THE WISCONSIN CONNECTION

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.

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Letter from the Office

Program Director

Joshua Escher

Welcome to Fall! The leaves are changing and the temperature is dropping. I for one am excited for the tree outside my office window to give me my sunlight back for a few months. Of course changing temperatures can be a big challenge for folks with Prader-Willi because of the challenges they have with body temperature regulation. Make sure you are ready for all kinds of temps, inside and out, for your loved one with PWS.

Check out page 5 of this newsletter for a little recap of our annual golf benefit. After a year away, we were extremely excited to have gotten back to the links. A great time was had by all. If you head over to our Facebook page you can see some pictures of the teams and other fun stuff from the day. We are already looking forward to next year.

Now that golf is done with I am already thinking about fun things we can do during the colder months. Virtual Bingo is coming back for sure and I may just have to resurrect the virtual pub quiz as well.

As always, if you have any questions or needs, reach out. I am here for you!

Our **Caring Is Sharing** fundraising campaign begins November 1st. As a small nonprofit organization, Prader-Willi Syndrome Association of WI, Inc is dependent on the generosity of people like you to maintain the programs and services we provide. Keep an eye on your mailbox as a letter requesting your support will be arriving soon. Every little bit helps so please consider making a donation. People like you are why we are able to continue doing what we do.

Soleno Therapeutics Announces Positive Data Showing Continued Significant Improvements In Symptoms of PWS Following One Year Treatment With DCCR

- Statistically significant reduction in hyperphagia and all other PWS behavioral parameters in Study C602
- Statistically significant improvements compared to natural history of PWS from the PATH for PWS Study
- On track for data submission to the FDA in Q3 2021

REDWOOD CITY, Calif., Sept. 08, 2021 (GLOBE NEWSWIRE) — Soleno Therapeutics, Inc. ("Soleno") (NASDAQ: SLNO), a clinical-stage biopharmaceutical company developing novel therapeutics for the treatment of rare diseases, today announced top-line results from the Company's ongoing open-label extension study, C602, evaluating investigational, once-daily DCCR (Diazoxide Choline) Extended-Release tablets for patients with Prader-Willi syndrome (PWS) and its comparison to data from the PATH for PWS natural history study.

A total of 115 subjects were enrolled into C602, the extension study in patients with PWS who completed DESTINY PWS, an international, multi-center, randomized, double-blind, placebo-controlled study of DCCR in 127 PWS patients at 29 sites in the U.S. and UK. There are 95 subjects continuing to receive DCCR in C602. The PATH from PWS (PfPWS) study is an ongoing study sponsored by the Foundation for Prader-Willi Research (FPWR) to advance the understanding of the natural history in individuals with PWS. Data from subjects who received DCCR for 26 and 52 weeks were compared to results from matched subjects from the PfPWS. In all of these studies, changes in hyperphagia and PWS-related behaviors were assessed using the same questionnaires: the Hyperphagia Questionnaire for Clinical Trials (HQ-CT), a caregiver completed 9-item validated questionnaire for assessing hyperphagia in PWS, and the Prader-Willi Syndrome Profile (PWS-P), which consists of caregiver responses to questions in six domains: aggressive behaviors, anxiety, compulsivity, depression, disordered thinking, and rigidity-irritability.

Key Top-line Results of Study C602

Hyperphagia: The mean (SE) improvement in hyperphagia, the primary endpoint in the DESTINY PWS study, represented by a decrease in the HQ-CT total score, of -9.9 (0.77), was highly significant ($p<0.0001$) after receiving DCCR for 52 weeks. Highly significant improvements in hyperphagia were also observed after receiving DCCR for 13, 26, and 39 weeks (all $p<0.0001$).

PWS related behaviors: Behaviors related to PWS were measured using the PWS Profile Questionnaire (PWS-P). After 52 weeks, there were statistically significant improvements in all behavioral domains (all $p<0.0001$) and after receiving DCCR for 13, 26, and 39 weeks (all $p<0.0004$).

