THE WISCONSIN CONNECTION

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PWSA of WI, Inc. is a
chapter of PWSA (USA)
Website: www.pwsausa.org

The Newsletter of the Prader-Willi Syndrome Association of Wisconsin, Inc.

The mission of the Prader Willi Syndrome Association of Wisconsin, Inc. is to Support, Educate and Advocate for persons with Prader-Willi Syndrome, their families and professionals in meeting the challenges of this disability.

Notes from the Office

Program Director-Joshua Escher

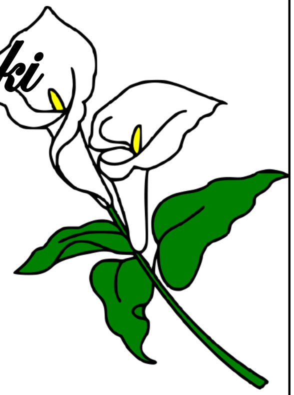
Hi folks, summer and the heat that comes with it are upon us. Remember that Prader-Willi Syndrome can cause poor temperature regulation. Keep an eye on your loved one and if you suspect the heat might be getting to them, help them to cool off. Encourage them to drink water as well!

We were really happy to see all the folks who came to our Special Needs Planning Presentations in June. Patrick shared tons of very useful information that families with a loved one with special needs have to know! Patrick volunteered a ton of his time for us so we told him to take this quarter off from writing his P.R.O.M.I.S.E. financial planning series. We have a new fundraising event coming up (see page 6) and of course out golf and bowling events. If you would like some support for an IEP or would like training for school or residential staff, let me know! Have a great summer!

In Loving Memory

Ryker Ploszczynski

Kelly Jones



Two beautiful PWS Angels left us recently. Many of their loving family and friends made generous donations to PWSA of WI in their memory.

We would like to congratulate Gavin Johnson, a staffer at Prader-Willi Homes of Oconomowoc, for receiving our Professional Service Provider Award. Gavin has shown himself to be a true champion of individuals with Prader-Willi. The person who nominated him wrote, "Gavin has become our favorite "go to" staff member, due to the high level of trust we have developed in him. He always has a ready smile and an upbeat attitude when we arrive to pick up our grandchild, which is such an important thing with our "kids." He remembers details and knows how to communicate therapeutically with this population. He is willing to go the extra mile. We've seen it firsthand several times.

Positive Behavioral Support is an essential tool for caregivers.

- Positive behavioral support can help decrease tantrums, reduce perseverative and OCD like behaviors, and increase cooperation and effective transitions through activities.
- Positive behavioral support can be used at a young age even before adverse behaviors begin.
- Positive behavioral support can be used to teach, reinforce, and strengthen desired behavior. As desired behaviors are strengthened undesirable behavior fades due to a lack of reinforcement.
- Positive behavioral support uses an individual’s strengths and interests as motivation to complete non-preferred tasks, meet goals, and learn new coping skills. Because individuals with PWS typically have low internal motivation, the use of external motivators is a must. External motivators include verbal praise, token rewards, sticker or other visual reward systems, or even special attention or one-on-one time.
- Always work with your loved one to determine what their individual motivators are understanding what may be motivating to one person might not be to another. It is important to work collaboratively with individuals with PWS to increase their investment in learning and growing.

This originally appeared on PWSAUSA.org as a blog post.

For more information about positive behavioral supports or suggestions of how to implement them, contact us!

PWSA of WI's 2019 On The Move Walkathon

2019 brought us our biggest walk yet! 357 people descended upon Watertown to celebrate PWS awareness month and to help us raise funds for PWSA of WI and PWSA (USA). Everyone looked super sharp in their tie dye. The weather was a little cool and breezy but mother nature kept us dry. We are already looking forward to seeing everyone next year!

We would like to thank all of our Corporate sponsors for supporting our walk!

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This year for May Awareness PWSA (USA) regularly posted some awesome "did you know" information about Prader-Willi on their Facebook page, we have decided to compile all these posts and share them here. This is a great start in sharing with friends and family about what PWS is.

