical Presentation of Obesity Hypoventilation and Right Heart Failure in Prader-Willi Syndrome**, "*The all-too-familiar clinical picture of shortness of breath, decreasing physical activity, and leg swelling seen in morbidly obese persons with Prader-Willi syndrome is due to hypoventilation during sleep, leading eventually to respiratory failure with or without right heart failure. The onset may be rapid or slow but is always insidious, that is, initially unrecognized.*"

In **Mortality in Prader-Willi Syndrome** from New South Wales (published in Am J Ment Retard. 2006 May), the authors of the study state: *"Prader-Willi syndrome is a substantial risk factor for death, above the risk related to intellectual disability alone. Those with Prader-Willi syndrome have higher mortality rates than those with intellectual disability in general. They have a much higher estimated mortality ratio than a comparison group with mild or borderline intellectual disability. The pattern of death in those with Prader-Willi syndrome suggests that obesity-related disease is a likely major risk factor. Management of caloric intake is truly a life-and-death issue for people with this syndrome."*

**Please remember, and remind your relatives and schools, when you are showing your child love during this holiday season, with Prader-Willi syndrome, you can love your child to death.**

## WE REMEMBER



*Leslie*
*DOB 2/19/83 DOD 2/21/11*



*Alden*

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# Online Auction!

We are having an online auction for the National Conference and need your help!

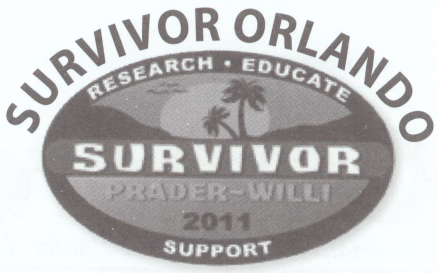
This year instead of a silent auction at the Gala Banquet, we will be holding an online auction two weeks prior to the conference and continuing through the conference weekend. Having an online auction will allow everyone a chance to participate even if you cannot attend the Conference.

In order to make this a success and help raise funds for PWSA (USA), we need lots of items! That is where you all can help. Please approach your local businesses and ask for donations for the online auction. Contact me, and I will provide you with a sample letter and donation form to use.

## Some great item suggestions are:

-Sporting events and attraction and show tickets
-Resort and hotel stays
-Vacation packages and vacation homes
-Hunting and fishing trips
-Airline tickets and cruises
-Jewelry, artwork and collectibles
-Gift certificates to nationally known chain restaurants
-Autographed sports items and more

All items must be new and in good condition. The items collected should be shipped to me where they will be cataloged and placed on the website for bidding. They will then be shipped to the winner after the auction closes.




***Please help us by obtaining items. The more we have, the more money we can raise.*** Please contact me with any questions you may have. I appreciate everyone helping to make this online auction a great success!

Watch the Web site or e-Bulletin for further information about bidding.

See you in November for Orlando Survivor 2011! ■

Michelle Torbert
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# Marcia Dunn ~ Our Shining Example of Courage

Marcia Dunn is 79 years old, the mother of 6 and the grandmother of 8. Ten years ago Marcia did not know how much her life would soon change. That year her grandson, Aiden Dunn, was diagnosed with Prader-Willi syndrome, she was diagnosed with leukemia, and she had just moved from New York to Sarasota, Florida, when she opened the paper and saw that volunteers were needed at PWSA (USA). Until that time she did not even know that the national PWS office was located in Sarasota.

Since then, even with more hardships thrown her way, Marcia has volunteered with a smile on her face every week at PWSA (USA) with only very short periods of absence. During that time, Marcia cared at home for her husband, Judd, until his death from Alzheimer's three years ago. In spite of his care, her own diagnosis of Parkinson's five years ago, and ten years of chemotherapy for her leukemia, Marcia still has managed to get to the PWSA (USA) office every week. Marcia said, "*I love to come here! Everyone is so nice and wonderful.*" At the office, we know that the most "wonderful" is really Marcia!

> Rainbows are people whose lives are bright, shining examples for others.
>
> *Maya Angelou*

Shine on, shine on, Marcia ~ the world needs more people like you. ■

# A Few Words about the 2011 YAP Program

We have unfortunately reached the 60-person limit for the YAP Program due to an overwhelming response the first week of conference registration. We had to make the tough decision to close registration for the YAP Program as of August 23, 2011. We can accept no more YAP registrations.

We know that many children and adults with PWS will be very disappointed at this news, and we wanted to offer an explanation of WHY we had to place a 60-person limit on registration. It has been a struggle in the past years to coordinate and manage a large YAP program that was largely staffed by volunteers. We felt like we could and should do so much more with this program and decided to make a change going forward. In 2011 we are piloting a new YAP program, led by Jackie Mallow and Marguerite Rupnow of Prader-Willi Homes of Oconomowoc, that will be directed and staffed by individuals who work professionally with people with PWS every day. The goal is to create a safe, well-structured, and fun program that we can reproduce every year that we hold a general conference. Please accept our sincere apologies if you weren't able to register for YAP this year. ■

# Exciting New Research for Prader-Willi Syndrome

***Janalee Heinemann, MSW***
***Director of Research & Medical Affairs***

There has been a lot of interest and excitement about three pharmacological products being researched that may have an impact on PWS. One getting a lot of publicity is **oxytocin.** I know the researchers, Professor Maithé Tauber and Catherine Molinas from France, who recently published their results. Professor Tauber states, "*Oxytocin is a key hormone in building social interactions and empathy.*" and "*Two days after administration of oxytocin, we noticed that our patients had increased trust, decreased sadness and showed less disruptive behavior.*" The following is an excerpt from their published study which can be found in the *Orphanet Journal of Rare Diseases* 2011.

> *To see whether oxytocin could benefit individuals with Prader-Willi syndrome, endocrinologist Maïthé Tauber of the Children's Hospital in Toulouse, France, and colleagues injected oxytocin or a placebo into the noses of 24 adult patients. The researchers monitored the patients' behavior; they also used cartoon stories to test patients' grasp of social interactions and pictures of faces to see how well they could recognize emotions.*
>
> *For the 2 days that patients were studied after treatment, those who were given oxytocin were significantly more trusting and less sad. They were less disruptive and had fewer conflicts with others. They also had higher scores on the tests evaluating social understanding, compared with the placebo-treated group.*

I have been in touch with a researcher in Australia who is also doing a study on oxytocin. The following is from the Australian researcher.

