

# Understanding Optimal Nutrition In Prader-Willi Syndrome



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[www.feedingphilosophies.com](http://www.feedingphilosophies.com)

# 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



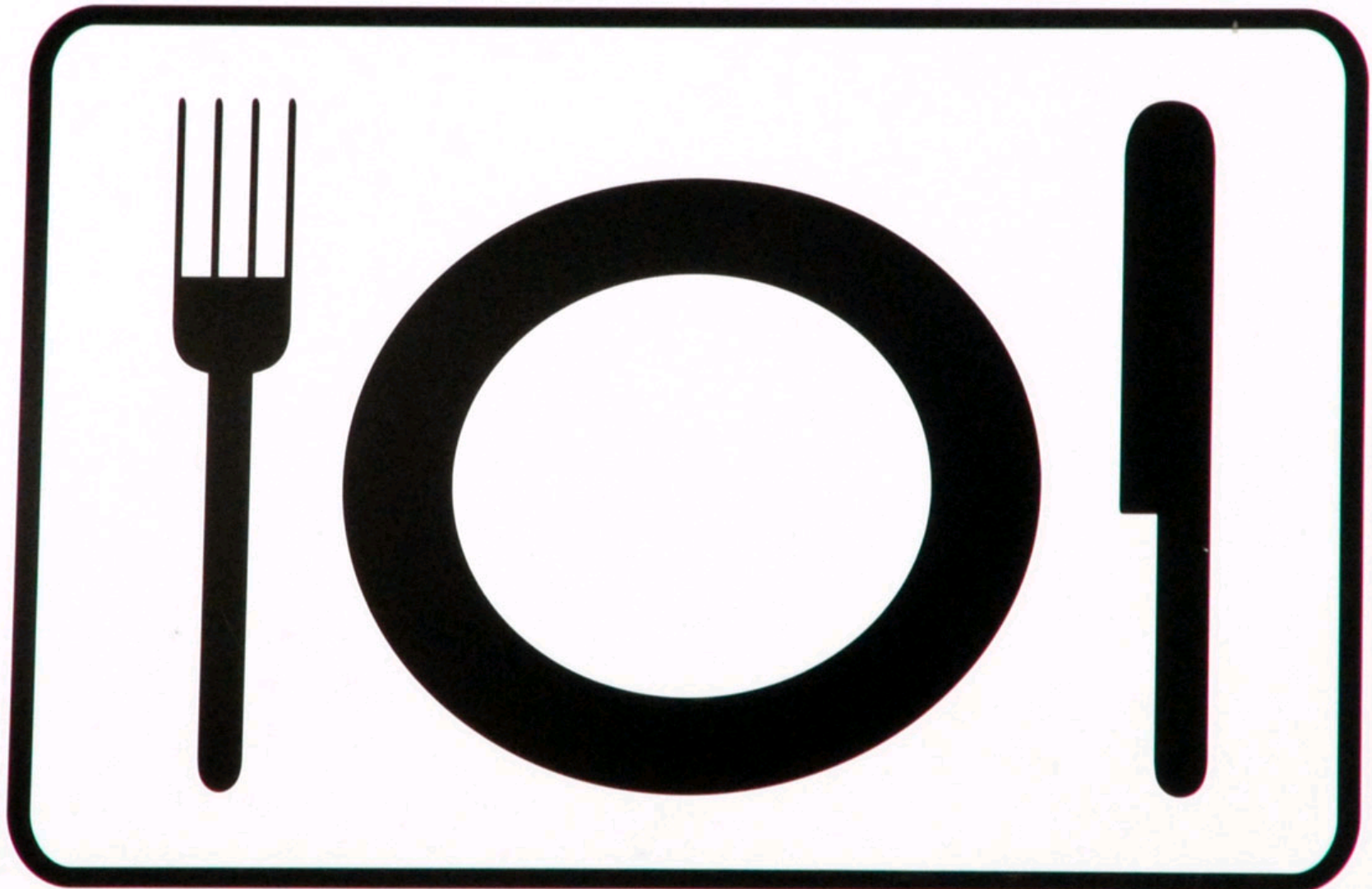
# Two Objectives for Today

- 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-  
anythings” at the  
table)