- May is Prader-Willi Syndrome Awareness Month! Republican Congressman Ed Royce and Democratic Congresswoman Jane Harman co-sponsored the original House Resolution thanks to efforts by Amy Porter (Aunt to Colorado's young Abby Porter who has PWS) and Prader Willi California Foundation members Tom and Renay Compere (parents to California's William Compere) who brought this important cause to their attention. The House Resolution passed in December 2009, which then led to the Senate bill being passed on May 27, 2010, thereby establishing National Prader-Willi Syndrome Awareness Month!
- Prader-Willi syndrome is classified as a rare genetic disorder. The incidence rate of PWS is generally thought to be between 1 in 12,000 to 15,000 which means somewhere between 21,000 - 26,000 Americans have PWS. Because PWS is a rare disease, awareness and advocacy are essential to ensuring the needs of the PWS community are understood by all.
- Based on the incidence rate, there are somewhere between 21,000 and 26,000 Americans who have PWS. However, only about 8,000 are known to PWS organizations across the US. This means most cases are either undiagnosed or that the families have not connected with PWSA (USA).
  PWSA (USA) works to increase awareness so that all individuals with Prader-Willi Syndrome can be diagnosed AND receive the support they need. One day ALL individuals with PWS will be properly diagnosed at birth because EVERYONE will know about Prader-Willi syndrome.
- Prader-Willi syndrome (pronounced PRAH-der WILL-ee) is caused by a genetic defect on the 15th chromosome. PWS was first identified in 1956 by Swiss doctors A. Prader, H. Willi and A. Labhart. There is no known cause for PWS nor is there a cure.
  PWS is one of the ten most common conditions seen in genetics clinics, and is the most common genetic cause of childhood obesity.
- PWS is a randomly occurring genetic abnormality that takes place at conception. It affects both males and females and occurs across all races.
  The two most common types of PWS are Deletion and Uniparental Disomy (UPD). Deletion accounts for approximately 70% of cases and happens when an individual has a non-inherited deletion in the 15q11-q13 region of the paternal 15th chromosome. Uniparental Disomy (UPD) accounts for approximately 25% of cases and happens when an individual receives two maternal 15th chromosomes and no paternal 15th chromosome. In rare cases there can be an imprinting error where the father's chromosome is present but does not work properly (this type is very rare and therefore PWS is considered to be non-inherited).
- PWS causes the hypothalamus (considered the command center of the brain) to function improperly. This malfunction impacts a number of things, including metabolism, temperature regulation, various hormones, muscle tone, emotional reactions, stress response, and of course, the hallmark trait of PWS, appetite regulation.
  PWS is a spectrum disorder, meaning that PWS is not a one-size-fits-all diagnosis and every person with PWS has his or her own unique abilities and challenges.
- Hyperphagia, or excessive appetite, is a hallmark trait of Prader-Willi syndrome. Though the exact cause isn't fully understood, individuals with PWS lack the normal signals of feeling full and therefore ALWAYS feel hungry. Hyperphagia is often the symptom most often associated with PWS, however, it is one of many.
  Another symptom of PWS is very low metabolism (up to 50% of what is considered "normal'). When combined with hyperphagia, very low metabolism is a recipe for obesity and all the health problems associated with it.
  Through diligence in food control, an individual with PWS can be thin. However, individuals with PWS who are overweight are not so due to parental or caregiver negligence. He or she might have an extremely low metabolism (even for PWS) or might be on the extreme end of the hyperphagia continuum. They might have received a late diagnosis and are just now trying to take off the weight, they might not be aware of the best nutrition recommendations for PWS, or they might not have access to a physician familiar with they syndrome. No matter what, weight management in PWS is a life-long, extremely challenging issue.
- Hyperphagia is a hallmark symptom of Prader-Willi syndrome. However, individuals with PWS don't start out with an excessive food drive. In fact, most are diagnosed with "failure to thrive" in infancy because they have hypotonia (low muscle tone) and are too weak to eat.
  One of the mysteries of PWS is in trying to figure out exactly what causes the "switch" to hyperphagia. In the past, the age of onset was considered to be around the age of two, and while that is still true for some, many are now finding the onset of hyperphagia to come much later.

Continued on page 5

- Individuals with Prader-Willi syndrome are often diagnosed with "failure to thrive" as infants. This is due to global hypotonia, or decreased or low muscle tone, negatively impacts the many muscles of the mouth making it difficult for infants with PWS to suckle to eat. As such, many infants require assistance with feeding by way of NG tubes (feeding through the nose) or g-tubes (directly into the stomach) until they are strong enough to eat on their own.
  Hypotonia continues to impact individuals with PWS throughout their lives. Developmental milestones and speech are often delayed. Articulation can remain an issue long after speech is developed, and anything involving gross or fine motor skills (writing, swimming, riding a bike, etc.) takes much longer to develop or master. Exercise can improve muscle tone to some extent, however, individuals with PWS will oftentimes remain weaker than their peers, struggling with endurance throughout life.

- Individuals with Prader-Willi syndrome have a much slower metabolism than the average person...up to 90 times slower!
  Imagine always feeling hungry but needing only half the calories your friends do to survive. For individuals with PWS, it can be far too easy to gain weight yet difficult to get proper nutrition on calorie-restricted diets. This is why there is NO room for empty calories. Every calorie counts and any special "treats" must be accounted for in the daily calorie intake or with extra exercise.

- With help, people with PWS can, and do, accomplish many of the things their “normal” peers do. With proper support, they can finish school, obtain employment, and pursue their interests and hobbies. However, they can't do these things without significant support from their families and from school, work, and care providers. Even individuals with IQs in the normal range need lifelong diet supervision and protection from food availability.
  The prevention of obesity (and the many health risks associated with it) is helping those with PWS live longer and more healthfully. New medications, including psychotropic drugs and synthetic growth hormone, are already improving the quality of life for many people with PWS. Ongoing research offers the hope of new discoveries that will help individuals with PWS thrive in the face of a rare genetic condition.

- Prader-Willi syndrome is classified as a rare genetic disorder. The incidence rate of PWS is generally thought to be between 1 in 12,000 to 15,000 which means somewhere between 21,000 - 26,000 Americans have PWS. Because PWS is a rare disease, awareness and advocacy are essential to ensuring the needs of the PWS community are understood by all.