*"The OXT neurons seem to be good candidates for playing a physiological role in ingestive behavior as "satiety neurons" in the human hypothalamus.*

- *One study provides further evidence for hypothalamic and oxytocinergic dysfunction in PWS. The associations between oxytocin, appetite regulation, and obsessive compulsive symptomatology in PWS warrant further investigation.*
- *We are currently conducting a trial of oxytocin nasal spray in PWS. We are still in the trial phase. Hopefully we will be able to form some conclusions by the end of the year."*

***~Stewart L. Einfeld, Chair of Mental Health***
***Senior Scientist, Brain and Mind Research Institute***
***University of Sydney***

Since our correspondence, the following was reported. A group of researchers from Sydney's Garvan Institute of Medical Research, including Drs. Alexander Viardot and Lisa Sze, Professor Lesley Campbell and Louise Purtell found that people with Prader-Willi syndrome experienced some significant fullness when given exenatide before food, but had no real fullness when given the placebo instead. The Prader-Willi group experienced no major side effects from the drug, whereas most from the obese-only group experienced bloating, nausea or vomiting. They state: "*Without further testing, we can't yet recommend that exenatide be prescribed for these people, unless they also happen to have Type 2 diabetes which is the recommended usage.*" These results are published in the *Journal of Clinical Endocrinology and Metabolism.*

The same researchers from France who did the oxytocin study (Tauber and Molinas) also did research on **Modafinil (brand name Provigil)** that was reported on in the last *Gathered View*. Basically, it has been shown to be safe and effective in treating excessive daytime sleepiness in PWS.

I am also keeping in touch with the research on **Exenatide which is marketed as Byetta**. It is an analogue of the gut hormone GLP-1, that is normally released after a meal. It is used as a novel daily injectable treatment for diabetes as it increases insulin secretion by directly acting on the pancreas. GLP-1 is what is called an 'incretin', i.e. increases insulin. It seems to have an advantage over some other diabetes medicines in that (like metformin) it does not cause weight gain and may even cause mild weight loss of under 5%.

It is unclear if this weight loss is just due to delaying stomach emptying (and so increased 'fullness') or also by its actions on nerve cells in the brainstem directly or indirectly via the vagus nerve to reduce appetite. A concern would be that many people with PWS already have delayed stomach emptying. So there is a theoretical risk of this getting worse with Byetta treatment in PWS, which might increase the risk of gastric necrosis associated with severe overeating.

An Australian group is doing a clinical trial on Exenatide (Byetta), the UK reported a positive case study, and Children's Hospital of Los Angeles will be looking at the effects of different obesity markers after six months of treatment with Byetta. ■

# ASK THE PROFESSIONALS:

## Newborn Genetic Screening Testing

**Q:** I would love to see PWS on the list Newborn Genetic Screening. I was told that testing for PWS is more complicated and involved than others. ***My question is this:*** Is there a current research project that would help further this in any way, such as research for a better test so that it could

**continued on page 4**

**Ask,** *continued from page 3*

be included on the screening? It just kills me that in this day and age you still hear of kids getting diagnosed at 2 1/2 years old or even later.

~Tammy Reals, Secretary, Conference Chair
Prader-Willi Alliance of New York
Mom to Greg (14), Zach (13, PWS), Emily (7), Dorothy (6)

**A: The following are the responses I received from two top geneticists.**

It would be relatively doable with PCR for methylation. The question is whether any states would be willing to fund it and whether there is an investigator willing to do a pilot study. If there are states where they routinely make DNA from the samples, perhaps a pilot study would not be that difficult. Deletions may be picked up prenatally in the future as new array technology comes into use for prenatal diagnosis. One question is whether noninvasive prenatal diagnosis becomes possible and routine.

*~Arthur L. Beaudet, M.D.*
*Henry and Emma Meyer Professor and Chair*
*Baylor College of Medicine*
*Dept. of Molecular and Human Genetics*

There has been discussion at several levels (e.g., Heartland Genetics Collaborative has discussed a pilot study) to consider and incorporate microdeletion syndromes (PWS, AS, VCFS) into the newborn screening arena, but it is not cost effective. As mentioned, the diagnosis of PWS and AS is not straightforward although methylation testing is very accurate at least for PWS.

*~ Merlin G. Butler, M.D., Ph.D., FFACMG*
*Director, Division of Research*
*Professor of Psychiatry, Behavioral Sciences and Pediatrics*
*ABMG Certified Clinical Geneticist and Clinical Cytogeneticist*

# Five Hyperphagia Grants Sponsored in 2011 by PWSA (USA)

## Transcranial Direct Current Stimulation (tDCS)

***~Dr. Merlin G. Butler and Dr. Albert Poje at the University of Kansas School of Medicine, Kansas City, Kansas; Dr. Filipe Fregni at Harvard University, Boston, Massachusetts***

This is a procedure whereby a weak electric direct current is transmitted into the brain via external electrodes without any known side effects. Previous data have shown that tDCS to the prefrontal cortex of the brain can change food craving in healthy subjects and preliminary data in 5 adults with PWS have shown encouraging results. This study is designed to test if tDCS is effective in PWS. If this pilot study is successful, it could lead to further, larger studies which if successful could lead to a therapeutic technique to reduce hyperphagia in PWS.

They will perform tDCS on 12 adults with PWS, 12 obese controls, and 12 non-obese controls. tDCS has been shown to impact craving via the prefrontal cortex, an area of known difference in PWS as shown by fMRI. Also, tDCS has been shown to moderate hunger/appetite in normal subjects, so it is possible that tDCS could have a beneficial clinical effect on hyperphagia in PWS for extended intervals after tDCS. This study is designed to test that possibility and understand if tDCS is effective in PWS.

## Leptin Resistance in Mouse Models of Hyperphagia

***~Rachel Wevrick, Ph.D., Department of Medical Genetics, University of Alberta, Canada***

The hypothesis that leptin receptor (LepR) signaling defects can cause congenital leptin resistance in PWS hypothalamic neurons preceding obesity and contributing to hyperphagia and obesity will be tested in PWS and similar genetic disorders such as Bardet-Biedl syndrome (BBS). The insensitivity to multiple hormones signaling energy needs (e.g., ghrelin) and broader phenotypes of PWS further suggests that more complex processes are affected in PWS. Defective intracellular signaling pathways may overlap with the leptin response pathways in the brain.

The aim is to examine leptin sensitivity in murine models of PWS and related disorders, including mice carrying targeted inactivation of the Snord116/MBII-85, necdin, and Magel2 PWS candidate genes, the Smith-Magenis gene Rai1, and BBS genes. The long-term goal is to determine whether defective LepR signaling is responsible for hyperphagia in PWS and related genetic disorders, and possibly contributes to hyperphagia in the general population.

The knowledge that individuals with PWS are congenitally leptin resistant would draw attention to this disorder as a model for other forms of leptin resistance, and would provide a sound and logical explanation for the severe post-weaning hyperphagia that is so characteristic of PWS.

