The Gathered View

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

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

Sandpaper

By Diane Seely, Plain City, Ohio

Reagan

It's another start to a summer day, weather perfect outside. Inside, a storm is in the making. I remind myself that summer is always difficult for my son, Reagan. I suppose that it is to some extent for most any child, typical or "not" so typical. We are part of the "not" so typical crowd.

The schedule has changed, and without his usual routine and structure, it has him uneasy, his anxiety all ramped up. He needs predictability, to know what will be expected from him today. But today I don't have answers to his twenty questions… "Who's coming today, mommy?"(referring to one of his aides). "Do I have camp today?" The questions start as soon as he wakes up. It's always so random… "Is Sam retiring, too? Will I be able to see the Wiggles Live Show again?" In my morning fog I try to explain it all once more… "Well, if Sam is retired…What do you think? Will you be able to see them at a show?" Reagan: "No."

Silence as he tries to process this piece of information. I gently remind him that right after breakfast he needs to get ready for camp and to wear the shirt provided for the field trip today. I yell up the steps to him: "Reagan, you need to wear this shirt, so find a pair of green shorts to match." He holds up a pair of shorts way too small, but they are green, so I'll give him that. I try to kindly explain he cannot wear those shorts, pick another pair. Five minutes, that's all I need to finish my cup of coffee and my toast, for the love of…

In his room I take out several pairs of shorts and show him any of these will work; which one? "But, mommy"… and the tears begin… "You said they needed to be GREEN"…! Yes, I did say that…what am I thinking? In his world, you say what you mean, and mean what you say. "I'm sorry, Reagan; these are ok. They don't have to be green. I shouldn't have said that"…still more tears…"But I don't understand, we always match"…Stay calm, stay calm…I get out his socks. "Wear these socks, please"…Reagan pulls out a pair of old socks, "Why can't I wear these?"…"Well, because they are old, and you need to wear the new bright white ones." Here we go again…crying loudly now…"But why do we have them if I can't wear them?"…Good question!

Downstairs ten minutes later: He has a birthday coming up; he'll turn nine. We have been working on convincing him to pack away the Wiggles and some of his toys that are too young for him. We started a pile last night, and he seemed ok with it. This morning, well…not so much! I want for him to have friends over and have things for them to do that are age appropriate. What I want is for him to grow up, but that reality scares the hell out of me…because I don't know what that will look like for him.

I begin to second guess what I'm trying to accomplish. Part of me worries that perhaps he is a hoarder, and I try to think back…Do I recall reading anything about PWS and hoarding? Have we fed into it, by keeping all of his toys? Have we allowed him to become too complacent in his comfort of these things that he holds so dear? Is this some sort of reality vs. fantasy thing that he needs to have in his life? I just don't know what to think…

I try to halt my racing thoughts and look at this little boy standing in front of me with big crocodile tears streaming down his face. He does not understand why he can't have them, why we need to pack them all away. Even the idea of getting new toys does not excite him; he's perfectly fine with what he has right now.

The Golden Granddaughter

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Volume 37, Number 5 ~ September-October 2012 ~ Our 37th Year of Publication

IMPORTANT!

Attention All PWSA (USA) Members

The PWSA (USA) Board of Directors is inviting all members to participate in the annual membership meeting via conference call

Monday, October 29, 2012, at 8 p.m. EDT.

Please join us!

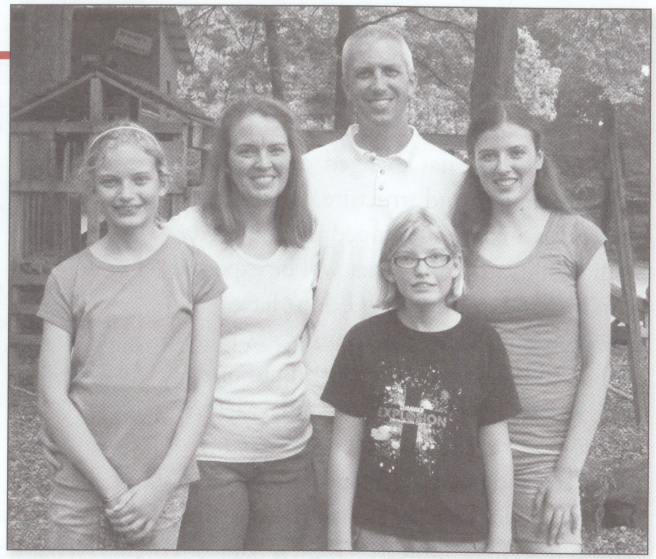
The call in number is 213-493-0800, access code is 1076930#.

Interim Executive Directors View

The Beauty of Prader-Willi Syndrome

On September 12, 2011, my husband Dale, daughter Shawn and I had the opportunity to attend the Georgia annual fall picnic for families with children affected by Prader-Willi syndrome. While we were ostensibly there to represent the PWS-Georgia Association Board of Directors, we really went to soak up the beauty and energy of the younger families and their precious children. What we received in return was even more than we had hoped for.

Almost 100 people were in attendance at the event, hosted by Neal and Angie Spradlin. Their family is so personable and attentive that we felt instantly welcomed, even though Shawn, who has PWS, was definitely older than the targeted age group. The weather and the Spradlin homesite were both perfect – adding to the beauty of the day. The children with PWS and their siblings were beautiful – both inside and out. The babies stole my heart, and the older children stirred my soul - so happy and loving and active! From the horse barn, to the chicken coop, to the rabbits, to the garden, to the playground, to the hay rides, there was plenty for them to explore, and they were clearly enjoying every minute of it.



Neal and Angie Spradlin with Brooke on left (curly blonde hair), Christina in front (PWS), Amy on right (dark hair)

Dale and Dottie Cooper

Parents loved letting the children play, picnicking on blankets and lawn chairs across the spacious farm, getting to know each other and their families, and chatting about the latest treatment or sharing ideas on what's working for them. It was most inspirational to see these young families tackle with hope and optimism the challenges of PWS which seemed so scary and overwhelming to so many of our families 20-30 years ago when our children were young. It was great spending time with other families who "understood", were flourishing and doing well, seeming to handle PWS as just one of many daily challenges that affects us all. If there is a legacy that we who have older children with PWS can be proud of, it is that yes – we can survive and survive well.

