United in Hope FOR PWS

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Prader-Willi syndrome: Nutrition Guidance and Recommendations

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The Endocrinologist’s Perspective
# PWS Nutritional Phases

<table>
<thead>
<tr>
<th>Phase</th>
<th>Median ages</th>
<th>Clinical characteristics</th>
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<tbody>
<tr>
<td>0</td>
<td>Prenatal to birth</td>
<td>Decreased fetal movements and lower birth weight than sibs</td>
</tr>
<tr>
<td>1a</td>
<td>0–9 months</td>
<td>Hypotonia with difficulty feeding and decreased appetite</td>
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<tr>
<td>1b</td>
<td>9–25 months</td>
<td>Improved feeding and appetite and growing appropriately</td>
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<tr>
<td>2a</td>
<td>2.1–4.5 yr</td>
<td>Weight increasing without appetite increase or excess calories</td>
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<tr>
<td>2b</td>
<td>4.5–8 yr</td>
<td>Increased appetite and calories, but can feel full</td>
</tr>
<tr>
<td>3</td>
<td>8 yr to adulthood</td>
<td>Hyperphagic, rarely feels full</td>
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</tbody>
</table>
Progression to Hyperphagia

**Phase 1a/1b**
- Dysfunctional Hypothalamus
  - Abnormalities in size & function

**Phase 2a**
- Peripheral Hyperinsulinemia
  - Increase in glucose uptake
  - Increase in lipogenesis
  - Weight gain

**Phase 2b**
- Decreased leptin sensitivity
  - Increased Appetite
  - Weight gain/Obesity

**Phase 3**
- Obesity
  - Leptin Resistance
  - Lack of Satiety

**Phase 1a/1b**
- Abnormal Central Insulin Signaling
  - Impacts hepatic glucose output
  - Increased hepatic glucose output
  - Increased insulin secretion

Graphs show:
- Fasting Leptin by nutritional phase
- Fasting insulin by nutritional phase
Glucose and Insulin in PWS

- Individuals with PWS can have hypoglycemia in infancy, which improves with growth hormone treatment.
- Glucose levels tend to be within normal range in individuals with PWS.
- There is increased insulin sensitivity in individuals with PWS.
- Elevated insulin signaling in the face of increased insulin sensitivity in young children with PWS is a probable trigger for weight gain and onset of hyperphagia in this population.
Insulin Response to Sweet Taste in PWS

![Bar chart showing insulin response to sweet taste in PWS with p-values.]

- Insulin (mU/mL)
- Categories: 1a, 1b, 2a, 2b, 3
- p-values: p=0.0158*, p=0.033*
Other Metabolic Changes in PWS

- As insulin release increases it begins to block leptin entry to brain, creating leptin resistance at blood-brain-barrier (this is not unique to PWS).

- Leptin levels rise because the brain starts to not see the leptin it was used to seeing.

- Leptin is a satiety hormone, so brain not seeing it = increased appetite
Hormone Changes in PWS through the nutritional phases

- As adipose tissue amount increases in PWS, insulin and leptin continue to increase.
- Ghrelin (hunger hormone) continues to go up as metabolic changes in nutritional phases cause increased appetite drive
- Adiponectin (cardioprotective hormone) decreases as adipose tissue amount increases
Natural History of PWS

Metabolic Changes
- Increased insulin
- Increased leptin
- Increased ghrelin
- Low adiponectin

Body composition changes
- Increased weight gain
- Increasing body fat

Appetite Changes
- Progression through nutritional phases
- Insatiable appetite
Nutritional Guidance for all ages

- Using complex carbs (whole grain) vs simple carbs (white flour, processed) helps mediate insulin response
- Pairing carbs with a protein or healthy fat helps mediate insulin response
- A very low carb diet is not recommended because too much of anything (e.g., dietary fat) will also cause increased insulin response
- Key to good nutrition is balance – a well-balanced diet results in appropriate weight gain, growth, muscle mass
Prader-Willi Syndrome
Nutrition Guidance and Current Recommendations