## Brain-Derived Neurotrophic Factor in PWS & MC4R Function-Altering Mutations

***~Joan C. Han, M.D., Senior Clinical Fellow, Unit on Growth and Obesity***

**continued on page 5**

**Grants,** *continued from page 4*

***Jack A. Yanovski, M.D., Ph.D., Head, Unit on Growth and Obesity Program in Developmental Endocrinology and Genetics, Eunice Kennedy Shriver National Institute of Child Health and Human Development, NIH***

Brain-derived neurotrophic factor (BDNF) is a protein that is important in nervous system development and function. BDNF is well-expressed in the hypothalamus, a key region in the brain for energy homeostasis, and appears to function downstream of the leptin-melanocortin signaling pathway to control appetite. In both animals and humans, diminished BDNF function is associated with hyperphagia and obesity. They are studying BDNF in two hyperphagic disorders: Prader-Willi syndrome and MC4R function-altering mutations. They hypothesize that patients with PWS may have increased BDNF during infancy, followed by a decline in BDNF that precedes the onset of hyperphagia and persists after the onset of obesity. They also hypothesize that patients with MC4R mutations will have decreased BDNF, the severity of which will be associated with the degree of MC4R functional loss caused by the specific mutation(s) in each individual.

To test these hypotheses, they are conducting 2 cross-sectional studies comparing serum BDNF concentrations in: 1) 75 subjects with PWS (25 infants, 25 non-obese children, and 25 obese children) and 75 healthy control subjects matched for age, sex, race, and BMI; 2) 50 subjects with MC4R mutations and 50 healthy control subjects matched for age, sex, race, and BMI. If alterations in BDNF are found to be associated with PWS and/or MC4R mutations, these investigations could lead to future studies of BDNF receptor agonists as mechanism-specific pharmacologic therapy for hyperphagia and obesity in PWS and MC4R mutations, or BDNF receptor antagonists for failure-to-thrive in neonatal PWS.

**Hypotheses:**

1) Patients with PWS will have increased BDNF during infancy, followed by a decline in BDNF that will precede the onset of hyperphagia and persist after the onset of obesity.

2) Patients with MC4R mutations will have decreased BDNF, the severity of which will be associated with the degree of MC4R functional loss caused by the specific mutation(s) in each individual.

## Abnormal Proteins Drive the Hyperphagia in PWS

***~Barbara Y. Whitman, Ph.D., Susan E. Myers, M.D. Jeffrey Teckman, M.D., Yie-Hwa Chang, Ph.D.***

**Hypothesis:** While both genetic and brain imaging studies offer some interesting possibilities in terms of research and pathophysiologic pathways, the results of these studies are only the first step; explanatory results from either of these methodologies depend on further elucidation of the protein metabolic abnormalities. A number of genes involved in protein processing are located in the PWS region. They assert that the mechanism of hyperphagia in PWS involves signaling via serum proteins and/or peptides that are unique in type or magnitude compared to obese non-PWS patients or normal controls. Thus they are investigating this hypothesis using state-of-the-art proteonomic analysis comparing rigorously defined PWS patients, obese non-PWS patients and normal controls.

## Probing Genes for Hyperphagia in Rare Obesity-related Syndromes

***~Merlin G. Butler, M.D., Ph.D., Professor, Psychiatry and Pediatrics, Kansas University Medical Center in collaboration with Dan J. Driscoll, M.D.,Ph.D., Professor, Pediatrics and Genetics, Departments of Pediatrics, Molecular Genetics and Microbiology, University of Florida, College of Medicine, Jan Marshall, B.A., Senior Professional Assistant at the Jackson Laboratory and the Genetics Coordinator of Alström Syndrome, Randi J. Hagerman, M.D., Medical Director, University of California-Davis, M.I.N.D. Institute and Professor of Pediatrics, Endowed Chair in Fragile X Syndrome Research***

The study of rare genetic obesity-related disorders with hyperphagia including Prader-Willi syndrome (PWS), Alström syndrome (ALMS) and fragile X syndrome (FXS) allows a window of opportunity to not only provide potential insights into genetic, biochemical and developmental pathways by probing the genes for hyperphagia and obesity impacting on each rare disorder, but also applicable to the growing problem of obesity in the general population. Defining the genetic cause of obesity syndromes should enrich our understanding of obesogenic pathways in common or exogenous obesity, a major public health problem resulting in increased morbidity and mortality with severe economic burdens on healthcare systems, loss in worker productivity and decreased quality of life for affected individuals.

They propose that the three rare disorders (PWS, ALMS, and FXS) will individually be associated with unique structural and functional genetic patterns identifiable with the latest microarray technology and bioinformatics tools. Integration of structural and functional genetic profiles for PWS, ALMS, and FXS along with their clinical, hyperphagia and obesity measures, when compared with obese and non-obese comparison subjects, will enable the

**continued on page 6**

**Grants,** *continued from page 5*

discovery and characterization of a more precise molecular signature for each rare syndrome useful for diagnosis and causation. Identifying gene targets or molecular pathways common to the obese phenotype seen in individuals with rare obesity-related disorders and those non-syndromic subjects with exogenous obesity will have the potential to stimulate new directions for study using pharmaceutical interventions tailored for each syndrome with hyperphagia and obesity in general. ■

# Aerobic Exercise Capacity in Youth with Prader-Willi syndrome

***~D.A. Rubin[1], Ph.D., D.A. Judelson[1], Ph.D., D. Mendoza-Castner[1], M.S., S. Clark[2], M.D. , and M. Mouttapa[3], Ph.D., [1]Department of Kinesiology, California State University, Fullerton, Fullerton, CA; [2]Department of Endocrinology, Children's Hospital of Orange County, Orange, CA; [3]Department of Health Sciences, California State University, Fullerton, Fullerton, CA***

The purpose of this study was to compare the aerobic work capacity and heart rate (HR) responses to exercise of youth with PWS (14 subjects, aged 8-15 years) to normal weight (17) and obese children without PWS (14, aged 8-11 years). Participants were measured for body mass, body height, waist circumference, resting blood pressure (BP), resting heart rate, and body composition.

They then completed a graded cycling test until volitional exhaustion. Criteria for terminating the test was inability to keep up with the workload or the subject requesting to stop.