I left the picnic feeling inspired. We found that these young families are committed and active – strong advocates for our children. How beautiful is that? It is very important our families continue to work together to advance the research and treatment options available to our children impacted by PWS. The Spradlins have shown us a beautiful example of how one family can really make a difference. ■

~ Dottie Cooper

Announcing PWSA (USA) 2012 Grants Awarded!

Nutritional Aspects of Prader-Willi Syndrome and Childhood Obesity: Understanding the Shift from Failure-To-Thrive to Hyperphagia

By Daniel J. Driscoll, M.D., Ph.D. – Principle investigator & Jennifer Miller, M.D., M.S. – Co-Investigator

The mechanism(s) governing the shift from poor appetite and failure-to-thrive to development of obesity and hyperphagia (abnormally increased appetite) in Prader-Willi syndrome (PWS) remain unknown. Ghrelin and oxytocin have been suggested to be involved in the pathogenesis of hyperphagia in PWS while dysfunction of the hypothalamic melanocortin signaling pathway remains an unexplored possibility. The goals of this grant are:

Aim #1 will evaluate mediators of the hypothalamic leptin-melanocortin signaling pathway in individuals with PWS. Serum α-MSH levels in young and old patients who have PWS will be measured via enzyme-linked immunosorbent assay (ELISA) and compared to age-matched normal control subjects and non-PWS subjects with early-onset morbid obesity (EMO). Proopiomelanocortin (POMC) mRNA expression in young and old PWS patients will be assessed via quantitative real-time PCR (qRTPCR) and compared with age-matched normal control subjects and EMO subjects.

Aim #2 will assess oxytocin levels in serum of individuals with PWS in various nutritional phases. Serum oxytocin levels in young and old PWS patients will be measured via ELISA and compared to age-matched normal control subjects and non-PWS subjects with early-onset morbid obesity (EMO).

In the long term the utility of knowing the hormonal and metabolic factors involved in the transition to each phase will then give scientists incredible insight into the pathophysiology of obesity in PWS and therefore lead to rationale treatment strategies (e.g., hormonal supplementation or blocking). In addition, insights into the causes of obesity in PWS will undoubtedly make valuable contributions into other causes of obesity in the general population. The development of biomarkers that can predict presymptomatically the development of obesity later in childhood would be invaluable, especially given the epidemic of obesity that is happening nationally.

A Pig Model for Hyperphagia and Obesity in PWS

By Robert Nicholls, D. Phil – principle investigator

Overcoming the intense drive to overeat and subsequent obesity that provides the greatest threat to life quality and expectancy is of upmost importance for PWS individuals and families. Despite advances in understanding genetic causes of PWS and generation of mouse models that mimic some clinical components, the underlying basis of PWS is unknown and **mouse models do not display overeating and obesity.** Although human studies have identified one candidate gene family (group of genes of similar function and structure) as likely playing the most important role in PWS, the lack of a suitable animal model and access to key tissues has meant little is known about what this gene family does. We have discovered this same gene family (and other PWS genes) in the pig and plan in the proposed research to: 1) characterize the unusual number and structure of this gene's family members, 2) determine exactly what this gene family does and how it does it, and 3) demonstrate that loss of this gene family in pig leads to the major clinical components of PWS. ***This project will provide an understanding of the major role of this gene family and how its loss causes PWS.***

Overcoming the intense drive to overeat and subsequent obesity that provides the greatest threat to life quality and expectancy is of upmost importance for PWS individuals and families.

The specific aims of the PWSA grant are to characterize the complex Snord116 gene locus of the pig, to begin to identify target genes of the Snord116 small RNAs, and to generate a targeting construct and to use gene targeting techniques to generate a deletion of the Snord116 locus in the pig cell lines. Researchers now know that the Snord116 family likely has a major role in PWS.

This work will also provide a suitable animal model

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Grants, *continued from page 3*

which can be used for clinical evaluation and treatment studies and to obtain critical tissues such as the brain (for developmental and behavioral features of PWS), muscle (for reduced muscle tissue and weakness in PWS) and adipose tissue (for increased fat tissue in PWS). A clinically accurate animal model is crucial for future studies of how the genes causing PWS control how much we eat and how that energy is used or stored and results in obesity. Having a pig model of the severe overeating and obesity in PWS will allow future development of therapeutic approaches involving dietary, surgical, pharmacological, genetic, stem cell, and/or neural transplantation.

Due to the complexities of the genetic basis of PWS, Dr. Nicholls' FPWR grant awarded last year and the new PWSA grant have very different scientific specific aims and will develop very different reagents to produce in the future two different types of pig models related to PWS. The FPWR grant is developing the reagents for generation of a pig model lacking expression of all (more than ten) of the pig-PWS imprinted genes and will represent a model for the imprinting defect class of PWS individuals. In contrast, the new PWSA grant will develop the reagents for generation of a pig model with a deletion of and lacking gene expression of only the Snord116 imprinted locus, the top candidate gene for a major role in the PWS clinical features. The new PWSA grant also seeks to begin to determine functions of the Snord116 locus, which are presently unknown, and also to assess whether the Snord116 locus differs in size between different pig strains, and thereby will also generate a very different knowledge set than will the FPWR research grant. The ultimate goal of these two projects is to develop much better animal models for PWS. The scientific approach of the two projects is different, but they share the goal of increased understanding of the genetic and clinical basis of PWS, and in the long-term development of therapeutic approaches for PWS.

Metabolic Risks Associated With Antipsychotic Medication Use In Patients With Prader-Willi Syndrome

By Gregory Cherpes, M.D. – Principle investigator
Jennifer Padden Elliott, Pharm.D. – Co-Investigator
The Children's Institute, Pittsburgh

This is a retrospective medical chart review at Children's Institute of Pittsburgh to identify the adverse effects associated with the use of atypical antipsychotic medication in about 500 people with PWS observed over a 10 year period. They will collect and analyze charted data from over 500 charts on weight gain/loss, diabetes, hypertension, hyperlipidemia, and liver function in adult and pediatric patients treated with atypical antipsychotics and an equal number of age, sex, IQ, and genetic type-matched controls not treated with atypical antipsychotics. They will also evaluate indications for treatment with all types of psychoactive medications.