- Hypogonadism (a condition in which the testicles are not working the way they should) and cryptorchidism (undescended testes) are both common newborn males with PWS and can lead to delayed puberty and infertility. A study that looked at the long-term effects of orchidopexy (corrective surgery for undescended testicles) in children with PWS found the surgery to be only minimally successful, therefore parents will want to carefully consider the risks verses benefits of this treatment option.

- The “Normal” body temperature in individuals with PWS can vary significantly from one person to the next. It is important to know your loved one’s usual “healthy” temperature which can be established by monitoring his or her temperature under different circumstances and at different times. In addition, individuals with PWS can easily become too hot or too cold meaning keeping comfortable might require wearing clothing in layers to allow for easy adjustments.

- A 2018 research study found children and adolescents with PWS experienced reduced salivary secretion, increased salivary viscosity, and mouth breathing. Investigators recommended that individuals with PWS have dental care focused on the prevention and treatment of tooth wear and gingivitis and limiting the negative influence of reduced saliva. Salivary secretion stimulation can be improved by the application of topical fluoride treatments and chewing sugar-free gum containing xylitol and CPP-ACP.

- Some clinicians suggest growth hormone (GH) therapy is contraindicated in the presence of scoliosis. However, a multicenter randomized controlled trial found no difference in onset or curve progression between children undergoing GH treatment and those children not on GH. In fact, some children treated with GH experienced less severe scoliosis suggesting a protective effect of GH treatment.

- Skin picking is a common behavior seen in people with Prader-Willi syndrome (PWS). Intensity and duration vary from person to person and each episode might be different. For some, stress is a contributing factor to an individual’s tendency to engage in skin picking. If the source of stress can be removed, skin picking should also reduce. Remember, though, that skin picking can become habitual and repetitive meaning it might take time to cease altogether. Refer to Medical A-Z on the PWSA (USA) website for ways to prevent and control skin picking behaviors.

2020-2021 Board of Director Openings

PWSA of WI will have 7 open board positions to begin in January of 2020. Board positions are 2 year positions and elections are held each October. If you are interested in running for a board position or would like more information contact our Program Director Josh at progdir@pwsaofwi.org. Board Applications must be submitted by September 15th.

Prader-Willi Syndrome Association of WI, Inc. 19th Annual Golf Benefit

PWSA of WI
Annual Golf Benefit
SUPPORT • EDUCATE • ADVOCATE

Sunday, September 29th, 2019
The Oaks Golf Course
Cottage Grove, WI

Prizes, Food, and Most of All Fun!

THE PRADER-WILLI SYNDROME ASSOCIATION OF WI
welcomes you to an evening that will inspire hope, raise community awareness, provide education and medical research, and encourage heartfelt support

Sparkle & Shine Gala

BE THE REASON FOR THE SPARKLE IN THEIR LIVES

Dinner | Cash Bar | Silent Auction | Live DJ
SATURDAY, AUGUST 17, 2019 | 5:00-10:30 PM

RED CIRCLE INN & BISTRO
N44 W33013 WATERTOWN PLANK ROAD | NASHOTAH, WI 53058

For more information on these events and to register, visit pwsaofwi.org.

CAREPWS

Clinical Study Recruiting for Prader-Willi syndrome (PWS)

CARE-PWS is a clinical study evaluating an investigational medicine for the treatment of Prader-Willi syndrome (PWS) in 7-18 year olds. People with PWS have a constant feeling of hunger, known as hyperphagia. Find out more.
https://www.carepwsstudy.com/qualify/?utm_source=Seeker&utm_medium=newsletter

Unmask the Possibilities

PWSA (USA) NATIONAL CONVENTION 2019

AWARENESS FAMILY SUPPORT RESEARCH EDUCATION ADVOCACY

Agenda of Events

Chapter Leaders Meeting - October 23rd & 24th

Medical & Scientific Conference – October 23rd & 24th

New Parent Mentors Meeting – October 24th

Professional Providers Conference – October 24th

Welcome Reception – October 24th (6pm – 9pm)

PWSA (USA) General Conference – October 25th & 26th

Youth & Infant Program

Youth & Adult Program

Sibling Program

PWSA (USA) National Convention

Date: October 23rd – 26th, 2019

Hotel: Caribe Royale

Location: Orlando, Florida

PWSA of WI provides scholarship to PWSA of WI members from Wisconsin. Visit our website pwsaofwi.org for more details or contact our program director progdir@pwsaofwi.org or 920-733-3077.

To purchase PWSA of WI merchandise visit pwsaofwi.org/merchandise

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PWSA of WI, Inc.'s Event Calendar

August 17th, 2019	**Sparkle and Shine Gala**	**Red Circle Inn & Bistro, Neshotah, WI**
September 29th, 2019	**PWSA of WI Golf Benefit**	**The Oaks, Cottage Grove, WI (New Location)**
October 12th, 2019	**Strike Out PWS Bowling Event**	**Bowlero, Wauwatosa, WI**