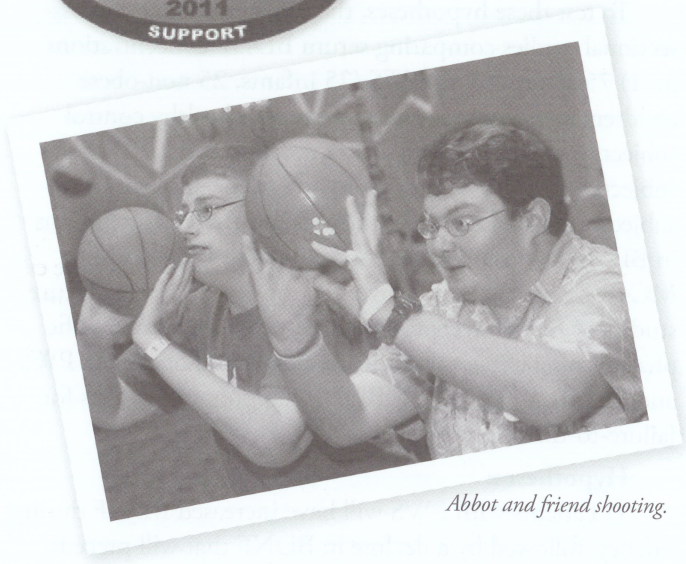
Participants with PWS attained a lower peak power and a lower peak heart rate than either of the other two groups. Those with PWS may have a lower exercise capacity. Although those with PWS exhibited similar body fat percentage to obese children, their capacity to do aerobic work was lower. However, it is important to note both that those with PWS can complete an aerobic capacity test and that there was a wide range of responses. ■

# Obstructive Sleep Apnea – reflux

In infants with PWS we have seen a lot of obstructive sleep apnea that is due to gastroesophageal reflux and thus persists even after a tonsillectomy/adenoidectomy. Sometimes the growth hormone seems to make this better (probably better muscle tone), but several of the young children that I have seen with obstructive sleep apnea that is present after a tonsillectomy have gastroesophageal or nasopharyngeal reflux. A swallowing study and/or a visit to a gastroenterologist may be of benefit to these kids because treatments are available.

I have not had much success with getting young kids to wear CPAP- -it is not a fun thing to wear. However, the best thing I have found is to let them play with it during the day (i.e., wearing it while watching TV, playing, etc.) so they get used to the feeling of it on their face. A few weeks of doing this seems to somewhat de-condition them so that they are not as bothered during the night. That being said, most kids and adults still end up taking it off during the night.

***~Jennifer Miller, M.D., Pediatric Endocrinology University of Florida, Gainesville, Florida***




*Abbot and friend shooting.*

# PWS Clinic Opens in Denver

In June the first Prader-Willi syndrome clinic was held at The Children's Hospital of Colorado in Denver. Seven patients were seen. Dr. Todd Porter, who has a niece with PWS, is the pediatrician. Drs. Jan Forster and Linda Gourash were there to lend their expertise and help kick off the new clinic. ■



# Farewell to a Friend

It is with great sadness that we bid farewell to our Executive Director, Evan Farrar. Evan has informed us that he is resigning in order to pursue a career in counseling. Evan has recently completed an advanced degree in counseling and in October will take the test to become licensed as a counselor. We thank Evan for his years of dedicated and professional service to PWSA (USA) as a Crisis Counselor and as Executive Director and wish him great success.

For the immediate future, David Crump, our Development Coordinator in Sarasota, has agreed to serve as Interim Executive Director. As you might remember from his professional resume, David was the executive director of a non-profit in Chicago prior to his relocating to Sarasota, so he brings solid experience into the interim role.

In addition, a transition team composed of the senior staff in the Sarasota office and the Executive Committee of the board is working together to manage the day-to-day operations of the organization and support David through the transition period.

In spite of the departure of a beloved employee, our organization is strong, from both the perspective of committed staff and financial performance, and is fully functioning and fully committed to continuing and advancing the mission of serving individuals and families affected by PWS. After our National Conference in Orlando in November, the board will begin the search for a new Executive Director.

Registration for the National Conference officially opened on August 15th, and the response has been very strong. The conference agenda, we believe, will be the best we have had yet in terms of scope and depth, with a wide range of presentations, workshops and social events to educate, inform and support our families, professional providers and research community.

And, of course, we are all looking forward to our conference keynote speaker, Clint Hurdle, manager of the Pittsburgh Pirates, parent of a child with PWS and strong supporter of PWSA (USA) as our national spokesperson.

Looking forward to seeing you all at Conference. ■

*The Executive Committee of the Board of PWSA (USA)*

# Cognitive Changes in the Aging Adult with Prader-Willi Syndrome

***By Lizabeth Moser, M.S.W., L.C.S.W. - Psychotherapist with Prader-Willi Homes of Oconomowoc, WI***

*(This is the third in a series of three articles that shares information about aging in adults with PWS with a focus on dementia.)*

With the increased awareness of Prader-Willi syndrome (PWS) leading to early diagnosis and treatment, life expectancy has grown dramatically over the years. When I first began working with adults with PWS in 1989, I learned that many did not live past their teen years due to obesity-related medical complications. Back then, I worked with 15 people diagnosed with PWS, most in their early twenties.

Twenty-plus years later, I am thankful to still work with many of those individuals, as we step into our forties together. We are entering the uncharted years of aging in PWS with the aim to achieve the same outcome as the frontier movements of awareness--early detection and treatment--to ensure continued quality of life.

This article explores how aging may influence cognitive processes in PWS and how we can prepare for future needs.

## What is Dementia?

According to Wikipedia, "dementia is a serious loss of cognitive ability in a previously unimpaired person, beyond what might be expected from normal aging." There are several types of dementia, but the most common is Alzheimer's (AD), which occurs in "50-80 percent of dementia cases." It is a progressive disease with symptoms getting worse over time (www.alz.org).

## Dementia Prevalence:

In the general population, Alzheimer-type dementia usually occurs after the age of 65 (alz.org). Dementia studies are not as abundant in the area of intellectual disability (ID) syndromes compared to the general population studies, but in March 2009, the State of Science on Dementia released a comprehensive review of studies from 1997-2008 relating to aging and intellectual disabilities.

This 2009 review reported that "the prevalence of dementia (particularly AD) among the ID population may differ from the general population, at least in specific subgroups such as Down syndrome (DS)" (Zigman, Schupf, Haveman, & Silverman, 1997). Later studies showed that the prevalence of dementia in the ID population was 6.1% in those aged 60 and over, which is comparable to the percentage in the general population (Janicki and Dalton 2000). The same study showed that adults with DS had much higher rates of dementia--56% for those 60 and older.

## The mean age of dementia onset:

- General population: 67 years
- Intellectual Disability excluding DS: 67.2 years
- Intellectual Disability with DS: 52.8 years

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**Aging,** *continued from page 7*

• Intellectual Disability with PWS: Unknown

## PWS Research Overview:

The State of Science on Dementia cited one PWS study by Sinnema, Maaskant, Van Schrojenstein, et. al. in 2008, which evaluated 74 individuals with PWS ages 18-63 years old and reported no cases of dementia.