Key Top-line Results of Study C602 Compared to Matched Subjects from PATH for PWS

For the comparison of C602 to PfPWS, statistical techniques were employed to minimize and/or reduce selection bias during subject matching and to account for the impact and timing of the COVID-19 pandemic. Matching of subjects to and comparisons with PfPWS were conducted by an external CRO independent of Soleno using a pre-specified Statistical Analysis Plan.

Hyperphagia: PfPWS subjects showed a mean (SE) change of -2.5 (0.38) and -3.4 (0.48) after 26 and 52 weeks compared with that of -8.4 (0.72) and -9.4 (0.74) in matched C602 subjects, showing a statistically significant improvement with DCCR ($p<0.001$ for both comparisons).

PWS related behaviors: As with hyperphagia, statistically significant improvements with DCCR in C602 subjects compared with matched subjects in the PfPWS study were seen in all behavioral domains of the PWS-P after 26 and 52 weeks ($p<0.003$ for all).

Additional Results from Study C602 after One Year

Body Composition Changes: Patients in study C602 showed no significant change in body fat mass and statistically significant improvements in lean body mass ($p<0.0001$) and the ratio of lean body mass to fat mass ($p=0.0005$).

Endocrine and Metabolic Changes: Subjects demonstrated highly significant improvements in leptin ($p<0.0001$), adiponectin ($p<0.0001$), and fasting insulin ($p=0.0004$). Significant improvement in insulin sensitivity measured using the Homeostatic Model Assessment of Insulin Resistance (HOMA-IR) ($p=0.03$) was also observed.

Safety: The safety profile of DCCR remains consistent with the known safety profile of diazoxide and the prior experience with DCCR with increases in blood glucose levels, hypertrichosis and peripheral edema being the most common adverse events. No serious, unexpected, related adverse events have occurred with DCCR in the program to date.

Cont. on page 3

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"Hyperphagia is one of the main problems associated with PWS and can have life-threatening consequences such as severe obesity. No treatments are currently available," said Dr. Evelien Gevers, M.D., Paediatric Endocrinologist and Reader at Barts Health NHS Trust and Queen Mary University of London and the Coordinating Investigator for the UK in the DESTINY PWS and C602 studies. "Collectively, these data further demonstrate that DCCR significantly reduced hyperphagia, as well as improved body composition, behaviors, and endocrine and metabolic measures over 52 weeks. If approved, DCCR could offer people with PWS a meaningful option for long-term management of their symptoms."

"The PATH for PWS study is an ongoing evaluation of the natural history of individuals with PWS," said Dr. Theresa Strong, Director of Research Programs for FPWR. "These data showing a significant improvement compared to the PATH data suggest that DCCR may provide a much needed and meaningful improvement to the lives of those living with PWS and their families. We look forward to supporting the Soleno team as they advance DCCR through the regulatory process."

"We are extremely pleased with these results from our ongoing open-label extension study and their comparison to the PATH for PWS study," said Anish Bhatnagar, M.D., Chief Executive Officer of Soleno Therapeutics. "We look forward to submitting the data from these studies to the U.S. Food and Drug Administration ("FDA") in this quarter and remain firmly committed to the goal of obtaining approval for DCCR as a new treatment for people with PWS as expeditiously as possible."

Soleno will submit these data to the FDA as part of an ongoing discussion with the Agency regarding the clinical data necessary to support the submission of a New Drug Application (NDA) to market DCCR for the treatment of PWS. The FDA has previously conveyed to Soleno that another clinical trial will likely be needed and that open-label data and comparisons with natural history sources such as PATH for PWS may have statistical and other limitations, but it has agreed to review the data to determine whether it is appropriate for the Company to submit an NDA.