Structure Supports →

Food Security

# 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

# Physical Activity

- Parks
- Sports
- Family Participation

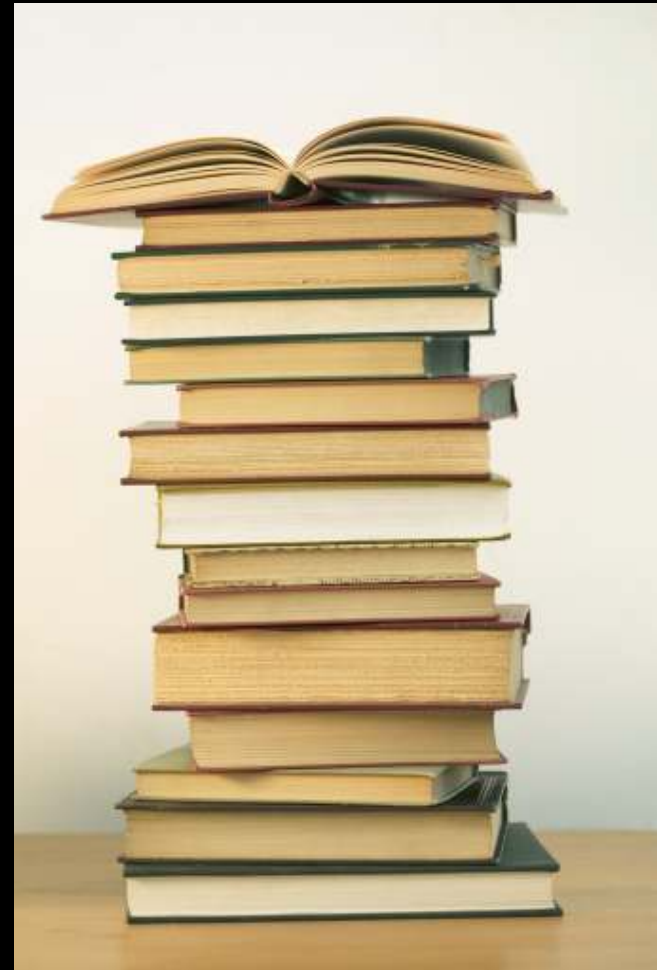


# WHAT to feed



# Nutrition 101

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





$$\sqrt{\frac{x_i - y^2}{i + \frac{1}{y_i^k} - k}}$$

# 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)

Protein (P)

Fat (F)

- Eggs: P, F
- Yogurt and Milk: C, P, F
- Nuts: P, F
- Beans: C, P, F
- Cottage Cheese: P, F