Due to the frequency of atypical antipsychotic use in the PWS population, the unknown effects of these agents on weight gain and metabolic risk within the PWS population, and the fact that complications of obesity remain the leading cause of death in this disorder, additional information is sorely needed. Their study will address this paucity of data.

Note: One additional grant may be awarded after revisions. ■

Social Responsiveness and Competence in Prader-Willi Syndrome: Direct Comparison to Autism Spectrum Disorder

By Anastasia Dimitropoulos, Alan Ho, Benjamin Feldman – J Autism Dev Disord DOI 10.1007/s10803-012-1547-3
A PWSA (USA) Sponsored grant

Prader-Willi syndrome (PWS), a neurodevelopmental disorder primarily characterized by an insatiable appetite and food preoccupations, is caused by the absence of expression of the paternally active genes of chromosome 15. Although the problematic behaviors and the cognitive profile of PWS have been thoroughly researched, the social functioning of PWS has only begun to be examined. Findings to date indicate that individuals with PWS have social impairments that may reflect specific difficulty interpreting and using social information effectively. In addition, evidence suggests that, of the two most common PWS genetic subtypes, there is an increased risk of social deficits in people with the maternally-derived uniparental disomy (mUPD) subtype in comparison to those with the paternal deletion (DEL) subtype. Furthermore, there is evidence indicating a genetic risk for autism spectrum disorder (ASD) is associated with the PWS chromosome 15 region, particularly in the mUPD subtype. Using parent/caregiver surveys to measure the ability to engage in and initiate social activity, our goal was to compare social functioning in individuals with PWS to

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Autism Spectrum, *continued from page 4*

those with ASD. Participants with mUPD scored similarly to the ASD group across most areas of social functioning. All groups had difficulty with social competence, although the DEL group scored higher than the other groups. These findings suggest that further understanding of social behavior in PWS is necessary to better understand the contributions of the PWS chromosome 15 region to ASD susceptibility, particularly with respect to the contribution of maternally inherited genes that define the mUPD subtype, as well as to improve in awareness and development/implementation of interventions. ■

Differences Between the Genetic Subtypes of Prader-Willi Syndrome

By Suzanne Cassidy, M.D.
President IPSWO Advisory Board

Why do different genetic changes cause PWS?

PWS is always due to the same thing: deficiency in the expression of certain genes on chromosome 15. Specifically, the deficient expression is from genes that are normally expressed from the member of the chromosome 15 pair that was inherited from the father, because the copy of these genes on the chromosome 15 inherited from the mother is normally not expressed ("imprinted"). This lack of expression on the maternally-inherited copy is the result of a chemical reaction called DNA methylation. So, in the general population, the genes relevant to PWS are only expressed (like reading a blueprint) from the chromosome 15 inherited from the father, and in PWS, that copy is missing (when there is deletion or uniparental disomy); or else the chromosome 15 inherited from the father is erroneously switched off ("imprinted") like the genes on the chromosome 15 inherited from the mother (an imprinting defect).

A deletion is a missing piece of a chromosome. In PWS, the missing piece is at the top of the long arm (called q) of chromosome 15, in a region called 15q11.2-q13. There are three common points along the chromosome in which the break occurs, two at one end with four genes between them and one at the other end. The type of deletion missing the larger piece of genetic material is sometimes called a Type 1 deletion, and the smaller deletion is sometimes called Type 2. A few deletions are larger or smaller than the common deletions, but most genetic testing for PWS does not identify those differences.

Maternal uniparental disomy (also called UPD) is a situation in which both of the members of the chromosome 15 were inherited from the mother (and the genes relevant to PWS are therefore not expressed from either copy). The chromosome 15 that should have been inherited from the father—the one with the expressed genes for PWS—is missing. This situation is somewhat more common among children with PWS born to women over age 35 years.

An imprinting defect is a condition in which there is one chromosome 15 from the mother and one from the father, but the one from the father behaves as though it were inherited from a mother, at least as far as the genes for PWS are concerned. Those genes relevant to PWS are switched off in both the chromosome 15 inherited from the mother (as is normal) and the one inherited from the father (which is not usual). Most cases of imprinting defect are random and of unknown cause, but a small percent are due to a small deletion in the i1mprinting center which is responsible for switching genes nearby off and on (applying or erasing DNA methylation). In some families that have a child with PWS due to an imprinting center deletion , there is a significant risk to have another affected child. Therefore, in families planning future children, further genetic testing and genetic counseling are important.

A few people who have PWS are found to have a "translocation", which is a rearrangement of two chromosomes such that part of one is stuck onto part of another. Most of the time that this happens in someone with PWS, the translocation involves one chromosome 15 and there is a deletion on the rearranged chromosome. In that case, the individual in effect is like people with PWS due to a deletion. Occasionally, a translocation can cause uniparental disomy 15, and the individual in effect is like people with PWS due to UPD. Sometimes with a translocation, there is an effect from another missing piece of chromosome in addition to it causing PWS.

What are the differences in the effects of the different genetic causes of PWS?

Researchers, by studying many people with each genetic type of PWS (primarily the most common deletion and uniparental disomy types) have identified a few differences between the groups. Most of these differences are in the frequency or severity of findings in the two groups. People with an imprinting defect appear to be most similar to those with UPD.

Before mentioning these differences, it is very important to explain that these are GROUP differences, not individual differences. *There is no feature that is exclusively found in one of the three genetic categories.* And it is also important to recognize

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Genetic Subtypes, *continued from page 6*

that even within a genetic type there is a lot of variation among people affected with PWS, so there is a lot of overlap in the genetic groups. For example, even though the people with UPD as a group have a slightly higher mean intelligence quotient (IQ) than people with deletion as a group (8 points, with an IQ of 100 being the average in the general population), there is a very wide range of IQ within each genetic group (about 40-90). So in an individual, knowing the genetic type is not very helpful in knowing the future abilities or problems of the individual.