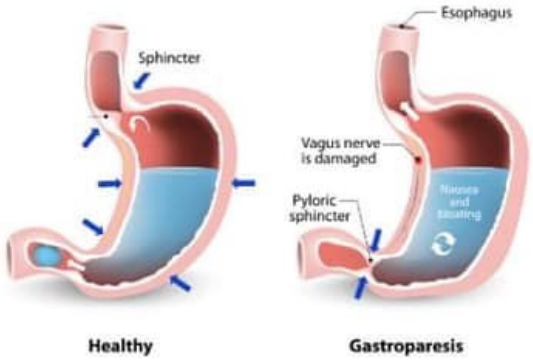
For further information about DESTINY PWS (NCT03440814) and the open-label extension study Study C602 (NCT03714373), please visit: www.clinicaltrials.gov.

Thank you to Dr. Ann O'Shea Scheimann!

On September 17th Dr. Scheimann did a virtual presentation for our organization about GI issues in Prader-Willi Syndrome. This event was attended by numerous parents and professionals and the recording has been shared with medical providers that were unable to attend. Prader-Willi Syndrome of Wisconsin has a goal to try to provide at least one educational opportunity per year. We were very lucky to have Dr. Scheimann with us once again.

AUGUST IS GASTROPARESIS AWARENESS MONTH

Nearly every person with PWS has **some degree of slow gastric emptying**, also known as gastroparesis.



What is Gastroparesis?

Gastroparesis is a chronic condition that affects the stomach muscles and prevents proper movement of food from the stomach to the small intestine.

Symptoms:

heartburn
acid reflux
vomiting undigested food
bloating
stomach pain

To help us learn more about GI issues in PWS, please complete the Gastrointestinal History survey in the Global PWS Registry.

www.pwsregistry.org

Do you have a teacher, doctor, care provider, or other professional that you think deserves to be recognized for their service to the PWS community? We want to reward them! Visit our website at https://pwsaofwi.org/ProRecognition and fill out an application today! Each winner will receive a certificate, a card, and a $50 gift card.

Did you know that you can donate to PWSA of WI just by shopping on Amazon? Go to **smile.amazon.com/ch/39-1732251** to set us as your preferred charity!

******We would like to thank anyone who has chosen to use employer matching to support our organization! We are not always provided with the information about who makes the donation so that we can individually thank you so we wanted to make sure all of you are acknowledged! ******

SUPPORT • EDUCATE • ADVOCATE

PWSA OF WI
ANNUAL GOLF BENEFIT

On September 26th thirty six golfers decended upon The Oaks in Cottage Grove to help us raise money at our annual golf benefit. We had the best weather we have had in years and a wonderful time was had by all!

A big thank you to all our donors:
James Jones
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Milwaukee Milkmen
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Barb & Don Dorn-In Honor of "Bud" Tony Dorn - Love Mom & Dad
Barb & Don Dorn-In Honor of Tony Dorn - Love Mom and Dad
Rebecca & Ryan Norman-In honor of Tony Dorn
James & Janis Becker-In honor of Tony Dorn
Mike Schultz-CGC Inc In Honor of Tony Dorn
Mike Schultz-CGC Inc In Honor of Tony Dorn
Mark Dorn-In Honor of Tony Dorn
Laurie Grinnell-In Honor of Tony Dorn
Larry & Mary Dall-In honor of Tony Dorn
Crystal & Clint Boser-In Honor of the Guys at Anthony Home
Crystal & Clint Boser-In honor of Kyle Page
Cathie Lehman-In honor of Martha Lehman
Prader-Willi Homes-Logo
Fresh Coast Swag-In honor of Tony Dorn
Friends of Tony Dorn-Cathy, Mary, Alice, Sandy, Vicki, Debbie, and Cindy-In Honor of Tony Dorn

Huge extra special thanks to our beverage cart sponsor the David & Sandra Nagy Family

Save the Orphan Drug Tax Credit!

The orphan drug tax credit is crucial to assist and encourage pharmaceutical companies to develop therapies for rare diseases!