# What Is Optimal Nutrition?

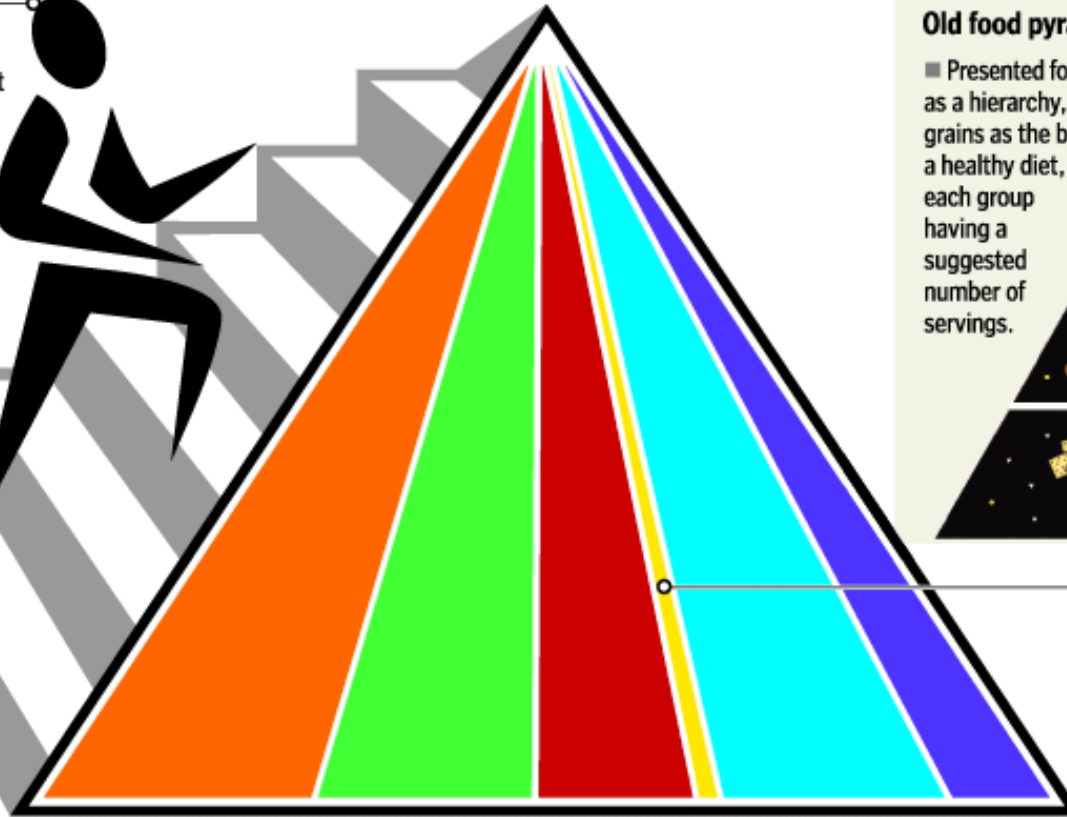




# Food Guide Pyramid

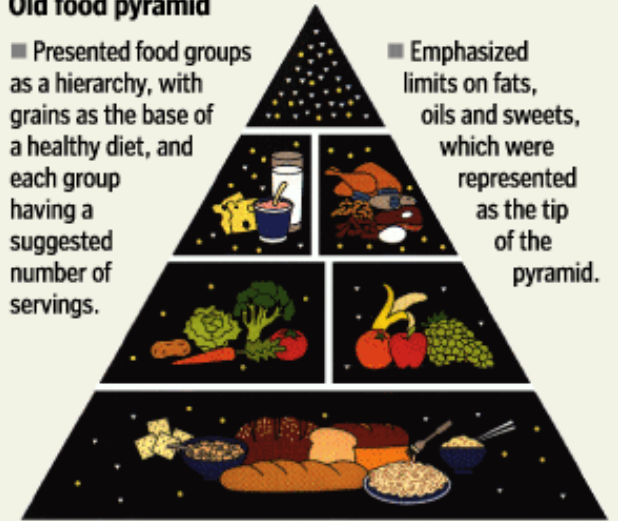
## Exercise

- 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

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



## Oils

- Most fat should be from fish, nuts and vegetable oils.
- Limit solid fats, such as butter, margarine or lard.
- Keep consumption of saturated fats, trans fats and sodium low.
- Choose foods low in added sugar.

## CATEGORY

**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

6 oz.