In general, and as a group, people with a deletion are more likely to have fair coloring (hair, eyes, skin), whereas those with UPD more closely resemble their parents. People with a deletion are more likely than those with UPD to have the characteristic facial appearance of PWS. And they are more likely to be skilled with jigsaw puzzles.

In general, and as a group, people with UPD are more likely to be born late. As noted above, they have a slightly higher average IQ than the group with deletion. In addition, they have somewhat milder behavior problems (temper tantrums, stubbornness, repetition, controlling behavior). However, it appears that people with UPD as a group have a significantly higher incidence of autistic characteristics and of psychiatric disorders, including psychosis, than those with deletion.

In addition to these differences, there are some studies suggesting somewhat worse behavioral problems in those with a Type 1 (slightly larger) deletion versus those with a Type 2 deletion. There are also some studies that have not shown these differences.

It is important once again to stress that knowing the genetic type does not predict the manifestations of PWS in an individual. It is primarily important for genetic counseling purposes, as far as we know today.

Why are there clinical differences between the different genetic types?

The genetic or biological basis for these differences has not been determined. In people with a deletion, a number of genes are missing, and some of those genes are normally expressed from both members of a chromosome pair. In people with UPD or an imprinting defect, the genes are not actually missing, but are just not being expressed (turned into messenger molecules, regulators or proteins). This presence of some genes that, as far as we know, are not the ones causing PWS but are also in the chromosome segment included in the deletion, may have an impact on the clinical manifestations in an individual. There is still much to learn about the genetics of PWS! *[Dr. Cassidy is currently President of the International Prader-Willi Syndrome Organisation (IPWSO). She also has served for many years on the PWSA (USA) Scientific Advisory Board.)* ■

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.

October 17-20, 2012
2nd International Conference on Hyperphagia
Pennington Biomedical Research Center, Louisiana State University, Baton Rouge, Louisiana, USA

HYPERPHAGIA
The Mechanism The Research The Treatment

PWSA (USA) Scientific Day, Providers Conference, and State Leaders Day

The 2nd International Conference on Hyperphagia
Wednesday, Oct. 17, 5:30 p.m. until
Friday, Oct. 19, 1:30 p.m.

The PWSA (USA) Professional Providers Conference
Thursday, Oct. 18, 8:30 a.m. until
Friday, Oct. 19, 12:00 p.m.
(see page 10)

The 26th Annual PWSA (USA) Scientific Day Conference
Friday, Oct. 19, 12:00 p.m. until
Saturday, Oct. 20, 12:00 p.m.

The State Leaders Day Conference
Saturday, Oct. 20, 8:00 a.m. – 5:00 p.m.

Register now at www.hyperphagia.org

Pennington Biomedical Research Center
Louisiana State University – Baton Rouge, Louisiana

Salivary Flow and Oral Abnormalities in Prader-Willi Syndrome

International Prader-Willi Syndrome Organisation 7th Scientific Conference - May 20-21, 2010, Taipei, Taiwan

By Ronnaug Saeves[1], Hilde Nordgarden[1], Ivar Espelid[2], Kari Storhaug[1]; [1]TAKO-centre, Lovisenberg Diakonale Hospital, Oslo, Norway; [2]Department of Pediatric Dentistry, University of Oslo, Norway.

INTRODUCTION: Persons with Prader-Willi syndrome (PWS) have sparse, thick and sticky saliva. High caries activity, poor oral hygiene and extreme tooth wear have been described in case reports. Oral and dental problems have received little attention by researchers. The aims of the study were to examine salivary flow rate and describe oral and dental characteristics in Prader-Willi syndrome.

METHODS: Fifty-one individuals with PWS, aged 5-41 years, and an age- and sex-matched control group were examined with regard to salivary flow rates, dental caries experience, gingival inflammation, enamel defects and tooth wear. Both unstimulated and chewing stimulated whole saliva as well as taste-stimulated parotid salivary flow rates were measured. The presence or history of dental caries was evaluated both clinically and on radiographs. Tooth wear was evaluated according to a 4-point scale, the Jonkoping-index. An individual tooth wear index (I_A) was created on the bases of the scores of incisal or occlusal wear for each tooth.

RESULTS: The average flow rate for unstimulated saliva (UWS) was 0.12±0.10 ml min^{-1} for individuals with PWS compared with 0.32 ± 0.20 ml min^{-1} for controls (p<0.0001). Chewing stimulated flow rate (SWS) was 0.41±0.35 ml min^{-1} for the PWS group compared with 1.06±0.65 ml min^{-1} for the control group (p<0.0001). Taste-stimulated parotid saliva was not found to differ significantly between the persons with PWS and healthy controls. There was no significant difference in caries experience in the primary dentition. Caries experience in permanent teeth (persons >18 years) was higher in the control group (p=0.04). The median GI-index (gingival inflammation) was significantly higher in the PWS group compared with the control group (p=0.04). The number of surfaces affected with enamel defects was 3.5(1.0-8.8) in the study group and 4.0(0.5-7.0) in the control group (p=0.76). The median tooth wear index I_A was 7.5 (0-100) in the PWS-group and 2.2 (0-10.7) in the control group (p<0.0001)

CONCLUSIONS: Low whole salivary flow and tooth wear are very common in individuals with PWS. Taste-stimulation may increase salivary flow rates in this group. The oral hygiene in the studied population with PWS was generally poor but the dental caries experience was not increased. This may reflect a low sugar diet and tight follow-up regimes. ■

Severe Tooth Wear in Prader-Willi Syndrome: A Case-control Study.

By Ronnaug Saeves, Ivar Espelid, Kari Storhaug, Leiv Sandvik and Hilde Nordgarden

Abstract (provisional)

BACKGROUND: Prader-Willi syndrome (PWS) is a rare complex multsystemic genetic disorder characterized by severe neonatal hypotonia, endocrine disturbances, hyperphagia and obesity, mild mental retardation, learning disabilities, facial dysmorphology and oral abnormalities. The purpose of the present study was to explore the prevalence of tooth wear and possible risk factors in individuals with Prader-Willi syndrome.