At the IPWSO conference in Taiwan last year, several studies examining aging in PWS were presented. Whittington and Holland reported the results of their study Recent Mortality Rates and Risk of Dementia in PWS. They found that out of the 26 individuals who were 40 years and older: 22 with no signs of dementia, one case of mild-moderate dementia, one potential case of mild dementia and evidence of cognitive decline in one person.

Sinnema and her research team from the Netherlands presented a case study of a 58-year-old woman with PWS. The assessment scores supported "the presence of dementia in very late stages."

The researchers agree that more studies are needed to understand how aging will affect older persons with PWS.

## Preparing for the Future:

Prader-Willi Homes of Oconomowoc (PWHO) currently supports 81 adults in residential care: 27% are in their 30s, 33% are in their 40s, and 2% in their 50s. One woman was diagnosed with dementia almost three years ago at age 54.

According to the article *Stopping Alzheimer's Before it Starts* posted on www.medicinenet.com, "the prevention process should begin at approximately age 40. That is because on the average, Alzheimer's disease begins 30 years before the first symptoms appear." Because it is unknown when onset may occur, PWHO has taken the approach of evaluating cognitive changes over time with the aim of early detection and treatment.

**The fundamentals of this procedure are as follows:**

1. All clients have a mental status baseline score from "The Short Portable Mental Status Questionnaire" (SPMSQ).

Date of assessment and cognitive range (normal mental functioning, mild, moderate or severe cognitive impairment) corresponding with score on the questionnaire are documented.

2. All clients thirty-five years and older complete the SPMSQ annually. If there is a change in range, further evaluation occurs (i.e. retesting, additional assessments, psychotherapist consult, psychiatrist consult, neurologist consult).

3. Clients that present with any significant behavioral and/or mental health changes complete the SPMSQ to assist in determining the possible cause(s) of decline in functioning.

4. If cognitive range declines and other medical or mental health causes have been ruled out, more in-depth assessments that measure cognitive and daily functioning are administered.

To date, the assessment scores have shown no indication of early onset of dementia in the 30s or 40s.

In diagnosing dementia and evaluating potential risk for early onset, an important contributing factor is family history.

## Treatment:

The National Institute on Aging (NIA) has the following statement on their Web site, www.nia.nih.gov:

AD is a complex disease, and no single "magic bullet" is likely to prevent or cure it. That's why current treatments focus on several different issues, including helping people maintain mental function, managing behavioral symptoms, and slowing AD.

Visit www.webmd.com under Alzheimer's Disease: Daily Care of the Alzheimer's Patient for support recommendations. One of the most important suggestions for caregivers is "to understand and act according to your own physical and emotional limitations. Be sure to take care of yourself, and allow yourself periods of rest and relaxation." Other factors in caring for somebody with AD are exercise, nutrition, socialization, structure, routine, and visual cues. As a PWS community, we are already one step ahead of managing the symptoms of dementia, as the approaches are similar to the ones effective in supporting persons with PWS.

In addition to daily supports, medication has also been used to manage and potentially slow down AD. The State of Science on Dementia reported, "Donepezil is the most commonly used anti-dementia drug used in intellectual disabilities to treat dementia, and there is some evidence for rivastigmine." The woman diagnosed with dementia residing at PWHO has had a positive response to receiving donepezil (Aricept).

As treatment strategies are planned, it is important to remember that dementia is a progressive disease in that "lost skills cannot be regained" (www.webmd.com). It is crucial to incorporate and continue with activities that are physically and mentally stimulating per individual's preference.

## Summary:

More studies are needed to understand how the aging process will affect persons with PWS. There is not enough information to make any conclusive statements about how or when dementia will present.

Based on my review of the literature and observations at PWHO, there is no evidence to suggest that dementia in PWS is comparable to Down syndrome with the potential age of mid-30s onset. Further research is needed to evaluate whether or not the onset could occur earlier than other ID syndromes and the general population.

Like PWS, dementia has no cure, but with early detection and the necessary supports in place, we can

**continued on page 9**

## Fundraising

*By Jodi O'Sullivan*

These incredible fundraisers invite others to hear the story of Prader-Willi syndrome and offer a way for them to help. With these personal events, more understanding and compassion is grown, and communities in different areas of the country effectively unite for a common purpose…our loved ones who have PWS. We thank each and every volunteer and participant, and we invite others to tell their stories by hosting a benefit event of their own for PWSA.

*Thank you to* **Al Heinemann**, father of **Matt** (38, PWS), who conducted an ***On The Move*** event, a **Personalized Fitness Lose-A-Thon**, from May 1 to June 30. Al's gym in Florida, Personalized Fitness, opened this event to its members and further contributed $10 for every pound lost, raising $1,225 for the crisis program.

*Thank you to* **Justin and Cynthia Wilson Loeb** of Texas, aunt and uncle of **Emerson Parker** (2 in October, PWS), who raised over $16,600 for PWSA. Cynthia and Justin, who own **Oliva Italian Eatery**, decided their restaurant would donate a portion of sales in May of their chef's feature. Mixing their main goal of PWS awareness with love, they cooked up an impressive fundraiser recipe.

*Thank you to* **John and Lori Lens**, parents of **Hunter** (13, PWS), for hosting an ***On The Move*** event. Their **2nd Annual Hunter Lens Golf Tournament for PWS** on May 21 in Connecticut netted over $11,500. That's no mulligan.

*Thank you to* **Sybil Cohen**, mom to **Rose** (16, PWS), from New Jersey whose birthday wish was that, in lieu of gifts, donations be made to PWSA (USA). Her wish came true with $185 in birthday dollars.

*Thank you to* **PWSA Indiana** who, after just reestablishing themselves this year, conducted a successful ***On The Move*** event! Their **PWS On the Move 5k Walk/Fun Run** on May 21 raised over $20,000 for their chapter and PWSA (USA). Talk about a walk in the park!

*Thank you to* **Aaron Carvajal** (13, PWS) from Texas, who in May celebrated his **Bar Mitzvah**, the attaining by a boy of his religious adulthood and responsibility. He chose on this occasion to perform a mitzvah, or good deed, by asking that donations be made to PWSA, resulting in contributions totaling $231. Mom **Susan Fisher**, could not be more proud.

*Thank you to* **Fran Baehr** from New York who in May sold **PWS Awareness bracelets** in honor of her son **Raymond** (6, PWS) at the West Babylon School District, raising $265.

*Thank you to* **Kerry Headley** and work colleagues of the late Jay Headley for organizing the **Jay Headley Memorial Golf Tournament to Benefit PWSA (USA)** on June 3 in Ohio. Jay passed away in November, survived by wife, Kerry, and son, **J.R.** (12, PWS). The event raised $3,330.

