



PWS FAMILY NEWSLETTER

Spring / Summer 2024 Issue

Endocrinologist's Corner: Promising Study Drug

by Dr. Sejal Kadakia, M.D.

Many of you may have already received the wonderful news that DCCR, also known as diazoxide choline extended-release, (Solenio Therapeutics Inc., medication study for hyperphagia), has received Breakthrough Therapy Designation from the FDA. We understand that there may be questions about what this designation entails. It's important to note that this designation is designed to streamline the development of new drugs for severe conditions, particularly when initial data suggests that the drug may offer significant advantages over current therapies. However, it's crucial to emphasize that the drug is not yet approved, and there's no guarantee of future approval. Nonetheless, this designation is expected to expedite the process.

Rest assured, our team is actively monitoring updates, and we'll keep you informed about any advancements regarding the approval and availability of this therapy. Stay tuned for further updates!! Here is a link to an article about it from the NIH.

[Diazoxide choline extended-release tablet in people with Prader-Willi syndrome: results from long-term open-label study - PubMed \(nih.gov\)](#)

Recipe Corner

Healthy Banana "Ice cream"



[A recipe worth repeating!](#)

Ingredients:

- 1 Small Banana
- 1 Tbs Unsweetened Vanilla
- Almond Milk, add more as needed to make it smooth.

Directions:

1. Cut ripe banana into 4 pieces and freeze.
2. Add frozen bananas and unsweetened vanilla almond milk to food processor or blender.
3. Blend until smooth and portion into 3 containers, serve as is or freeze for firmer consistency.

1/3 of recipe: 30cal, 6.7g carb, 0g fat, 0.3g pro.

Optional: Swap out the banana for your favorite fruit, such as strawberries or blueberries, to create a variety of flavors!

Behaviorist and Dietitian Corner:

Understanding the No Doubt, No Hope, No Disappointment Concept: for Families with Prader-Willi Syndrome and Implementing it in Daily Life

By: Christina Wright-Yee, MPH, RD, CSP and Geeta Grover, MD, FAAP

If you have a family member with Prader-Willi Syndrome (PWS), it's important to understand the concept of "No Doubt, No Hope, No Disappointment." This concept means that we should avoid creating uncertainty (doubt), false expectations (hope), or setting someone up for disappointment. In this article, we will explain the concept and provide some ways to implement it in your day-to-day lives to create a more positive and supportive environment for your loved one with PWS.

Understanding the Concept:

"No Doubt, No Hope, No Disappointment" is a behavioral management strategy that emphasizes "high warmth, high structure parenting" that encourages clarity and consistency, which are essential for individuals with Prader-Willi Syndrome. It means avoiding situations where someone might not be sure what's going to happen (doubt), making promises, or creating expectations that might not be met (hope), and setting unrealistic goals that can lead to frustration (disappointment). An example is that there will be no extra meals beyond scheduled mealtimes.

Implementing the Concept in Daily Life:

- ✓ **Clear Communication:** Be clear and direct in your communication with your family members with PWS. Use simple language and give precise instructions, avoiding vague or ambiguous statements that can create doubt or confusion.
 - Example: Instead of saying, "We will eat later," say, "We will eat at 5:30 p.m."

- ✓ **Predictable Routine:** Establish a consistent daily routine for your loved one. Knowing what to expect helps reduce anxiety and uncertainty. Stick to a schedule for meals, activities, and bedtime, ensuring everyone in the family is aware of and follows the routine. This way, you can avoid creating false hope or disappointment caused by sudden changes in plans. When life necessitates a temporary change, prepare your loved one if possible, by explaining the schedule or routine will be different in advance.
 - Example: "Tomorrow we are going to the zoo, so our meal times will be a bit different based on traffic and the fun we are going to be having. We are going to have so much fun walking at the zoo and seeing different animals."
- ✓ **Realistic Goals:** Set achievable goals that take into account the abilities and limitations of your family member with PWS with high warmth along with high structure.
 - Celebrate small victories and progress, rather than focusing on unreachable milestones.
 - By setting realistic goals, you can encourage their self-confidence and help them avoid disappointment.
 - *Set a SMART goal using the tool on the next page! Bring your filled out SMART goal for a prize at your next clinic visit!*
- ✓ **Supportive Approach:** Offer unconditional support and understanding to your loved one with PWS. Encourage them to express their emotions and concerns and provide reassurance and guidance when they face challenges. Remember, by embracing the "No Doubt, No Hope, No Disappointment" concept, you can help create a safe and nurturing environment for your family members.
 - "I will always love you, but I am not happy with the way you are behaving right now".



In conclusion, the concept of "No Doubt, No Hope, No Disappointment" is a valuable mindset for families with members who have Prader-Willi Syndrome. Remember, embracing this concept helps foster a positive and understanding atmosphere, benefiting everyone in the family.