METHODS: Forty-nine individuals (6-40 years) with PWS and an age- and sex-matched control group were included. Tooth wear was evaluated from dental casts and intraoral photographs and rated by four examiners using the Visual Erosion Dental Examination (VEDE) scoring system and the individual tooth wear index IA. In accordance with the VEDE scoring system, tooth wear was also evaluated clinically. Whole saliva was collected.

RESULTS: Mean VEDE score was 1.70 +/- 1.44 in the PWS group and 0.46 +/- 0.36 in the control group (p<0.001). Median IA was 7.50 (2.60-30.70) in the PWS group and 2.60 (0.90-4.70) among controls (p<0.001). In the PWS group tooth wear correlated significantly with age (VEDE; r=0.79, p<0.001, IA; r=0.82, p<0.001) and saliva secretion (VEDE; r=0.46, p=0.001, IA; r=0.43, p=0.002). Tooth grinding was also associated with tooth wear in the PWS group, as indicated by the mean VEDE 2.67 +/- 1.62 in grinders and 1.14 +/- 0.97 in non-grinders (p=0.001) and median IA values 25.70 (5.48-68.55) in grinders and 5.70 (1.60-9.10) in non-grinders (p=0.003). Multivariate linear regression analysis was performed with tooth wear as the dependent variable and PWS (yes/no), age, tooth grinding and saliva secretion as independent variables. PWS (yes/no), age and tooth grinding retained a significant association with tooth wear, VEDE (p<0.001) and log IA (p<0.001).The only factor significantly associated with tooth wear in the control group was age.

CONCLUSIONS: Our study provides evidence that tooth wear, in terms of both erosion and attrition, is a severe problem in Prader-Willi syndrome. There is therefore considerable need for prosthodontic rehabilitation in young adults with PWS. ■

Credit: PubMed.gov - US National Library of Medicine National Institutes of Health - BMC Oral Health. Published 28 May 2012; 12(1):12

Weight Loss Drug Approved by FDA

By Janalee Heinemann, M.S.W. PWSA (USA) Director of Research & Medical Affairs

A new weight loss drug, **Qsymia**, has been approved by the FDA. Weight loss was more with this drug than with two others recently reviewed by the FDA with one approved (Lorcaserin/ Belviq) that was not nearly as remarkable regarding weight loss. Qsymia is a combination of two older drugs that have long been known to help with weight loss: phentermine (the safer half of the old fen-phen drug that was banned) and topirimate – a drug that was studied back in April 2000 under a PWSA (USA) grant, ***"Open-Label Pilot Study of Topirimate in Adults with Prader-Willi Syndrome."***

Topirimate is an anti-convulsant drug that makes people feel more satiated after eating, which is why we sponsored the study by Nathan A. Shapira, M.D., Ph.D. The study showed that Topirimate did not significantly change the calories consumed, Body Mass Index, or decreased self-reported appetite in PWS; there were no significant changes in compulsions. Surprisingly though, Topirimate treatment resulted in a clinically significant improvement in the self-injury (i.e., skin-picking) characteristic of PWS.

The researchers of Qsymia state that it targets multiple brain signals that drive people to overeat. We cannot say at this time if it will be effective with PWS. Please know that we will keep you informed of any new outcomes on this and other obesity drugs that might be helpful with PWS. As with any new medication, if you wish to try it before it has been proven to be effective in PWS, it is most useful to do so as part of a clinical trial.

Currently, I have been working informally and under strict agreements of confidentiality with two pharmaceutical companies which are working on potential products that might impact on PWS. We are also working with FPWR on financially supporting the Best Idea Grants post our 2nd International Hyperphagia Conference (go to www.hyperphagia.org) with 2012 One Small Step funds. We never forget that the #1 deadly enemy of PWS is the appetite. ■

CHUCKLE CORNER

I was reading to my oldest son (5) tonight and one of his pages in "I'm a Good Friend!" says "Friends share their snacks." Well, in our family, we have talked about the dangers of doing so (my kids and I have food allergies, our niece/cousin has PWS, etc.). My son said, "We don't do that." And I assured him that was OK, reminding him of allergies and PWS. He said, "When Josi (age 10 with PWS) does not eat, we don't eat." Then he paused and said, "Mommy, when I grow up, I want to be a scientist so I can figure out Silly Wyndrome." Then in the next breath he said, "And I'll also teach the president how to fight." This perfectly characterizes my son of extremes—loving and, um, 'lively.'

~ Jodi O'Sullivan, Cincinnati, OH

Fundraising News

paint the yellow

Another Way to Fund Raise!

It started simply enough.

Parent Ed Peoples from Pennsylvania enjoys simulated auto-racing and joined the league Paint the Yellow. During races, he would chat with others racing. Over time, Ed developed friendships with other drivers, even though they've never met. Sometimes, the other drivers would hear Ed's daughter Victoria, 12 with PWS, cheering on her dad in the background. They questioned Ed about her here and there, and little by little they learned about Prader-Willi syndrome.

Then in June members of the Paint the Yellow league approached Ed with an idea. They wanted to do a charity race to raise money to help Ed with his bills. Ed was deeply touched but asked for a change. He wanted the funds to go to PWSA because the organization has been helpful to his family. He wanted to give back, and he wanted to do it in a way that would help all who have PWS.

Within days, the July 5th race was scheduled with a $1,900 purse generously donated by a league member. Anyone who is a member of iRacing.com would be eligible to enter and could attempt to qualify for a minimum suggested donation of $5 per qualification attempt. A full month of activities would lead up to the race, including nearly 50 qualifying sessions to lock in the fields for the Twin 50 races that would set the stage for the main event.

The Prader-Willi Syndrome Charity Race, as it was ultimately named, was shown live on ETV at http://www.etv-eplay.net. When approached about the benefit race, ETV also suggested a pre-race interview and on-air announcements about PWS during the race, allowing for more PWS awareness to potentially hundreds of people.

To top it off, Ed learned that the friends he never met desire to make this an annual event. They've gone flat-out, burning rubber from the start to make this first annual event very special. And very special it was, raising $3,023.51.