Michael Tan, MS, RD, LDN, CDCES
Nutrition Guidance and Current Recommendations

Overview

- **Traditional** Guidelines
- **CURRENT** Recommendations
  - Quality
  - Quantity (portion sizes)
  - Balance (protein, carbs/fiber, fat)
  - Variety
  - Drinks
- Convenience
- Meal Scheduling
- Special Occasions
- Family Responsibilities
- Summary
TRADITIONAL PWS Nutrition Recommendations

- Strict calorie-restriction targeting a specific weight
- Disadvantages:
  - Limits dietary fat intake due to high calorie content
    - Dietary fats are important for brain health, growth, development, satiety
  - Does NOT limit sugar substitutes/non-nutritive sweeteners (NNS)
    - NNS may increase appetite
  - Calorie counting can be tedious, labor intensive, and time-consuming
  - Inappropriately associates weight/lower calorie to health
Nutrition Guidance and Current Recommendations

CURRENT PWS Nutrition Recommendations

- Unfortunately, there is a lack of clear, standardized medical nutrition therapy for PWS
- Current nutrition recommendations are not necessarily unique or highly specialized, but typically are stricter and more disciplined
  - Focuses on sustenance (physiologic nourishment) vs indulgence
    - De-emphasizes indulgence, celebratory/glorification of food
    - Minimizes/limits NON-hunger eating (emotional, behavioral, time-based, food as a reward, etc.)
    - Due to significantly lower calorie needs, there are less opportunities for “empty” or low-nutrient density foods
  - Limited flexibility (due to absolute/rigid thinking, “black and white” thinking)
- Recommendations are based on Mediterranean diet
- Recommendations focus on four key aspects
  - Quality
  - Quantity (i.e. portions)
  - Balance
  - Variety
Nutrition Guidance and Current Recommendations

QUALITY

- Focus on
  - WHOLE grains, beans, legumes
  - Vegetables, fruits
  - Protein (emphasizes fish and plant-based proteins: nuts/seeds)
    - Other animal-based proteins and dairy are OK too
  - Extra virgin olive oil (unsaturated fat)

- Limits
  - Highly processed, “convenience” foods (“bars”, breakfast cereals, jelly/jams, etc.)
  - Refined grains
  - Excessive added sugar
  - Sweets and treats and other “sweet-tasting” foods

- NO sugar substitutes/sweeteners
Examples of Sweeteners

- Acesulfame Potassium - Sunnett, Sweet One
- Aspartame - Nutrasweet, Equal
- Neotame
- Soluble Corn Fiber
- Saccharin - Sweet 'N Low, Sweet Twin, Sugar Twin
- Sucralose - Splenda
- Stevia/Rebaudioside - A Sweet Leaf, Sun Crystals, Steviva, Truvia, PureVia
- Allulose
- Monk fruit
- Sugar alcohols – xylitol, erythritol, mannitol, sorbitol, maltitol
- Tagatose
- Advantame
- Inulin (chicory root)
- Tapioca Fiber (prebiotic fiber)
Nutrition Guidance and Current Recommendations

QUALITY

- Guidance on “sweet tasting” foods
  - Fruit
    - Fresh fruit only
    - Limited frequency (twice per day)
    - Limited portion sizes
    - Always balance with protein/fat
  - Common “sweet tasting” foods generally NOT recommended
    - Sweetened DRINKS
    - Breakfast cereals
    - “Bars” (granola, protein, fruit bars, etc.)
    - Dried/processed fruit
      - Jelly, jam, fruit spread, marmalade
      - Applesauce
      - “Fruit snacks”
      - Canned or highly processed fruit, “fruit cups”
    - Flavored yogurts, flavored oatmeal
    - Granola
    - Muffins
    - Jello/pudding
    - Sweet condiments (ketchup, teriyaki, BBQ, fruit-flavored vinaigrettes, honey mustard, etc.)
    - Syrup, honey
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QUALITY

- Examples of sugar-containing foods that are generally OK to give (because they are not sweet)
  - Plain Cherrios
  - Tomato Sauce
  - 100% whole grain bread/products
  - Sausage
  - Refried beans
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QUALITY

• Low nutrient-density foods are not recommended because they (1) tend not to be satisfying or satiating and (2) offer minimal nutritional benefit

