

Prader-Willi California Foundation's 2009 General Education Meeting
Broadening Our Understanding of Anxiety, Endocrine & Gastric Issues in Prader-Willi Syndrome

PWCF's annual General Education Meeting was held on November 7 at the Long Beach Marriott Hotel. Families and professionals in attendance received important information on significant issues that affect persons of all ages with Prader-Willi syndrome: stomach and bowel issues, endocrine issues, and anxiety.



When 'Enough!' Isn't Enough: Moving Beyond Limit Setting to Help People With PWS Cope More Effectively With Frustration and Anxiety was presented by **Dawn Huebner, Ph.D.**, a psychologist expert in anxiety related disorders. Dr. Huebner explained that behavior is shaped by thoughts, feelings, and experiences: what we think influences how we feel and what we feel influences our behavior. Therefore, we can help kids and adults with PWS better manage their anxiety and frustration by teaching them tools to identify their thoughts, then alter them, and ultimately better manage their own behavior.

Dr. Huebner talked about the importance of clear, consistent, posted, and enforced routines to keep worries to a minimum. Routines should be created around eating, sleeping, homework, work, chores, TV and other electronic games, etc., and incorporated into the process of transitioning from one activity to the next. Discuss changes before they happen and create a visual picture of what is expected to occur. Dr. Huebner recommended using Collaborative Problem Solving techniques, an approach developed by Ross Greene, Ph.D. [for more information, visit the Center for Collaborative Problem Solving at www.CCPS.info] and explained that our use of words can create resistance or enhance cooperation. For example, in the sentence "You want to ride in the front seat of the car but I want you to ride in the back seat," the word "but" negates what our child/adult child wants, while the word "and" takes into account his desires. "You want to ride in the front seat *and* I want you to ride in the back" paves the way to using Collaborative Problem Solving techniques to resolve the disagreement without argument.

Dr. Huebner outlined seven teachable skills to manage a Worry – Externalize the Worry; Quantify the Worry; Distraction; Relaxation; Flexibility; Stop & Shift; and Containment – and provided a detailed overview of how to teach each of these skills (as outlined in her book, *What To Do When You Worry Too Much* available from PWCF). Embedded within her presentation was the importance of slowing down your rate of speech to help the person with PWS relax; listen without offering solutions; don't problem-solve until everyone is calm; don't try to teach a lesson while your child is still distressed; don't announce a negative consequence or make threats; don't worry that your child is getting away with something; don't worry about what other people are thinking. The concept of a "Zigger Zagger" can help teach flexibility. Containment strategies, such as utilizing a Worry Box, Worry Time, or Question Cards, do not work well for habits, which include skin picking, nail biting or picking, hair pulling, or sucking. Habits are partly learned and partly hard-wired and almost impossible to stop through willpower alone, but can be managed in other ways as outlined in *What to Do When Bad Habits Take Hold*. Each of Dr. Huebner's *What to Do Guides for Kids* can be successfully used with teens and adults and are available from PWCF (PWCF earns a percentage of each book sold). Additional helpful books include *What to Do When Your Temper Flares: A Kid's Guide to Overcoming Problems with Anger*; *What to Do When Your Brain Gets Stuck: A Kid's Guide to Overcoming OCD*; *What to Do When You Grumble Too Much: A Kid's Guide to Overcoming Negativity*; *What to Do When You Dread Your Bed: A Kid's Guide to Overcoming Problems with Sleep*.

A Comprehensive Overview of GI Issues in Prader-Willi Syndrome was presented by **Ann Scheimann, M.D., MBA**, a PWS expert and national consultant on PWS gastro (GI) issues. The prevalence of GI issues in persons with PWS of all ages is quite high. In addition to early feeding difficulties, reflux and aspiration symptoms are frequent problems in infancy. Problems with digestion affect approximately 35% of adults, constipation and diarrhea problems occur in 20-35%. Gastroesophageal reflux disease, GERD, is common. Dr. Scheimann outlined suggestions to treat GERD in infants: use thickened feedings, avoid overfeeding, use more prone positioning, and eliminate all exposure to tobacco smoke. In older children and adults she recommended avoid lying down after eating a meal, elevate the head of the bed, lose weight, avoid all tobacco, and avoid foods and medications that may cause reflux. Fundoplication is a surgical option when lifestyle changes aren't enough.