2.5 cups

2 cups

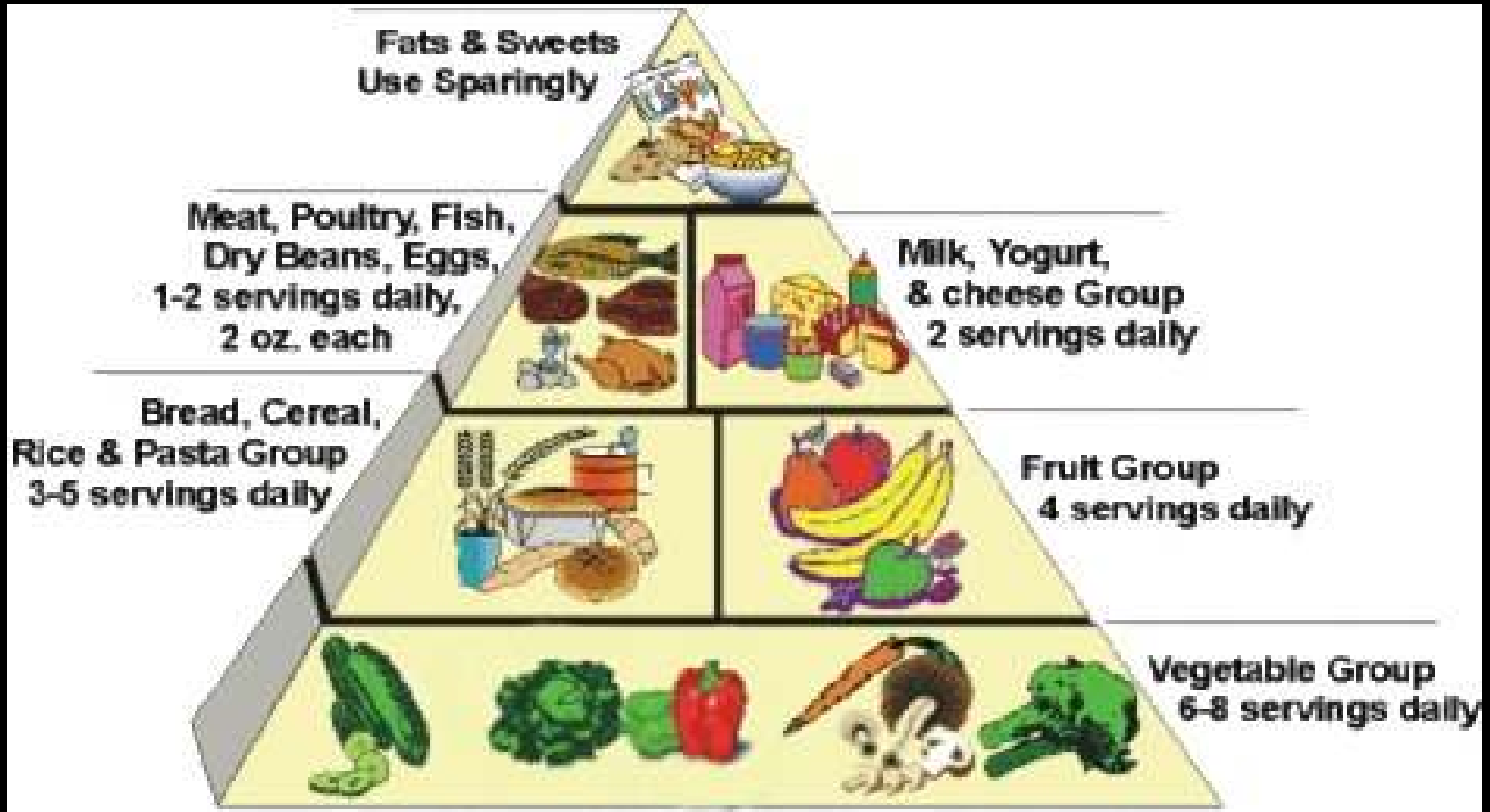
3 cups

5.5 oz.