• Examples:
  • White flour
  • Rice cakes
  • Cassava flour
  • Tapioca flour
  • Rice flour
  • Potato starch
  • Veggie straws
  • Pirate’s booty
  • Pretzels

• Look for 100% WHOLE grain products
Nutrition Guidance and Current Recommendations

**QUANTITY**

- Portion sizes are *highly* variable/individualized to specific needs
  - Based on growth, weight trends, activity level, body composition, genetics, etc.
  - General recommendations/handouts are *only a starting point*
  - **Most important: appropriate growth and weight gain**

- Generally, portions are significantly smaller when compared to general population
  - Due to low tone/lean mass, decreased energy needs

- Fruit is limited to twice per day and recommended to be served with a meal or with protein/fat

- Carbohydrates (whole grains, starchy vegetables, legumes, beans)
  - 1 fist (at each meal)
- Protein – 1 fist (at each meal)
- Non-starchy vegetables – 1 fist (at each meal)
- Nuts & Seeds – flat (*not piled up*) in the palm of hand (2x per day maximum)
- Fruit – flat (*not piled up*) in the palm of hand (2x per day maximum)
Nutrition Guidance and Current Recommendations

QUANTITY

• Guidance on snacks
  • Do NOT assume a snack(s) is needed
    • Do not offer food unless requested/showing signs of hunger
  • If your child is easily distracted, they may not be hungry
  • Lighter snacks are typically preferred
    • i.e. Non-starchy veggies (carrots, cucumbers, tomatoes, etc.)
  • No carb-only snacks
    • If serving fruit or whole grain/starch, serve with protein/fat (nuts, seeds, cheese, meat, eggs, olives, avocado, etc.)
  • Do NOT schedule snacks (or meals)
    • Keep snacks “as needed”
    • Allow flexibility in mealtimes; intentionally build “noise” into the schedule
Nutrition Guidance and Current Recommendations

BALANCE

1. **Protein**
   - Unstated

2. **Carbohydrates**
   - Starchy source of fiber ("complex carbs")
     - WHOLE grains, starchy veggies, beans/peas/legumes
   - Non-starchy source of fiber
     - Non-starchy veggies
   - Do not recommend carb-free (or low carb) meals
   - Do not recommend carb-heavy meals

3. **Fats**
   - Unsaturated/plant-based fats are preferred
   - Typically, fats are already included with the protein or added to the cooking process

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1. **Vegetables (NON-starchy)**
   - Carrots, Cucumbers, Green Beans, Broccoli, Cauliflower, Spinach, Lettuce, Tomatoes, Celery, Zucchini, Yellow Squash, Asparagus, Bell Peppers, Onions, Brussel Sprouts, Artichoke, Cabbage, Kale, Snap Peas, Okra, Collards, Bok Choy, Eggplant, Spaghetti Squash, Swiss Chard, Turnips

2. **Protein**
   - Beef, Pork, Chicken, Turkey, Salmon, Tuna, White Fish, Shrimp, Crab, Lobster, Scallops, Clams, Eggs, Greek Yogurt, Nuts, Seeds
   - Cottage Cheese, Duck, Soy, Tofu, Tempeh, Lamb, Goat

3. **Carbohydrate**
   - WHOLE grain Bread, WHOLE Grain Pasta, Quinoa, Brown Rice, Wild Rice, Barley, Oatmeal, Black Beans, Kidney Beans, Pinto Beans, White Beans, Lima Beans, Chickpeas (Garbanzo), Green Peas, Hummus, Black-Eyed Peas, Edamame, Lentils, Sweet Potato, Potato, Whole Grain Couscous, Farro, Millet, Pumpkin, Butternut/Acorn Squash, Popcorn, Buckwheat, Amaranth
Nutrition Guidance and Current Recommendations

Building a Balanced Meal

**BALANCE**

**PROTEIN**
- Fish (haddock, tilapia, cod, etc.)
- Shellfish
- Poultry - chicken, turkey, etc.
- Nonfat dairy - yogurt, milk
- Eggs whites
- Pork loin/chops
- Beans
- Peas
- Lentils
- Quinoa
- Edamame
- Beef
- Eggs whole/yolk
- Cheese
- Fatty Fish (salmon, herring, etc.)
- Bacon/Sausage
- Full-Fat Dairy - yogurt, milk, tofu
- Peanuts
- Tree Nuts - almonds, walnuts, pistachios, pecans, etc.
- Seeds - sunflower, flax, chia, hemp, pumpkin, etc.