*Thank you to* mom **Michele Shingleton,** and **Shawna Bush** who planned their **5th Annual PWS Walk-A-Thon in Honor of Carter Shingleton** on June 11 in Connecticut, raising over $5,000. Despite buckets of rain and cold cloud-cover, reported Michele, over 100 people came out to walk in honor of **Carter** (10 in October, PWS).

*Thank you to* the dynamic duo, **John Boughton and April Boughton** (22, PWS), of Indiana who rode in the **Great Ohio Bicycle Adventure** in June, a week-long bicycle adventure covering at least 50 miles a day. Although the father and daughter team had registered for the event many times before, this was their first time doing it as a fundraiser for PWSA (USA). They raised over $2,000, with half designated for research. Even more, several Ohio families affected by PWS met up with them at rest or camp stops to cheer them on. ■

## Tax Free Charitable Rollover Gift (May Expire Soon)

Did you know that ***if you are 70 ½ years old*** or older, current law allows you to donate, *tax free*, up to $100,000 from your Individual Retirement Account (IRA) to a public charity? This is one creative way you can make a significant gift to support the resources, services and research carried out by PWSA (USA). Although Congress is considering an extension, the current law expires at the end of this year. So it is important to act soon. If this is something you are considering, be sure to consult with your financial advisor. ■

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**Aging,** *continued from page 8*

provide the best opportunity for the physical, mental and emotional well-being of the individual.

As we make this venture into unknown territory together, we will continue to do what the PWS community does best--unite forces--by sharing experiences, contributing to the knowledge base, applying what is successful and growing together--stronger and more informed than the day before. ■

*[Don't miss Barb Dorn's review of the book Prader-Willi Syndrome: Growing Older, by Terrance James, Ph.D., which will appear in the November/December issue.]*

## Fundraising

**Tax Free,** *continued from page 9*

# Employer Matching Gifts

Earlier this year, PWSA (USA) received a gift of $7,000 from one of our members. A few weeks later we received an additional $14,000 – a double match – from that member's employer. Many employers will match your charitable contribution to PWSA (USA) with an equal or higher amount, if you are an employee, retiree, or board member of a company, or if your spouse is. This is another creative way for you to **increase the impact of your own giving!**

PWSA (USA) is a 501(c)(3) nonprofit organization, an important qualification for many companies' matching gifts programs. Check to see if your company has a matching gifts program. If so, please request a matching gift form (or sometimes an online application process) from your employer and send the completed form or information with your gift. We will do the rest! ■

# A Creative Opportunity for Federal Employees

Combined Federal Campaign

Celebrating its 50th Anniversary this year, the **Combined Federal Campaign** is "the largest workplace charity campaign in the United States." It provides a creative opportunity and channel for employees of the federal government in the U.S. to contribute, through their workplace, to qualified charities of their choice.

PWSA (USA) is qualified to receive contributions through the CFC. If you or someone you know is employed by the federal government, they can designate contributions to PWSA (USA) through this campaign, supporting the services and resources provided by the Association, helping fund research, and also raising awareness of PWS.

**The PWSA (USA) CFC ID Number is 10088** ■

*The Mission of PWSA (USA) is to raise awareness, offer support, provide education and advocacy, and promote and fund research to enhance the quality of life of those affected by Prader-Willi syndrome.*

*If you have questions about the creative giving opportunities covered here or about other ways you can support and help make this mission a reality, please contact PWSA (USA)'s Development Coordinator, David Crump, at 941-312-0400 or at dcrump@pwsausa.org.*

CHUCKLE CORNER

*Silver Sparrow* by Tayan Jones quote:

"People say, 'That which doesn't kill you makes you stronger'. But they are wrong. What doesn't kill you, [just] doesn't kill you."

## Moms Speak Out

*The next several pages will contain articles written by mothers of children with PWS. They offer hope, observations and sage advice, not just about the children but also for their parents.*

# Independence Day

***By Linda Huckelberry, Maryland Heights, MO***

Months before my daughter Natalie was diagnosed, I first heard of Prader-Willi syndrome from a doctor who described it dramatically with images of an out-of-control monster, clawing at cabinets and screaming nonstop to be fed. Honestly, that was the description given to me. Obviously, when Natalie was ultimately diagnosed with PWS, I was devastated. I remember looking at my gentle, sweet-natured, happy, loving baby and imagining that she would eventually slip away from me and morph into the description of PWS given to me.



*Natalie*

While PWS has definitely been a challenge for us, it seems to dole itself out into mostly manageable servings. We started off with what seemed like endless therapy sessions and doctors appointments. That gradually changed into school evaluations and IEP meetings. Sprinkle some time spent in hospitals throughout Natalie's life for back braces, spine surgeries, gastroparesis, etc. Slowly, behavior issues, elopement, OCD and skin-picking crept into our lives and were met with visits to psychologists, psychiatrists and massive numbers of band-aids. Now, at age 12, we are approaching the teenage years, middle school, and puberty and hormone therapy.

Natalie's biggest challenges never seemed to be food related. I figured we managed to dodge that bullet. She was gradually becoming more persistent, hungry and even aggressive when it came to food, but still manageable. Last week after a couple of intense episodes involving food, I mentioned to my husband that we should think about locking up the food. I told him that even though she's not a food seeker, I was aware of kids that were relieved to have food locked up so it's not a cause of stress. At that point, Natalie began crying because she felt we didn't trust her. I told her we didn't have to and I was just trying to think of ways to help her.

Imagine my surprise an hour later when Natalie timidly told me, "Mom, maybe we should lock up the food." She went on to tell me that she had snuck food on one occasion. She emphasized over and over that she only did it once. I praised her for telling me the truth and being such a smart

**continued on page 11**

**Independence,** *continued from page 10*

girl! I told my husband about Natalie's request, and he heaped on the praise. She immediately called Grandma and Grandpa to revel in their admiration, too. More than ten years after Natalie's diagnosis, the decision to lock up food wasn't a result of trying to protect an out-of-control child, but instead, it was the result of a request from a sweet, smart young girl determined to stay healthy.

We went to the hardware store together to make our purchase. Locking the refrigerator was simple, but the pantry would require a little more work so I was going to wait for help from my husband. When Natalie noticed the locked refrigerator and un-locked pantry, she demanded to know, "Why isn't the pantry locked? How are we going to keep me safe?" I managed to install the lock on the pantry, still somewhat in disbelief that *she* was the one so adamant to lock up the food.

Later that night as Natalie watched fireworks on TV it occurred to me that Independence Day had extra significance for her this year. This was the year she gained independence and freedom from the anxiety an unsecured kitchen had caused her. ■

# PWS Community = Room for Everyone

***By Jennifer Bolander, Fairview Park, Ohio***

I remember the moment I was told that our daughter Sophie had Prader-Willi syndrome. I had never heard of Prader-Willi syndrome before, neither had my husband, neither had any of my family.