To learn more, please visit: http://charity.painttheyellow.com/ http://www.iracing.com/ or http://www.etv-eplay.net ■

Spotlight on Publications

By Julie L. Doherty, Committee Co-Chair

Booklets, brochures, DVDs, *The Gathered View*. If you have been to a state or national conference, or have received a Packet of Hope, you have seen the work of the PWSA (USA) Publications Committee. PWSA (USA) currently has nearly 30 publications in stock – booklets, brochures and DVDs covering topics such as Weight and Behavior Management, Nutrition for each age group, and material for Physicians and Educators. Have you ever wondered how these fabulous resources were developed and continue to be updated?

Well, the answer is the all-volunteer and very hard-working Publications Committee. The committee members review publications regularly, solicit the services of more volunteers such as physicians, speech and physical therapists and other professionals to update and in some cases completely re-write the information, and coordinate with graphics artists and printers to keep our shelves at the National Office well stocked. They also review new publications, searching for topics that would be of benefit to our members. A complete listing of our products can be found by clicking the Product tab on our website.

Janalee Heinemann, Director of Research and Medical Affairs, recently heard from the sibling of a 54-year-old adult with PWS and noted how remarkable it was that the family had not contacted PWSA (USA) for services before now. The sister stated that with all of the information we put out and *The Gathered View,* they have been able to get along just fine until now. It was a nice reminder of how valuable our information is. It is the team effort that makes it possible for us to help thousands of families and professionals every year.

Jackie Mallow and I co-chair the Publications Committee. I joined the "PWS Family" in 1997 when my niece, Leslie Torbert, now 15, was diagnosed. Michelle Torbert, my sister, and I began attending conferences in 1998 partly because Michelle's husband didn't like to fly. It was a nice opportunity for Sister Weekends! We began to volunteer with PWSA (USA) by helping Annie and Norma sell publications and logo items in between conference sessions. In 2005 I learned there was a vacancy for the position of secretary of the Board of Directors, and volunteered. I have served in that position ever since. It was my thought that if I had a child with the syndrome, I would not have as much time to volunteer. So I wanted to do what I could to help. Besides, I have gained more than I have given in service to PWSA (USA).

Our life experiences often determine our passions. For those of us involved with PWS, that is truly the case. We all have a talent that can be used in the work that is done by PWSA (USA). Think about your talent and step forward to volunteer. You'll be glad you did!

[Dottie Cooper notes: As chair of the Publications committee, Julie Doherty spends countless hours coordinating all the volunteer efforts for content, editing, proofreading, and final approval. In addition, Julie serves as secretary to the National Board of Directors, moderator for the Chapter Leaders Yahoo Support group, and is an active member of the new State Leaders Team. Thank you, Julie, for all you do.] ■

Our life experiences often determine our passions. For those of us involved with PWS, that is truly the case. We all have a talent that can be used in the work that is done by PWSA (USA).

Sandpaper, *continued from page 1*

I put my arms around him and hold him. I pull him onto my lap. I softly whisper to him "It's ok, buddy, if you're not ready, it's ok. We don't have to do this today."

All of this happened in the first two hours of his day. This is Reagan's world, typical of what his days are like. Every day. The world exists around him and moves faster than he can keep up with. I want for him to be more typical and less "not" typical. Yet I know that every time I try to push too hard, he resists even more. I have to remind myself that for him the world around him rubs him like sandpaper on his skin. It hurts, it is uncomfortable…it scratches and leaves a mark. ■

Stuart Mitchell carries the torch.

PWS Carries the Olympic Torch

PWSA UK (United Kingdom) reports that not one but four people with PWS plus another who is the grandfather of a person with PWS were chosen to carry the Olympic Torch in their areas. You can read their inspirational stories on their Web site by going to the Home Page at www.pwsa.co.uk and clicking the "Readmore" button on the featured sliding photo banner. ■

Raising an Army of Advocates

By Evan Farrar, Crisis Counselor

In July, it was my great privilege to represent PWSA (USA) and the PWS community at the Institute of Special Education Advocacy (ISEA) in Williamsburg, Virginia**.** The five-day training program, hosted by the William & Mary School of Law and co-sponsored by the PELE Special Education Advocacy Clinic, Wrights Law, and The Oklahoma Disability Law Center, included 25 sessions on applicable laws, ethics, best practices in advocacy, strategies in working with parents and schools, and dispute resolution procedures, taught by national leaders in the field.

Limited to 40 participants, attendees were accepted after completing a rigorous written application process. I was very proud to be one of them and discovered I was the only advocate representing an agency from the rare disease community, which is a tribute to PWSA (USA)'s long standing commitment to being on the cutting edge of providing special educational advocacy and support to the families we serve. The experience was a once in a life time opportunity to formally and informally interact with leaders and other professional colleagues in the special education advocacy field. I learned many things including:

- Special education advocacy is not a regulated or certified profession in any state, which means parents need to be very careful when hiring an advocate to make sure the person is well qualified for the job.
- Every parent should take time to read the Individual with Disabilities Education Act (IDEA) to understand their child's legal educational rights.
- The parent/school relationship is the key to a child's success in school, and so developing a collaborative, professional, and effective working relationship is essential.
- Parents should never assume taking legal action against a school will be successful or is the best option.
- Two concepts are critical for parents to remember in working with schools: **document everything** and **data collection**. Parents should document all interactions related to their child's school experience. For example, always ask for policies and decisions in writing, and put parent concerns and requests in writing to the school. And keep all of this documentation in a well-organized file. At the same time parents should make sure the school is measuring their child's progress frequently and effectively. This data collection combined with strong documentation improves educational outcomes.

One lesson of the institute was very clear. PWSA (USA) must move even more creatively in the direction of equipping parents to be the best advocates they can be for their child with PWS. During the ISEA graduation ceremony Pete Wright, co-founder of Wrights Law, said the goal of the ISEA was to raise an army of trained special education advocates across the country. My hope is PWSA (USA) will join this effort by raising our own army of PWS parent advocates who are equipped with the knowledge and strategies they need to support the children they love. So that someday soon a free and appropriate public education, still only a promise for too many, will at last be a reality for students with PWS in every school across the United States. ■

The Army in Virginia - Can you see Evan?