**FAT**
- Oils
- Butter
- Mayonnaise
- Sour cream
- Cream cheese
- Avocado/Guacamole
- Olives
- Coconut meat

**FIBER**
- Whole Grains - oatmeal, brown rice, whole wheat bread, whole wheat pasta, kamut, barley, buckwheat, millet, rye, bulgur
- Fruit
- Non-starchy Vegetables - broccoli, spinach, green beans, cauliflower, carrots, etc.
- Starchy vegetables - sweet potatoes, butternut squash, etc.
- Chickpeas

**Example Breakfast**
- Omelet with bell peppers, tomatoes, onions. Whole wheat toast with peanut butter. Blueberries.

**Example Dinner**
- Chicken and brown rice served with asparagus, summer squash.
Nutrition Guidance and Current Recommendations

VARIETY

- Encourage a large variety of foods, textures, and flavors from a young age
- Do not stop offering disliked foods
- Do not rely too much on favorite foods
  - Do not let your child’s food preferences determine what is being served
- Continue to encourage new and different foods/flavors/textures etc.
- Nutritional variety:
  - Ensures the body is properly nourished
  - Reduces risk of nutritional deficiencies
  - Encourages diverse gut microbiome
  - Can reduce any selective (“picky”) eating
Nutrition Guidance and Current Recommendations

DRINKS!

- **Unsweetened** drinks only
  - Plain water
    - Ice-cold may work better
  - Milk or milk alternatives
  - Unsweet tea
  - Squeeze of *sour* fruit (lemon, lime) into water

- Beware of clever marketing: “no sugar added”, “no artificial sweeteners”, “sugar free”, “zero calorie”, etc.
  - These products are likely sweetened with alternative sweeteners (i.e. stevia, monk fruit, erythritol, sucralose, etc.)

- Build PLAIN water into *daily* routine
- Start EARLY, be consistent, do NOT offer alternative
# Nutrition Guidance and Current Recommendations

## MEAL AND SNACK IDEAS

|        | Monday                                                                 | Tuesday                                           | Wednesday                                              | Thursday                                               | Friday                                                 | Saturday                                              | Sunday                                                |
|--------|------------------------------------------------------------------------|----------------------------------------------------|--------------------------------------------------------|--------------------------------------------------------|--------------------------------------------------------|--------------------------------------------------------|
| **Breakfast** | Egg, Sausage, Raspberries, Whole grain toast                         | PLAIN Greek Yogurt Blueberries Cinnamon Oats      | Omelet w/ Tomatoes Peppers Whole grain toast Strawberries | PLAIN Greek yogurt Whole grain pancake with peanut butter Cherries | Egg Oatmeal Cinnamon Strawberries                      | PLAIN Greek Yogurt Blueberries Whole grain toast      | Peanut Butter on Whole grain pancake Sliced almonds Blackberries |
| **Lunch**   | Steak Cauliflower Carrots Sweet Potato                                | Chicken and Bean Chili Onions Tomatoes Yellow Squash | Salmon Broccoli Wild rice                               | Ground Turkey Tomatoes Cauliflower Black Beans Avocado | Salmon Spinach Quinoa                                 | Steak Asparagus Butternut Squash                      | Chicken Carrots Lentils                                 |
| **Snack**   | Apple Sunflower seeds                                                 | Pear Pecans                                       | Cherries Walnuts                                        | Apple Peanut butter                                    | Pear Sunflower seeds                                   | Orange Peanuts                                      | Apple Peanut butter                                  |
| **Dinner**  | Turkey Meatballs Broccoli Whole grain or Chickpea Pasta               | Salmon Brussel sprouts Brown Rice                  | Turkey Spinach Tomatoes Onions Black Beans Avocado       | Shrimp Green beans Quinoa                               | Fish Roasted carrots Whole grain couscous              | Baked chicken Green beans Sweet potato               | Pork Asparagus Peas                                    |
## CONVENIENCE - meals