Oral problems are common including small mouths causing teeth crowding and enamel erosion. Salivary flow is generally far less than normal [dry mouth products, such as Biotene can be helpful]. Factors predisposing someone to choking, a serious and not uncommon occurrence in persons with PWS, include hyperphagia (high drive for food), thick saliva, weak pharyngeal muscles, and reflux. Dr. Scheimann advised all care providers to learn the Heimlich maneuver, treat reflux and gastritis symptoms, encourage chewing during meals, and of course supervise persons with PWS at all times. Risk factors for developing gallstones, also not uncommon, include obesity, low fiber/high fat diet, and diabetes mellitus. 70-80% of adults with PWS in one study had no biliary symptoms when their gallstones were detected. Dr. Scheimann also reviewed issues involving the gallbladder, bladder, and liver.

Constipation (passage of hard stools fewer than 3 times/week in adults) and encopresis (involuntary "fecal soiling") are common problems. Factors that add to constipating conditions include developmental factors (cognition, genetics, fluid intake, etc); metabolic and endocrine factors (hypothyroid, diabetes, etc.) and altered anatomy (low muscle tone, malrotation, etc.). Rectal ulcers can occur when there is chronic constipation. General guidelines to treat constipation in infancy include the careful use of glycerin suppositories or softening agents such as Karo syrup, and to increase fiber intake when solids are introduced. She cautioned against using enemas, suppositories, and finger dilations. For the school age child and adult, her suggestions included a fiber rich diet plus water and the continuous use of medications, such as Miralax.

Gastric motility (the rate at which the stomach empties) and an impaired vomit reflex (controlled by the central nervous system) contribute to serious stomach expansion and stomach rupture problems that can cause death. Warning signs and immediate hospitalization for potential gastric rupture or gastric necrosis include a binge eating episode followed by abdominal discomfort, recent history of gastritis or ulcer.

While a variety of bariatric surgery techniques have been attempted in persons with PWS, the long-term results have been very poor. Research continues to explore viable treatment options, but bariatric surgery is not currently one of them.



The Endocrine Perspective of Prader-Willi Syndrome from Infancy to Adulthood was presented by **Michael Gottschalk, M.D., Ph.D.** Dr.

Gottschalk reviewed the anatomy of the Hypothalamus and Pituitary Gland functions which play key roles in the body's production of important hormones including growth hormone and the sex hormones. The body of a person with PWS is deficient in its production of growth hormone. Dr. Gottschalk outlined the pros and cons of Growth Hormone treatment throughout the lifetime. On the Pro side: increased height, increase head size, decrease fat mass, increase lean body mass, increase strength, increase agility, increase ventilatory drive, stabilizes behavior. On the Con side: development or worsening of obstructive sleep apnea. He noted that Growth Hormone treatment does not cause scoliosis (side to side curve of the spine), which occurs frequently in persons with PWS.

The body of someone with PWS is also deficient in the production of sex hormones. At birth, most male infants with PWS have cryptorchidism, the absence of one or both testes from the scrotum. In most cases, there is a failure of the testis to move, or descend, during fetal development from an abdominal position, through the inguinal canal, into the scrotum. HCG injections are effective to descend the testes about 15% of the time, while surgery (which generally should be performed before age 12 months) is effective greater than 95% of the time. While pubertal signs may begin to develop in adolescence, sex hormone deficiency interferes with the body's ability to complete pubertal development. Women with PWS may begin menstruating in adolescence or adulthood but generally have irregular or non-existent menses. Sex hormone replacement therapy can be helpful. It is believed that both men and women with PWS are usually infertile, though there have been two pregnancies reported in two women with PWS.

Dr. Gottschalk outlined the risks factors associated with obesity: hyperlipidemia, the presence of raised levels of fat in the blood, hypertension, cardiovascular risk, sleep apnea, Blount disease (a growth disorder of the shin bone that causes the lower leg to angle inward, resembling a bowleg), and Diabetes Type 2.

