Understanding Optimal Nutrition In Prader-Willi Syndrome

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Registered Dietitian
Lactation Consultant
In the office...

- Picky Eaters
- Poor Weight Gain
- Overweight & Obesity
- Food Allergies & Intolerances
- G-tube Fed
- Breastfeeding
- Prader-Willi Syndrome
The Road to PWS

• First patient---2004

• **HOW?**
  • Early pediatric growth
  • Lactation/Formulas
  • Healthy eating recommendations
  • Psychology of kids’ eating and parents feeding
  • Pediatric/Adult weight management
  • Metabolic Carts and RQ measurements
  • Continuous drive to learn
Two Objectives for Today

• **HOW** to feed

• **WHAT** to feed
Good Advice
How To Feed
How To Feed

Structure

- Essential
Structure: MENUS/RULES

- Separate meals and snacks by at least 2 ½ hours
- Post menus and rules
- Family meals
- No distractions at meals *(No “i-anything” at the table)*
Principles of Food Security in PWS

• **No doubt** when meals will occur and what foods will be served

• **No hope** of getting anything different from what is planned

• **No disappointment** related to false expectations
Principles of Food Security in PWS

- **No doubt** when meals will occur and what foods will be served—**SCHEDULE/MENU**
- **No hope** of getting anything different from what is planned—**SCHEDULE/MENU**
- **No disappointment** related to false expectations—**SCHEDULE/MENU**
Ways to Achieve Food Security

1. Secure food accessibility across all settings
2. Avoid spontaneity related to food
3. Supervise food exposure
4. Post the meal schedule
5. Post the menus
6. Try to avoid places and social situations with excess food

Janice L. Foster, MD and Linda M. Gourash, MD  Pittsburgh Partnership
Physical Activity

• Parks

• Sports

• Family Participation
WHAT to feed
Nutrition 101

- Calories
- Carbohydrate
- Protein
- Fat
- What to eat?
Calories Required

The Food

Your Body
Calories

A unit of energy

Carbohydrates, Protein & Fat provide this energy
Carbohydrates
Protein
Fat
Foods Can Provide C, P and F

Carbohydrate (C)
- Yogurt and Milk:  C, P, F

Protein (P)
- Eggs: P, F
- Nuts:  P, F
- Beans:  C, P, F

Fat (F)
- Cottage Cheese:  P, F
What Is Optimal Nutrition?
Food Guide Pyramid

- Exercise:
  - Adults should be physically active for at least 30 minutes most days of the week, children for 60 minutes.
  - Sixty to 90 minutes of daily physical activity may be needed to prevent weight gain or sustain weight loss.

- Old food pyramid:
  - Emphasized limits on fats, oils and sweets, which were represented as the tip of the pyramid.
  - Presented food groups as a hierarchy, with grains as the base of a healthy diet, and each group having a suggested number of servings.

- New food pyramid:
  - Grains
  - Vegetables
  - Fruits
  - Milk
  - Meat and beans

- Recommendation:
  - Half of all grains consumed should be whole grains.
  - Vary the types of vegetables you eat.
  - Eat a variety of fruits. Go easy on juices.
  - Eat low-fat or fat-free dairy products.
  - Eat lean cuts, seafood and beans. Avoid frying.

- Daily amount based on a 2,000 calorie diet:
  - Grains: 6 oz.
  - Vegetables: 2.5 cups
  - Fruits: 2 cups
  - Milk: 3 cups
  - Meat and beans: 5.5 oz.

Recommended nutrient intakes at 12-calorie levels can be found on mypyramid.gov.
Prader-Willi Food Pyramid

- Fats & Sweets: Use Sparingly
- Meat, Poultry, Fish, Dry Beans, Eggs: 1-2 servings daily, 2 oz. each
- Bread, Cereal, Rice & Pasta Group: 3-5 servings daily
- Milk, Yogurt, & cheese Group: 2 servings daily
- Fruit Group: 4 servings daily
- Vegetable Group: 6-8 servings daily
My Plate

ChooseMyPlate.gov
Proposed Prader-Willi Plate

- Grains
- Protein
- Fruits
- Vegetables

Dairy
Optimal Diet

2012
“A reduced energy intake, well balanced diet improves weight control in children with Prader-Willi Syndrome”
(J.L. Miller, C.H. Lynn, J. Shuster, D.J. Driscoll)

• Children 2-10
• 30% fat, 45% carbohydrate, 25% protein, 20 grams of fiber
• Energy restricted diet
Results

Balanced macronutrient at 30% fat, 45% carbohydrate and 25% protein (20 grams of fiber) IMPROVES weight and body composition in children with PWS compared to a simple energy restricted diet.
## Calorie Percentages

<table>
<thead>
<tr>
<th>USDA</th>
<th>PWS</th>
</tr>
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<tbody>
<tr>
<td><strong>CARBS:</strong> 50-70%</td>
<td><strong>CARBS:</strong> 45%</td>
</tr>
<tr>
<td><strong>PRO:</strong> 15-20%</td>
<td><strong>PRO:</strong> 25%</td>
</tr>
<tr>
<td><strong>FAT:</strong> 30-35%</td>
<td><strong>FAT:</strong> 30%</td>
</tr>
<tr>
<td></td>
<td><strong>FIBER:</strong> 20 grams per day</td>
</tr>
</tbody>
</table>