Georgia State House Designates Kindness Week in November

In February, the Georgia House of Representatives adopted a House Resolution sponsored by state Representative Andy Welch marking the second week of November as Kindness Week. The campaign, begun in 2004, was initiated in honor and memory of Hunter Welch, the son of Kit and Buddy Welch, and brother of State Representative Welch. Hunter was diagnosed with Prader-Willi syndrome. He died in 2004 at age 12.

Advocates of the campaign observe the anniversary of Hunter's birthday, November 5, in conjunction with international observance of "Random Acts of Kindness Week."

Hunter's mom Kit said that he showed kindness to those he met. The family had received much caring and support during difficult days, and started the week because they wanted to give back. Many, including businesses, friends, schools, and church groups have joined the campaign to spread awareness of the benefits of kind actions. ■

A Golden Granddaughter from the Gods

By Michael Tate

She arrived as a Christmas gift in December 2003. My sixth grandchild. Blond and delicately beautiful with the requisite blue eyes. All was well. Then – the SHOCK. There was a possible problem. Questions were asked. Does anyone else, except her mother, in the family have blond hair? Yes, I did as a child as did my brother. A week later it was confirmed. Ashley was a child with Prader-Willi syndrome!!

"Prader-Willi" – What was that? No one knew. Never heard of it! The Internet description gave detailed and alarming coverage of every detail of what might be Ashley's future. Our daughter Suzanne, Ashley's mother, said later that Ashley's birth date was the day that changed her life forever. She also said that God had chosen her to be the guardian of this little girl. That being so, it followed that He had chosen me to be her Grandfather.

In those early days I watched as Suzanne coped with an infant that would not feed normally and to whom she had to give nightly Hormone Growth injections. I was impressed with the ways she sought information and direction from the many medical facilities, while at the same time I was horrified at the lack of awareness of the syndrome, both in the general public and in the day-to-day medical profession. As Ashley grew, I found myself constantly looking for signs that would confirm the descriptions of behavior as outlined very explicitly online. Thankfully, outwardly, there was very little to distinguish Ashley from any normal little child, which confirmed my intent to treat her as such whatever may happen. Yes, Ashley had floppy limbs, speech problems, but Suzanne worked continuously to get the correct help. Early on, a speech therapist worked wonders.

Ashley now has a sister Meghan, a fish and half shares of a kitten, Belle. She is very active, swims and does Yoga. She played softball and tried soccer. I see that she does tire easily and is ready for bed early, perhaps because she wakes full of energy early each day. She will ask me when I am having dinner or whichever meal is next. I do not make a big fuss about her frequent requests for food as she is aware that her intake must be limited. So far there has been no serious need to lock the fridge. I am delighted that she loves her school where her teachers and school leaders have been briefed on her condition and her needs. She does well scholastically, and her reading is excellent. She is inquisitive and asks many questions. She is the right height for her age with weight only slightly high.

Two possible concerns. She does have "meltdowns" both at home and in public which Suzanne deals with expertly, making it clear that it is not appropriate behavior. Second, she is extremely friendly and will readily ask questions to complete strangers. OK now but will it be later?

Suzanne asked me to write something about being a grandfather to a child with PWS. I don't look at it that way. I see a bright, pretty, healthy little blond girl who has her own personality! Just as important I see a mother who has schooled herself to get her daughter the very best of attention and medical opinion, who despite the stress is always positive and cheerful and treats Ashley just like her younger sister.

Ashley looks after me, too! If she's around, she will bring me my pills saying "Here, you must not forget to take your medicine." Recently we travelled to New York to visit with one of the top specialists in PWS – about 85 miles each way. She visits him every 4 months. For 30 minutes all three of us played "I Spy" in the car; then Ashley fell asleep. The doctor put her through various tests, weighed her and recorded her height. Ashley answered questions without hesitation. At the close he proclaimed how delighted he was with her progress and congratulated Suzanne on attention to all Ashley's requirements. It was good to get official confirmation.

[We are delighted to receive an article from a grandfather and encourage other grandparents to consider sending a piece about their grandchild with PWS.] ■

Excerpts from a recent letter

to Janalee from Dawn Taylor in West Australia

Ryley with a friend.

One year after Ryley was born in 1984 I became a founder member of the Australian Association. Those were the days of snail mail and printing out monthly or bi-monthly newsletters. PWSA was a life-line as you seemed to have so much more support from professionals within the medical fields and also with group homes. I remember your personal support was vital to my ongoing participation in the association.

After devoting myself for four years to the association and putting my two other children at the bottom of my "to do" list, I realized that life just ticks along and all my children equally needed my care, attention and support. I decided then to focus on my family and balance our lives.

So Ryley became one of the gang and we tried to play happy family, but, of course, over time cracks appeared. PW needs just ARE greater, and Ryley's tantrums and outbursts became destructive. By 13 we were all just shattered, and our family's cracks became canyons. The very hard decision to put Ryley in care was made. We were all in a dark place at that time, and each family member tried hard to heal deep wounds from the separation of Ryley from our family unit and felt guilty for feeling some sort of release and relief from the day-to-day hassles. I'm not sure that Anouska and Tristan, now 32 and 29, have ever recovered from those feelings of guilt and deep sadness.

Ryley has round-the-clock caregivers and lives with three other men in a suburban home. He has a social life that rivals any young person. He is out dancing, acting, bowling and is involved in other artistic and social pursuits. We joke that we find Ryley now the easiest of our children to deal with. We know who he is with, what drugs he is taking, where he is and who is driving!!!

Living with PWS means that you have challenges that are really hard but you get to delve down deep within yourselves to overcome these challenges. Life is magnified, and the good is always VERY good and the bad is VERY bad. In the end the joys far outweigh the lows, and you know you can survive anything.

Looking back, I think we made mistakes, but like all parents we did the best we could. Drugs played a big part in Ryley's development, and we failed as parents by refusing to give up Ryley's unique personality to drugs. His caregivers could not cope and sought the referrals of specialists who all prescribed mood-altering drugs and over the years he has been on a number of medications and finally has a balance in his life. The anti-psychotic drugs he is taking now enhance his lifestyle and give him balance. He is happy, really happy, and that makes us happy.