Dr. Gottschalk outlined his reasons why he's not in total agreement that Central Adrenal Insufficiency is as common in persons with PWS as is currently speculated.

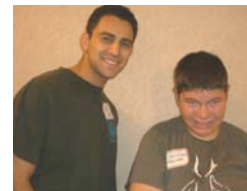
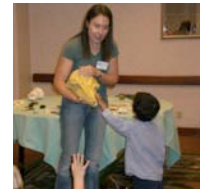
A Panel Discussion provided an opportunity for audience questions to be addressed in a concise, succinct manner for the benefit of all.

The Youth & Adult Program was written by **Jenn Paige Casteel** and Directed by **Kimberly Postal** (Youth Program), **Theresa McGrath** (Teen & Adult Program) and **Kristen Amarasekera** (Teen & Adult Program). PWCF is grateful to **Valerie** of the **Roundhouse Marine Studies Lab & Aquarium**, located in Manhattan Beach, who brought cool fossils of all kinds of sea life. Not only does the Youth &



Adult Program allow parents to attend the General Education Meeting, it provides an opportunity to make new friends and allows participants, both with PWS and without, to feel surrounded by soulmates.

Julie Casey reached out to local universities and colleges for the volunteers to staff the Program, and with help from **Renay Compere**, recruited students studying such fields as special education, speech & language pathology, occupational therapy, and physical therapy. These students now have a much broader and more personal understanding of Prader-Willi syndrome.



PWCF's Business Meeting was held during the lunch hour. **Ken Knox** presented his **2009 President's Award** to the following deserving awardees:

Jennifer Rinkenberger In recognition and appreciation of her work to create the Central California Prader-Willi Syndrome Support Group to provide information and support to families of a young child with Prader-Willi syndrome

Debbie Martinez In recognition and appreciation of her work to create the Central California Prader-Willi Syndrome Support Group to provide information and support to families of an older child or adult with Prader-Willi Syndrome

Jenn Paige Casteel In recognition and appreciation of her sustained dedication to families and her continuous work to improve the quality of fun for children and adults with Prader-Willi Syndrome

Nay Lay In recognition and appreciation of her nomination of Prader-Willi California Foundation for Southern California Edison's 2009 Philanthropy Budget Award

Carina Chaij In recognition and appreciation of her work to educate Spanish speaking families by translating educational materials

Kim Morgan In recognition and appreciation of her work on Prader-Willi California Foundation's Board of Directors to improve the quality of life for all persons with Prader-Willi syndrome

PWCF is grateful to each of the exhibitors, including Pfizer Endocrine Care. We are especially grateful to the major sponsors of this year's program, **Confidence Foundation** (who provided a generous grant in memory of Board Member **Chris Patay's** mother, **Sally Patay**) and **Southern California Edison** (whose award was made after nomination from PWCF member Nay Lay).

This year's attendees rated the meeting overall 3.6 out of a possible 4: "Thanks for the GEM!!!! It was great, especially on the anxiety talk! Kids loved it and lasted all day really well. Thanks a million!" "The presenters were fantastic, the information was extremely useful, and everything seemed to run very efficiently." "Great networking opportunity." "Thank you so much. I am really grateful for all the information. It was an excellent and well planned informative conference." "*Coping with Anxiety* was excellent and very useful - best part of the day." "The meeting was very informative, educational. The panel are [sic] definitely knowledgeable in their field. All three look like they will continue to collect data for PWS population and this is what we need." PWCF thanks the members of the 2009 Program Committee for their hard work to produce this year's exceptional program: **Julie Casey, Jenn Paige Casteel, Fran Moss, Wendy Young, Mary Hill, Kim Morgan, Ken Knox, Renay Compere, Lisa Graziano, Tracy Goatcher, and Diane Kavrell.**

Don't miss the 2010 General Education Meeting in Northern California featuring PWS experts Janice Forster, M.D. and Linda Gourash, M.D.!