GOOD Carbs vs. BAD Carbs

“GOOD”
- Fruits/Vegetables
- Whole grains (>3 grams per serving)
- Beans, Peas, Legumes
- Brown rice
- Quinoa

“BAD”
- Candy
- Cakes
- Cookies
- Juice
- MANY MANY muffins
- Ice Cream
- Donuts
- Low fiber crackers
- White bread, pasta, rice
- “Snacky snacks”
Before/AFTER

**Before**
- Frosted Flakes, Milk, Grapes
- Tuna Sandwich, Pretzels, Apple
- Spaghetti, Salad and Garlic Bread
- Chicken Noodle Soup with Bread sticks, Salad

**After**
- Oatmeal, Pecans and Raisins
- Tuna Salad on Cucumber Slices, apple
- Eggplant lasagna (roasted eggplant as the noodles), berries
- Chicken, Asparagus, Salad
Before/AFTER

**Before**
- Tacos with shell
- Spaghetti and sauce
- Bagel and cream cheese
- Buttermilk Pancakes, Eggs, Bacon

**After**
- Taco salad (no shell)
- Spaghetti SQUASH and sauce
- Yogurt and granola
- Whole Wheat waffle (>3 grams per waffle) eggs, bacon
How Many Calories?

• For children and adolescents (varies):
  – 10-11 calories per centimeter to maintain growth velocity
  – 8-9 calories per centimeter for slow weight loss or support linear growth

• Adults vary:
  – 1,000-1,200 per calories day
  – About 60% of a typical person’s diet

ADA Pediatric Nutrition Assessment, 2008
Flip the Box
FOOD LABELS

- Serving Size
- Serving Per Container
- Calories
- Fat
- Sugar
- Fiber

**Nutrition Facts**

**Serving Size**: ½ cup (114g)
**Servings Per Container**: 4

<table>
<thead>
<tr>
<th>Amount Per Serving</th>
<th>Calories</th>
<th>90</th>
<th>Calories from Fat</th>
<th>30</th>
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<tbody>
<tr>
<td><strong>% Daily Value</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total Fat</strong></td>
<td>3g</td>
<td>5%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Saturated Fat</td>
<td>0g</td>
<td>0%</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Cholesterol</strong></td>
<td>0mg</td>
<td>0%</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Sodium</strong></td>
<td>300mg</td>
<td>13%</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total Carbohydrate</strong></td>
<td>13g</td>
<td>4%</td>
<td></td>
<td></td>
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<tr>
<td>Dietary Fiber</td>
<td>3g</td>
<td>12%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sugars</td>
<td>3g</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Protein</strong></td>
<td>3g</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

- Vitamin A 80%
- Vitamin C 60%
- Calcium 4%
- Iron 4%

*Percent Daily Values are based on a 2,000 calorie diet. Your daily values may be higher or lower depending on your caloric needs:

<table>
<thead>
<tr>
<th>Calories:</th>
<th>2,000</th>
<th>2,500</th>
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<tbody>
<tr>
<td>Total Fat</td>
<td>Less than</td>
<td>65g</td>
</tr>
<tr>
<td>Sat Fat</td>
<td>Less than</td>
<td>20g</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>Less than</td>
<td>300mg</td>
</tr>
<tr>
<td>Sodium</td>
<td>Less than</td>
<td>2,400mg</td>
</tr>
<tr>
<td>Total Carbohydrate</td>
<td>300g</td>
<td>375g</td>
</tr>
<tr>
<td>Dietary Fiber</td>
<td>25g</td>
<td>30g</td>
</tr>
</tbody>
</table>

Calories per gram:
- Fat 9
- Carbohydrate 4
- Protein 4
Look at **Fiber** on a Food Label

### Nutrition Facts

Serving Size: 1/2 cup (114g)
Serving Per Container: 4

<table>
<thead>
<tr>
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<tbody>
<tr>
<td><strong>Calories</strong></td>
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<td>Calories from Fat: 30</td>
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<tr>
<td><strong>Total Fat</strong></td>
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<td>5%</td>
</tr>
<tr>
<td>Saturated Fat</td>
<td>0g</td>
<td></td>
</tr>
<tr>
<td>Cholesterol</td>
<td>0mg</td>
<td>0%</td>
</tr>
<tr>
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<td>4%</td>
</tr>
<tr>
<td>Dietary Fiber</td>
<td>3g</td>
<td>12%</td>
</tr>
<tr>
<td>Sugars</td>
<td>3g</td>
<td></td>
</tr>
<tr>
<td><strong>Protein</strong></td>
<td>3g</td>
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- Vitamin A: 80%
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- Calcium: 4%
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<th>2,500</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Fat</td>
<td>Less than 65g</td>
<td>80g</td>
</tr>
<tr>
<td>Saturated Fat</td>
<td>Less than 20g</td>
<td>25g</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>Less than 300mg</td>
<td>300mg</td>
</tr>
<tr>
<td>Sodium</td>
<td>Less than 2,400mg</td>
<td>2,400mg</td>
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<td>375g</td>
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<tr>
<td>Dietary Fiber</td>
<td>25g</td>
<td>90g</td>
</tr>
</tbody>
</table>