I have always dreamt of living in France, and we now spend five months in France and seven in Australia. We live a simple life and we enjoy the simple things that other families may take for granted. We value time together as a family, and we listen to our children. I really think we owe this fabulous lifestyle to Ryley. Living with PWS means that you have challenges that are really hard but you get to delve down deep within yourselves to overcome these challenges. Life is magnified, and the good is always VERY good and the bad is VERY bad. In the end the joys far outweigh the lows, and you know you can survive anything.

So Janalee, thank YOU for being part of our family's formative years and for providing me with the courage, the information and the knowledge that allowed us to survive and prosper. ■

Sibling View

To The Prader-Willi Association Board of Directors,
My name is Libby Elaine DeLemos, I'm 17 and the younger sibling of a person with PWS. My sister Abby is 21 and lives at the ARC in Gainesville; FL.

Libby, 17, with sister Abby, 21.

Abby has been both a blessing and a burden in my life. For the longest time I did not understand why it was my family who had to have a family member who was disabled. As I grew up, everything became clearer and while Abby has no idea, she has made me the person I am today. I want to share a little bit of that with you.

As a child, I did not get the attention I wanted or needed because most of it was given to Abby and her problems that needed more time and parental involvement. When you are a kid all you want is someone to listen to your stories and tell you that they are proud of you. I felt I did not get that, and although it did have an effect on how my childhood will be remembered, it definitely made me more grateful for the love and care I get now. Do not get me wrong, I would not go back and change anything. Abby helped me realize what was important in life and showed me that life was more than getting everything I wanted. An example would be simply wanting ice cream before bed but I could not have any because my mom wanted to avoid problems.

Over the years, I have become more proud of being a sibling of a child with PWS. I am not embarrassed to tell people who Abby is and what her disability is. Abby has made me be 100% more thankful for the things I have and this is why I am writing to you.

No one understands better than other siblings or parents. When I write letters to colleges, I tell them about Abby, but they do not understand her disability and the strain it places on families.

Honestly, they never will. A vast majority of our financial resources have gone to Abby and her care. My number one goal is to spread the word about kids with PWS and raise awareness and money to find a cure.

I want to make a suggestion regarding a scholarship fund that could be made available for siblings. There are so many of us out there that need help with college. When we write about being a sibling of a person with PWS, they might feel bad, but they do not understand exactly what it means and how PWS has shaped the women and men we have grown up to be.

Maybe this is something PWSA could do in the future. We are all 'still hungry for a cure' and only God knows when the hunger will be filled. I want to thank the readers of this letter for your time and consideration. Writing this means a lot to me, and I hope in some way it touches each of you as well.

I love someone with Prader-Willi Syndrome,
~ Libby Elaine DeLemos, Florida

[John Heybach, Chairman of the Board of Directors, notes that a scholarship fund for siblings is not within the parameters of our mission.] ■

Chapter View

Across the Country, our Chapters are Busy!

Washington, Oregon and **Idaho** joined for a PWS Picnic and Family Campout in early August… the Annual PWSA of **Maryland, D.C. & Virginia** Chapter Picnic is slated for early September…PWSA of **Ohio** Fall Weekend camp will be Friday evening, October 19, to Sunday afternoon, October 21. Children and adults, ages 8 and above, who have PWS are eligible to attend the weekend.... Prader-Willi **California** Foundation is proud to announce a new educational series for parents, extended family members, friends, care providers, teachers, and ***all*** professionals who serve someone with PWS. The Prader-Willi Syndrome Behavior Training Series will be held from 1:00 - 4:00 p.m. on August 5, September 1, and October 6 in Redondo Beach, California.…Debbie Lange, Executive Director of the **Georgia** Association, has joined the **State Leaders Team**….**Pennsylvania** held its annual golf outing in early June and is sponsoring a Family Day on September 30 to attend a Pirates baseball game and cheer on the team. Clint Hurdle, our national spokesperson, is manager of the team… **Wisconsin** is sponsoring a daylong conference on "Planning for the Future—Strategies to Support our Children's Lifelong Needs" on November 3 at Appleton, Wisconsin…**Indiana** plans a one-day Midwest Conference October 6, 9 am-4 pm, at the Riley Hospital for Children in Indianapolis. ■

Contributions

Thank you for Contributions in June and July 2012. 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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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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E-mail Support Groups:
We sponsor nine groups to share information.
Go to: www.pwsausa.org/egroups

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Medical information published in *The Gathered View* is not a substitute for individual care by a licensed medical professional.

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Sign up today at www.pwsausa.org!

Insurance Tips

Handling insurance is challenging for anyone, but it is especially complex for families with children with special healthcare needs. Here are some basic tips for better understanding and interacting with your child's insurance.

- Identify the effective dates of your child's coverage and how long your child is covered on your plan.
- Know what your insurance covers by keeping a copy of your plan documents.
- Find out how to review claims with your insurance provider—do they mail statements or can you check claims online?
- When you speak with someone on the phone, write down the name of every person you speak with, the phone number (if applicable), and the time and date of every contact.
- Keep all insurance documents, including explanation of benefit (EOB) statements, prior approval letters, letters you write, and receipts.

The Clint and Karla Hurdle Endowment Fund for Prader-Willi Syndrome Support

"To act generously is to awaken a certain kind of freedom: freedom from the stranglehold of self-concern, and, consequently, freedom to choose a level of responsibility beyond the minimal charge most of us have for ourselves. To give unselfishly is at least momentarily to be free of ourselves, free of greed and attachments, resentments and hatreds, habitual and isolating acts of self-protection."
- Dale Wright

Clint Hurdle, PWSA (USA)'s national spokesperson, manager of the Pittsburgh Pirates (Go, Bucs!), and founder of the Endowment Fund, has a wonderful habit of sending out frequent emails with inspirational and encouraging quotes. You'll be finding some of them, like the one above, sprinkled in issues of *The Gathered View.*

To donate, mail your donation to PWSA (USA), and in the memo field or in letter form, note that it is for the endowment.