We weren't alone, though. I soon made the call to PWSA(USA) and felt like we had found a source of hope for us and for Sophie. We had found our new "family", our new community that extended around the globe.

At this point I can't begin to accurately count how many families I have met because of our membership with PWSA(USA). These families have come to feel like my family. This is a community where we all "get it". We all know what's involved in living with PWS. We all know there are good days and bad days. We all know, I think, that being a special-needs parent does not automatically mean we are "Perfect PWS Parents"…rather it means we all try hard to give our children good, healthy lives, and that sometimes life isn't always going to cooperate with those efforts.

To me, the word "community" implies inclusion of all realities and all perspectives. In the PWSA(USA) community there is room for all kinds of families. There is room for parents whose child is in a healthy, stable phase of PWS, and for parents and families where life is *not* calm and the family is struggling. There is room for families whose child never eats fast food, and for families where the parents are okay with the occasional fast food or restaurant meal. There is room for families where the parents have decided the whole family will be vegan/gluten-free/low-carb…and for families who have found success without doing those things. In this community, in my opinion, it is okay for PWS parents to vent, to be open about the sometimes horrifically bad days PWS can cause for an entire family. It is also okay to brag about the successes of a child with PWS, because we know those successes are hard-won.

While I would never overload parents of a newly-diagnosed child with details of every difficult situation PWS can cause, I am not going to sugarcoat anything. I think every PWS parent has the right to say "hey, this is really difficult, we are having an awful time right now" if they need to. Some days, PWS does stink. If it was easy and our children never suffered, then we would not have much to feel frustrated about.

But I firmly believe that even as we all fiercely love our children, that love can coexist with honesty about how difficult living with PWS can be. I believe in keeping it real, in telling it like it is, when appropriate, and to those parents who are ready to hear it. Every PWS family out there IS going to face their own variety of PWS challenges….that is the nature of the PWS beast.

It is okay to comment on those challenges. I think the last person who should ever shut down the venting comments of a PWS parent is…another PWS parent. If a parent who is going through a hard time with their child with PWS is "scolded" for talking about their less-than-sunshiny experiences, what's the consequence? That parent is made to feel alone, as if they are doing something wrong with their child, as if the complex challenges of PWS are controllable and anything less than calm in their house must be due to their bad parenting.

As special-needs parents, we should support each other on the hard days--and cheer each other on--on the good days. ■

# Waiting for the other shoe to drop

***By Kristi Rickenbach, Blaine, Minnesota***

Sometimes parents of kids with PWS talk about how worried they are for the "other shoe to drop"--the first shoe being the diagnosis, the other the many challenges that come with PWS. This shoe comes in many shapes and sizes and, at times, can become so consuming that we forget everything

**continued on page 12**

**Waiting,** *continued from page 11*

else. We just sit and wait, analyzing anything different that our kids do, fearing that this is the day. To some "the shoe" is a fear of the meltdowns and temper tantrums; to others it may be food seeking or skin picking. We all have our "shoe" that we fear; overcoming this fear is difficult, but crucial. I have known this fear, and I have succumbed to it on many occasions. Until one day it was all so clear, I was missing out on so much while waiting in fear.

The joy that I had with all of my other children at times seemed to be nonexistent with Justice Faith. I read all I could about PWS and tried to imagine my life in the shoes of those who had already walked this lonely road. I would get lost in dark thoughts of what would become of my beautiful baby. I worried so much about the future that I lost the present. My sadness, grief and fear had taken away precious moments that I should have been enjoying with my baby, moments I can never get back.

It was at a national conference in Florida when my head cleared and my fear no longer consumed my every thought. I met so many amazing people who taught me to live for today and enjoy every moment with my child. I no longer wait for the other shoe to drop. We live every moment to the fullest and cherish each day. If I could, I would go back in time and redo all those desperate moments when Justice was a baby. I look at her now, and I wish I could have had a crystal ball to see into the future so I could see how amazing she was going to be.

I can't get back the time I lost, but I can encourage others to live every day to the fullest and not miss out on one precious moment. We don't know for certain the future of our kids or when the other shoe will drop. We do know that time passes quickly and children grow fast. Cherish every moment with your child and don't let fear keep you from truly enjoying your amazing, beautiful child. ■

# Sexual Interests and Hypogonadism in PWS

***By Harry J. Hirsch[1], Talia Eldar-Geva[2], Fortu Benarroch[3], Orit Rubinstein[1] and Varda Gross-Tsur[1]***
***The Israel Multidisciplinary Prader-Willi Syndrome Clinic***
***[1]Neuropediatric Unit, Department of Pediatrics, and [2]Reproductive Endocrinology and Genetics Unit, Department of Obstetrics and Gynecology, Shaare Zedek Medical Center, Jerusalem, Israel; and [3]Department of Child and Adolescent Psychiatry, Hadassah Mount Scopus Hospital, Jerusalem, Israel***

*Note: This study was partially funded by PWSA (USA).*

**Background:** Hypogonadism is a major feature of Prader-Willi syndrome, but clinical manifestations are variable. Sexual interests and behavior in this population have not been previously described.

***Objectives: We studied PWS adolescents and young adults to assess***
(1) satisfaction with physical and sexual development,
(2) frequency of romantic and sexual experiences,
(3) aspirations and expectations regarding marriage,
(4) investigate the relation between sexual interests and hormone levels, and
(5) assess the desire for hormonal replacement therapy.

Methods: The study population consisted of 27 individuals (13 males) ages 17 to 32 (mean 23.5) years with genetically confirmed PWS. Mean IQ was 75 (range 50 – 100). We conducted structured interviews using questionnaires specifically designed for this study.

**continued on page 13**

## Counselors Corner

# Document, Document, Document!

That's what we all hear over and over, but it is true. When you have a child with PWS, it starts with documenting the diagnosis and sub-type and continues for the rest of the individual's life. It's difficult keeping all those records, but it is essential.

At PWSA (USA) we have been working on different forms to help with this documentation. On our website we have samples to help document for SSI, a school portfolio that not only documents a child's progress from year to year but also supplies teachers and parents with valuable handouts, and a Care Notebook to document all the medical contacts, lab work and medications record our children and adults experience. Make use of them, and make your record keeping easier.