Calories per gram:
- Fat: 9
- Carbohydrate: 4
- Protein: 4
INGREDIENTS: ENRICHED BLEACHED WHEAT FLOUR [FLOUR, FERROUS SULFATE, “B” VITAMINS (NIACIN, THIAMINE MONONITRATE (B1), RIBOFLAVIN (B2), FOLIC ACID)], SUGAR, CORN SYRUP, WATER, HIGH FRUCTOSE CORN SYRUP, PARTIALLY HYDROGENATED VEGETABLE SHORTENING (CONTAINS ONE OR MORE OF: SOYBEAN, CANOLA OR PALM OIL), DEXTROSE, WHOLE EGGS. CONTAINS 2% OR LESS OF: MODIFIED CORNSTARCH, CELLULOSE GUM, WHEY, LEAVENINGS (SODIUM ACID PYROPHOSPHATE, BAKING SODA, MONOCALCIUM PHOSPHATE), SALT, CORNSTARCH, CORN FLOUR, CORN DEXTROINS, MONO AND DIGLYCERIDES, POLYSORBATE 60, SOY LECITHIN, NATURAL AND ARTIFICIAL FLAVORS, SOY PROTEIN ISOLATE, SODIUM STEAROYL LACTYLATE, SODIUM AND CALCIUM CASEINATE, CALCIUM SULFATE, SORBIC ACID (TO RETAIN FRESHNESS), COLOR ADDED (YELLOW 5, RED 40). MAY CONTAIN PEANUTS OR TRACES OF PEANUTS.
What is high fructose corn syrup?

• Common sweetener derived from corn
  – Process corn with enzymes/acids

• Cheaper than regular sugar

• Found in: cereals, yogurts, snack foods, soda, soups, condiments, breads
Percent Daily Values are based on a 2,000 calorie diet.

CARBONATED WATER, HIGH FRUCTOSE CORN SYRUP, CARAMEL COLOR, PHOSPHORIC ACID, NATURAL FLAVORS, CAFFEINE.

CAFFEINE CONTENT: 34 mg/12 fl oz.

2007-5517 12 FL OZ
All of us need to limit how much sugar we eat...whether it’s high fructose corn syrup or not.
Sweeteners
Artificial Sweeteners

- Aspartame
- Sucralose
- Sugar alcohols
- Stevia

- Natural Food Markets may be best choice

- *Individual decision*
Should We Use Them?

- Personal choice
- Artificial means not real
- Intensify cravings?
Constipation

• To poop...one needs:

• **SOLUBLE FIBER:** Fruits/Vegetables
• **INSOLUBLE FIBER:** Whole Grains
• **FAT:** Oils, Butter
• **LIQUID:** Flavored water, Juice (no juice!), Tea (fruits and vegetables hydrate)

• Physician assistance
Supplementation
Supplements

• Fish Oil
• Carnitine
• Coenzyme Q10
• Creatine
• Multivitamin
• Iron
• Calcium
• Vitamin D
DISCUSSION WITH PHYSICIAN

• Carnitine—cellular metabolism, hypotonia, alertness

• Coenzyme Q10—energy levels, muscle function, metabolism
Some Supplements Seen in PWS

- **Multivitamin:** “Insurance plan”
- **Omega 3’s:** inflammation, blood clotting, cell membranes, brain development
- **Calcium:** bone health (if diet is low in dairy)
- **Vitamin D:** “sunshine vitamin”, bone health
- **Vitamin B12:** red blood cell formation, neurologic function and DNA synthesis
- **Probiotics:** promote gut and immune health

* Harvard School of Public Health, Web MD*
Best **Food** Sources of Supplements

- **Carnitine**: beef, milk
- **CoQ10**: salmon, tuna, liver, whole grains
- **Omega 3’s**: fish, walnuts, spinach
- **Calcium**: dairy, spinach, kale, beans, cereals
- **Vitamin D**: salmon, egg yolk, fortified milk and cereal
- **Vitamin B12**: meat, eggs, dairy
- **Probiotics**: yogurt, kefir
What You Need To Know

- Fresh food & better balance of CHO, PRO, FAT
- Shop the perimeter of the grocery store
- Visit Farmer’s Markets
- Cook
- Read food labels closely
- No juice, soda, sport drinks, etc. (small splash in water)
- Limit sugar intake
- Watch/learn about artificial sweeteners
- Supplements per physician recommendation
- Structure meals/snacks
- Intensive physical activity
Follow Me...

FeedPhilosophy

Feeding Philosophies
Private Consultations

- **Call/Email me:** melanie@feedingphilosophies.com
- **Insurance:** Call ahead about coverage
- **Referral:** Best to get physician to write referral
Thank you!

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