- Quick, “no cook” meal examples

<table>
<thead>
<tr>
<th>Breakfast</th>
<th>Lunch</th>
<th>Dinner</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peanut butter, whole grain toast</td>
<td>Deli meat, Triscuits, carrots</td>
<td>Tuna, whole grain bread, cucumber</td>
</tr>
<tr>
<td>Cashews, Josephs Pita</td>
<td>Pre-boiled egg, popcorn, bell peppers</td>
<td>Sunflower seeds, canned black beans, carrots</td>
</tr>
<tr>
<td>Almonds, PLAIN/Original Cherrios</td>
<td>Plain Greek yogurt (+ fruit, cinnamon), edamame, tomatoes</td>
<td>Deli turkey, chickpeas/hummus, steam-in-bag green beans</td>
</tr>
</tbody>
</table>
CONVENIENCE – snacks

- Harvest Snap Pea Crisps
- Whole grain crackers/bread (+ protein/fat)
- Hippeas
- Crunchsters
- Bada Bean Bada Boom
- Cheese
- Meat
- Eggs
- Popcorn (+ protein/fat)
- Nuts/seeds
- Beef jerky/meat stick
- Yogurt (PLAIN)
- Dried chickpeas
- Peanut butter
- Peanut butter puffs
- Chicken/tuna salad
- PLAIN Cherrios (+ protein/fat)
- Fresh fruit (+ protein/fat)
- Veggies (carrots, tomatoes, cucumbers, peppers, celery, etc.)
- Dried veggies
- Hummus
- Dried seaweed
- Edamame or dried edamame

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MEAL “Schedules”

- A strict schedule can exacerbate behaviors when schedule is not/cannot be followed
- Maintaining flexibility in schedules is preferable and recommended
  - May need to intentionally build “noise” into daily routine
  - Base meals around activities, not the clock/time
  - Ensure your child knows that food will be provided, but is not dependent on a specific time
  - Avoid “feeding the clock”
SPECIAL OCCASIONS

- Sweet treats (cookies, cakes, cupcakes, brownies, ice cream, etc.) are OK on extremely special occasions (~5 times per year)
- Birthdays
- Special holidays
- Big vacations
- Examples of NOT special occasions:
  - “on weekends”
  - Friday nights
  - “Wednesdays”
  - After dinner (i.e., dessert)
  - “when we go out to eat”
  - Uncle’s birthday
Nutrition Guidance and Current Recommendations

FAMILY ROLES

- **TEAM effort**
  - Whole-family approach
  - Proper nutrition is beneficial for everyone in the family

- **Parents**
  - Do *not* cater to children’s likes and dislikes; your children are not “customers”, and you are not a “waiter/waitress”
  - Should treat all children similarly
  - Need to set aside own food issues
  - Do *not* let own food preferences determine what is being served
  - Understand your food anxiety will spill over to your children
  - Acknowledge potential risk of disordered eating in other family members

- **Grandparents**
  - Food is *not* a reward or treat (or punishment)
  - Do *not* show love through food (use other forms of rewards: toys, books, game, quality time, money, adventures/trips, etc.)

- **Siblings**
  - No sharing of food
  - Awareness of access to food
  - Maintain a healthy relationship with food
Nutrition Guidance and Current Recommendations

SUMMARY

- Quality – no sweet-tasting foods! Focus on Mediterranean Diet
- Quantity – small portions, especially fruit
- Balance – protein, complex carbs, non-starchy veggies (no carb-free meals & no carb-heavy meals)
- Variety – do not serve the same foods over and over. Continually offer new and non-preferred foods.
- Drinks – UNSWEETENED only
- Convenience – meals/snacks do not have to be fancy or complicated!
- Meal schedules – maintain flexibility; snacks not necessarily needed
- Special occasions foods ~ 5 times per year
- Family Roles – parents are in charge! Do not cater to too much to your child
Nutrition Guidance and Current Recommendations

Questions?
THANK YOU

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