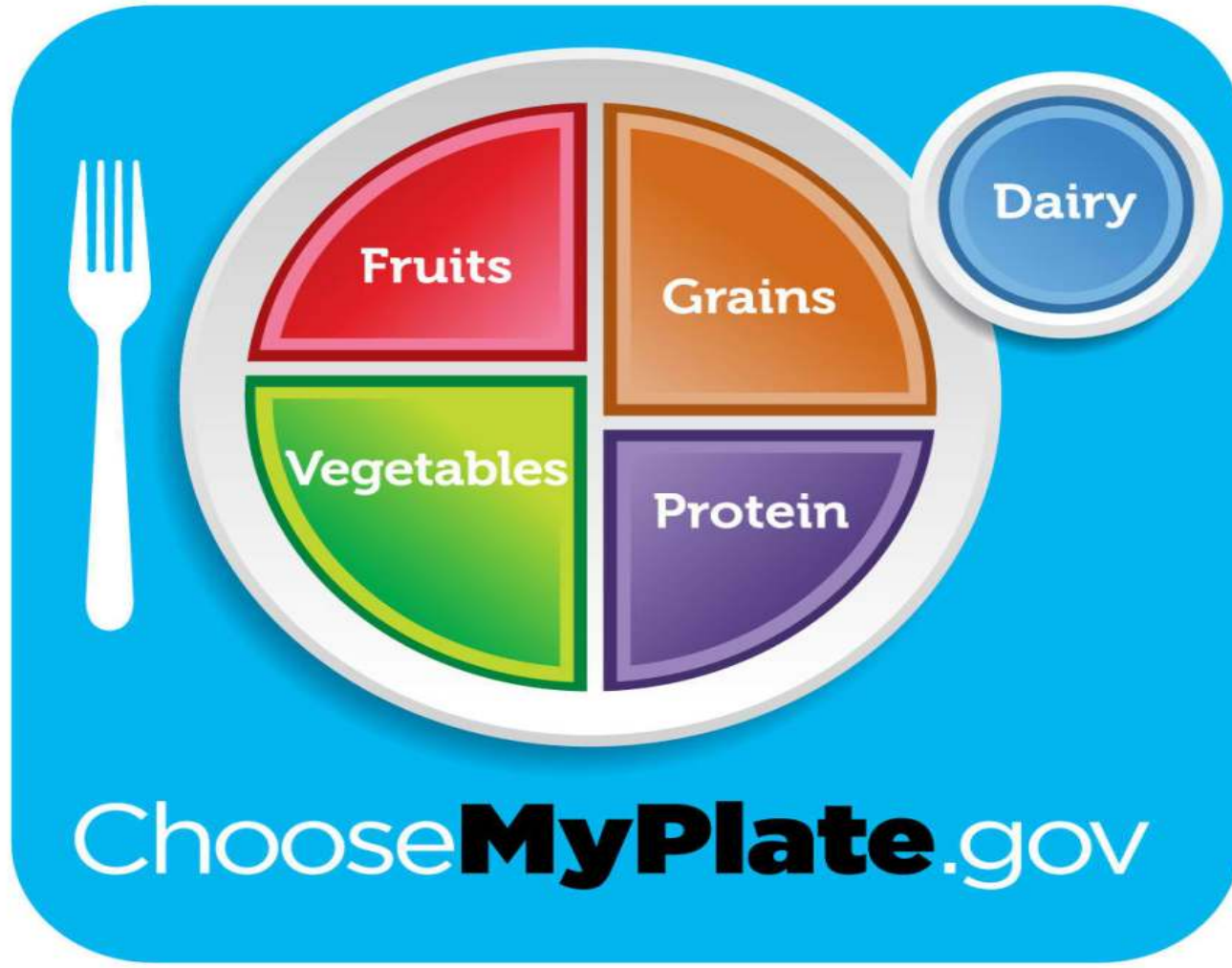
Based on a 2,000 calorie diet.

Recommended nutrient intakes at 12-calorie levels can be found on [mypyramid.gov](http://mypyramid.gov).