If you have any questions about these samples, just call and ask for a crisis counselor. Until next time. ■

*~Kate Beaver, Alterman Crisis Counselor*

**Sexual Interests,** *continued from page 12*

***Results:***

- There was a significant negative correlation between IQ and body image in both males and females.
- IQ showed a positive correlation with interest in dating and romantic activities.
- Approximately half of PWS males and females reported having gone on a date and kissing romantically.
- All males and 64% of the females wished to be married.
- Seventy-seven percent of PWS males wanted hormonal treatment to increase phallic size.
- We found no correlation between hormone levels and sexual interests.
- Only 43% of PWS females wanted hormonal medication to achieve regular menstruation

***Conclusions: Despite documented hypogonadism, PWS young adults are interested in sexual and romantic issues. The range of sexual activities and expectations is variable. Understanding specific sexual characteristics of each individual is important in order to offer proper anticipatory sexual guidance counseling and for appropriate recommendations for hormone replacement.*** ■

*[Parents and caregivers are encouraged to send comments on this important topic.]*

# Let's Talk about SEX

***By Linda Thornton***

In the past we swept the subject of sex under the carpet. We knew that women with PWS menstruated infrequently, if at all, and that males were infertile. In those days, sex hormone therapy for males and females wasn't really an option, so the whole chapter of sexuality was closed for many years.

It was rudely opened in 1999 when the first birth of a healthy baby girl to a woman with PWS was recorded in Sweden. This was followed in 2001 when a 32-year-old woman with the deletion diagnosis was recorded as having given birth to a baby with Angelman's Syndrome in Denmark. The third recorded birth was in New Zealand in 2004 when a baby girl, also with Angelman's syndrome, was born to a young woman with a genetic deletion diagnosis.

Sex hormone therapy is now recommended for both males and females with PWS. This "encourages the development of secondary sexual characteristics and potentially improves bone mineral content and density."

It will also induce menstruation and the likelihood of pregnancy. This means greater observance of lifestyle, good sex education and support, as well as a better understanding of how to cope with the resulting birth. It is also recommended that males have testosterone therapy, and many youngsters are on growth hormone therapy and may well continue to be on GHT into adulthood. Combined with sex hormone therapy, could this mean an increase in fertility? What about sex itself? Marriage, even?

Back in 1999 I visited a residential home in Seattle where a couple (both with PWS) were engaged to be married. I asked the young man why he wanted to marry, and his reply has stayed with me ever since…"because I want to see her in the shower".

Many of our young people want to experience a close relationship, to "have" a boy or girlfriend, and nearly every young woman with PWS that I have met expresses a desire to have babies, to look after babies and to have them to cuddle and hold. Most are genuinely wonderful with small children and babies.

However, I know from talking with the young woman's family in New Zealand, that there were many difficulties involved, even from the beginning when no one knew she was pregnant until a visit to the doctor in her last trimester brought the surprising news. Birth was by caesarian section as they did not think the mother would be able to tolerate a natural birth. The mother was overjoyed to have her own baby, but not so overjoyed when the baby cried and interrupted her dinner time. She did not prove to be a natural mother, able to care for a small baby or answer its constant demands. She did not have maternal instincts.

Somewhere along the line there has to be an extra element of responsibility and care from parents and caregivers alike. There still needs to be educated and intelligent management and support around such important decisions as choosing to share your body or life with another. There have been many changes in the way we care for and manage the lives of those with PWS, and doubtless there will be further improvements as we understand the needs of those with disabilities.

At the end of the day I expect every parent will share the same thought, regardless of whether there is a disability or not; "I just want my son/daughter to be happy", but let's make sure we know what they want, and how we can ensure that happiness and support. ■

*~ Linda Thornton, from New Zealand, is the mother of an adult daughter with PWS and the Secretary of the International Prader-Willi Syndrome Organisation.*

**Production, printing, and mailing of this newsletter was underwritten by a generous grant from Eastside High School student-sponsored "Spirit Week" Fundraiser in Greenville, South Carolina.**

# Contributions

**Thank you for Contributions in June and July 2011*.** We try to be accurate in recognizing contributions above $25, and apologize for any errors or omissions. If you notice an error, please tell us.

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**Correction: Last issue's Contributions were received in April and May 2011*

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Prader-Willi syndrome (PWS) is a birth defect identified in 1956 by Swiss doctors A. Prader, H. Willi, and A. Labhart. There are no known reasons for the genetic accident that causes this lifelong condition, which affects appetite, growth, metabolism, cognitive functioning and behavior. Prader-Willi Syndrome Association (USA) was organized in 1975 to provide a resource for education and information about PWS and support for families and caregivers.
PWSA (USA) is supported solely by memberships and tax-deductible contributions. To make a donation, go to www.pwsausa.org/donate

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**Our Mission:** Prader-Willi Syndrome Association (USA) is an organization of families and professionals working together to raise awareness, offer support, provide education and advocacy, and promote and fund research to enhance the quality of life of those affected by Prader-Willi syndrome.

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**Deadlines to submit items to *The Gathered View* are: Dec. 1; Feb. 1; Apr. 1; June 1; Aug. 1; Oct. 1**

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# ATTENTION!

Calling all SURVIVORS OF PRADER-WILLI SYNDROME!

## It's NATIONAL CONFERENCE TIME!

November 11-13, 2011 ~ Orlando, Florida

### In a Nutshell-

*(Check the PWSA (USA) website for more details.)*

### ✔ Registration opened August 15. You can:

- register online at www.pwsausa.org/conference/2011 or
- get the information on our home page at www.pwsausa.org or
- register by mail by calling 800-926-4797 to request a registration packet.

### ✔ Make hotel reservations at the Buena Vista Palace Hotel & Spa

- by calling the hotel reservations department at 1-866-397-6516 – mention PWSA (USA).
- Rates until October 23, 2011: $119 a night for a single or double room, $139 for a triple and $159 for a quad room, plus tax and resort fee. These rates are available for the 3 days before conference begins and 3 days after conference ends.

### ✔ Conference costs

- Conference registration is $225 per person until 9/30 and $250 after 9/30. This fee includes breakfast and lunch Saturday and Sunday. Companion registration fee, which includes the two days of meals, is $100, for those not attending the lectures.
- Saturday night Gala Dinner will be a ***separate cost.***
- YIP Infants (under age 2) $100 – **capped at 20**
- YIP Children (ages 2-9) $125 – **capped at 50**
- YAP Participants - $175 – **filled; Registration closed**

***(See Page 2)***

Siblings (ages 7-15) - $125 – **capped at 30**

We know that many of you were disappointed that the YAP (Youth and Adult Program) registration closed so quickly. We're working to meet your needs. In an effort to include as many families and children as possible in our November conference, we are increasing the age limit on the YIP (Youth and Infant Program) from 0-6 to 0-9. The children ages 6-9 will not be in with the toddlers, but will be engaged in fun, age-appropriate activities for them. If your child is in this age range and you wish to register them for the YIP, there are a limited number of spots available so don't delay registering!

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Reminder – stay informed and stay current with PWSA (USA)'s free weekly e-Bulletin.
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