Valentine's, Easter, or birthdays can be particularly difficult for someone with PWS. The challenge is in taking a holidays or specials days represented by gifts and candy and changing the meaning. On these days, when children traditionally receive a bounty of sweets, try creating a cheerful holiday in which food and candy or Easter egg hunt is filled with games and activities that are predominantly educational. Rather than candy and chocolate, students hunt for alternative items that can sometimes be great sensory items. We suggest:

- Jump Rope— a perfect tool for exercise and gaining balance skills.
- Silly Putty Eggs— a great sensory item.
- Bubbles— another great sensory item that also promotes good deep breathing skills.
- Pinwheels or Wooden Whistles— items that also help with breathing.
- Flower Seeds for Spring Garden— we love this idea because it not only promotes planning and organizing, but also provides an opportunity to spend quality family time together in the warmer days ahead.
- Memory Games— these are an awesome way to improve children's memory and executive functioning skills.
- Legos or Coloring Books Made for Older Kids and Adults— these are fun but also enhance children's fine motor skills.

Social Work Corner: Legal Issues, Conservatorship for PWS

By Bobbi McGann, LCSW

If your PWS child is developmentally disabled and will not be able to make medical decisions and care for themselves independently upon reaching the age of 18, you will need to go through a legal process called Limited Conservatorship. Regardless of your child's intelligence, level of insight and verbal acuity, adults with PWS lack adequate judgement and not capable of making appropriate self-care decisions.

Rights which are frequently conserved for PWS adults are the following:

- The right to decide and fix the residence. Most adults with PWS cannot make appropriate decisions regarding their residence because they will have difficulty navigating food independently.
- The right to access confidential records and papers. Most patients with PWS are unable to understand abstract and complex concepts. They can also be hypersensitive to criticism. PWS adults will need help to interpret records.
- The right to consent to marriage. Most PWS adults want to marry and have children, however most adults have egocentric (thinking of oneself without regard to others) can interfere with the ability to make informed decisions about relationships.
- The right to contract. Adults with PWS lack the cognitive ability, judgement, and skills to understand contracts.
- The right to give and withhold medical consent. PWS adults have inadequate body awareness to assess the severity of illness and injury. They



- also lack the ability to weigh potential consequences or outcomes to their actions.
- The right to control social and sexual contacts and relations. All PWS adults crave social connections, however they can be manipulated and taken advantage of. PWS adults need guidance with this area of their life.
- The right to make decisions regarding education. PWS adults should be supervised in the community, including educational training.

Since there is a wide spectrum of PWS, not all adults with PWS will need the same degree of conservatorship support. It will be important to work with a legal representative to discuss what your child may need.

Please reach out to the PWS Foundation for assistance. Info@pwfc.org

New update from CHOC/Cal Optima

For those Cal Optima patients who are in the process of obtaining conservatorship who are over 18, the provider can now write a capacity letter. This letter can be provided by the treating physician and would authorize the personal representative/individual identified in the letter to make healthcare decisions including access to PHI and the authority to make changes on behalf of the member. This authority would expire within 90 days, at which point a new capacity letter would need to be resubmitted. Ideally a more permanent result would be to obtain a power of attorney, conservatorship, or advance directive. However, until a permanent solution can be obtained a capacity letter can be submitted.

Fun Candy Swaps

By: Christina Wright-Yee, MPH, RD, CSP

If you and your family celebrate “food-based” holidays, where food is a large focus (Halloween, Easter, Holi, Ramadan), it can be hard for children with Prader Willi Syndrome to feel they can participate. Rather than giving candy and chocolates or food in gifts or baskets, try some of these ideas instead:

- Jump Rope
- Silly Putty Eggs
- Bubbles
- Pinwheels or Wooden Whistles
- Flower Seeds for Spring Garden
- Memory Games
- Legos or Coloring Books Made for Older Kids and Adults– these are fun but also enhance children’s fine motor skills.
- Temporary tattoos
- Balls
- *Adapted from Living Healthy with PWS Cookbook and Nutrition Tips.* lathamcenters.org



“Lower Calorie” Deviled Egg Chicks

- 12 eggs, hard-boiled
- 1/4 cup plain unsweetened Greek yogurt
- 1 tsp Dijon mustard
- 1 TBSP dill relish
- 1/2 tsp paprika
- 1/2 tsp salt
- 1/8 tsp dried parsley
- 1/2 tsp paprika
- 1/2 tsp salt
- 1/8 tsp dried parsley
- dash of lemon juice or white vinegar
- 6 black olives
- 1 carrot for feet/nose sliced for nose



1. Boil the eggs.
2. While the eggs are boiling, combine everything in a bowl except for carrots (feet and nose) and peppercorn (eyes). Refrigerate for the next step.
3. Wash & peel the carrot and cut into 12 rounds. Cut each round in half (these are the 24 chick feet). Next cut two triangles from the straight side of the half-round. Save the triangle cuts for the noses.
4. Once the eggs are cooked, peel them. Cut each egg into half. The cooked yolk should just fall right out. Place half of the yolk into the Greek yogurt mixture. Save or throw away the other half of the egg yolk. Repeat this step on all eggs.
5. Use an electric mixer to whip the yolks together with the Greek yogurt mixture. Scoop this filling back into the eggs and put the top of the eggs back on.