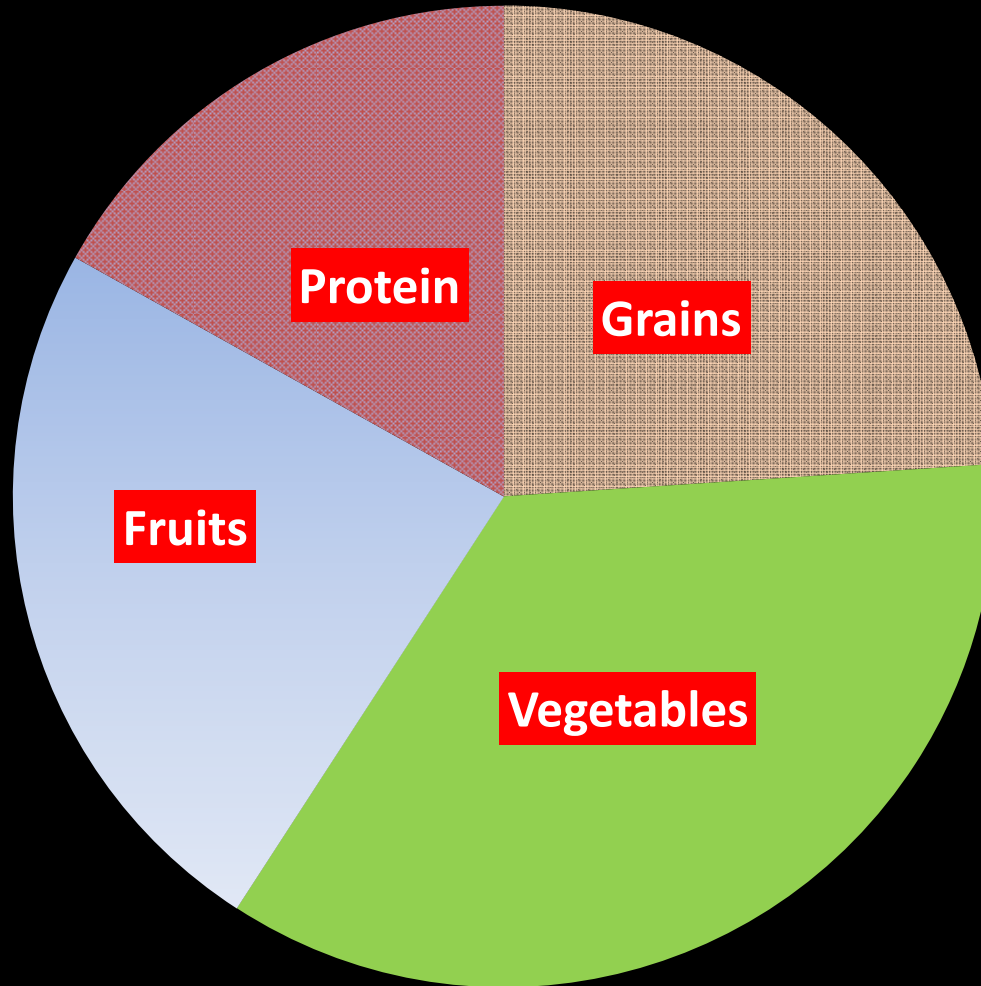
# Prader-Willi Food Pyramid



# My Plate



# Proposed Prader-Willi Plate



# 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

## USDA

CARBS: 50-70%  
PRO: 15-20%  
FAT: 30-35%

## PWS

CARBS: 45%  
PRO: 25%  
FAT: 30%  
FIBER: 20 grams  
per day

\*Miller, et al. A reduced-energy intake, well balanced diet improves weight control in children with Prader-Willi syndrome, J Hum Nutr Diet, 2012

# 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

# Flip the Box





# Look at Fiber on a Food Label

<b>Nutrition Facts</b>	
Serving Size ½ cup (114g) Servings Per Container 4	
<b>Amount Per Serving</b>	
<b>Calories</b> 90	Calories from Fat 30
<b>% Daily Value*</b>	
<b>Total Fat</b> 3g	<b>5%</b>
Saturated Fat 0g	<b>0%</b>
<b>Cholesterol</b> 0mg	<b>0%</b>
<b>Sodium</b> 300mg	<b>13%</b>
<b>Total Carbohydrate</b> 13g	<b>4%</b>
Dietary Fiber 3g	<b>12%</b>
Sugars 3g	
<b>Protein</b> 3g	
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:	
	Calories: 2,000 2,500
Total Fat	Less than 65g 80g
Sat Fat	Less than 20g 25g
Cholesterol	Less than 300mg 300mg
Sodium	Less than 2,400mg 2,400mg
Total Carbohydrate	300g 375g
Dietary Fiber	25g 30g
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 DEXTRINS, 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

*Bull*

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2007-5517 12 FL OZ

# The bottom line

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

# Some Supplements in PWS

## DISCUSSION WITH PHYSICIAN

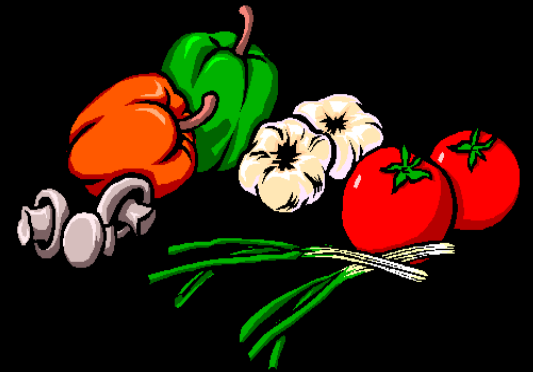
- Carnitine—cellular metabolism, hypotonia, alertness
- Coenzyme Q10—energy levels, muscle function, metabolism

# 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

# 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...

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# Private Consultations

- Call/Email me:  
[melanie@feedingphilosophies.com](mailto:melanie@feedingphilosophies.com)
- Insurance: Call ahead about coverage
- Referral: Best to get physician to write referral



# Thank you!



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