History

*An Orphan Drug is a pharmaceutical that is created in order to treat a rare disease. The orphan drug tax credit (ODTC) is a federal tax credit available to life science companies working to find cures for diseases that affect small populations. The credit is designed to encourage the development of treatments for rare diseases.

*Between 1983 and 2018, the orphan drug tax credit provided a 50% credit for qualified clinical trials with human subjects, and research grants to promote the development of new treatments for orphan diseases.

*A 2017 overhaul of the tax code under the Trump administration reduced the credit from 50% to 25% beginning in 2018.

*The ODTC has led to approvals for more than 780 products to treat more than 250 rare diseases (most of which are cancer treating therapies). However, there are more than 7,000 rare diseases and only a few hundred have therapies that are indicated for their disease.

*The central rationale behind this piece of legislation was to provide an incentive to invest resources into developing drugs to treat an extremely small patient pool

*Since the FDA grants orphan drug status to a specific use of a particular drug, it's possible to obtain orphan drug status for multiple uses of the same drug.

Current Situation

On September 13th, the House Ways and Means Committee released a first draft of the tax provisions to be included in the Biden Administration Build Back Better Act. It has a provision that would amend the Orphan Drug Tax Credit (ODTC) in ways that would severely undercut the original goal of the 1983 Orphan Drug Act. Section 138141 of the Build Back Better Act would remove the pharmaceutical tax credit for all but the first approved orphan use of a new drug. There was a push at this week's committee meeting to take out that provision, but it failed. Rare Disease Advocacy organizations and patients are meeting with Members of Congress and staff to advocate for the removal or refinement of this provision to ensure it does not hurt communities that would benefit from further study and approval of an on-market therapy.

Contact your representative and let them know that this is important to you, one of their constituents!

Mediterranean Diet Mini Omelets

Oven Baked Mini Omelets

Ingredients

- Olive Oil – 2 teaspoons or as needed.
- Eggs- 8.
- Half and Half- 1/2 cup. Feel free to sub in other dairy products. Dairy-. You can use milk, skim milk, half and half, sour cream, Greek Yogurt or a mixture of two. Your choice of dairy should have something to do with how much fat, calories, carbs, you want in your eggs versus the consistency and taste.
- Cheese- 1/4 cup. I am a fan of shredded cheddar cheese in this recipe. But, if I am feeling 'extravagant' I may use shredded Gruyere cheese.
- Chopped Vegetables- 1 cup. This can include chopped peppers, broccoli, green onions, or even spinach.
- Seasonings to taste- This can include Italian Seasonings, Salt, and/or Pepper.

Instructions

Preheat your oven to 350. Prep your muffin pan or ramekins with the olive oil as this recipe is prone to sticking. You can also spray a bit of nonstick over this.

In a mixing bowl use a whisk to beat together the eggs and dairy until they are fluffy.

Next, stir in the cheese and any desired seasoning.

Fold in any vegetables or remaining ingredients and then evenly transfer the mixed ingredients to the prepared ramekins or muffin pan.

Bake your omelets until the appear to be firm. Depending on what you are baking them i, this can be 25-30 minutes. Remove from oven and allow them to set for five to ten minutes before serving.

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Keep an eye out, we will be bringing back family bingo nights as the weather gets cooler. Make sure you follow our Facebook page and that we have a current email address for you so you don't miss out!

Do you know someone with Prader-Willi who no longer has family to provide them with gifts on their birthday or Christmas?

PWSA of WI has a gift giving program for these folks! Please contact us for more information so we can make their day.

Did you know PWSA of WI has a card club for individuals with PWS?!

Contact our program director at progdir@pwsaofwi.org to have birthday cards and seasonal cards sent to your loved one with PWS!

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WISCONSIN Prader-Willi SYNDROME ASSOCIATION (USA) SAVING AND TRANSFORMING LIVES

PWSA of WI, Inc.'s Event Calendar		
Snowflake Ball	January 2022	TBD
On The Move Walkathon	May 8th, 2022	TBD