

Prader-Willi Syndrome

1. Purpose

a. Nutrition Indicators

- Decreased metabolic/caloric needs
- Overweight, obesity
- Excessive overeating and preoccupation with food

b. Criteria to Assign the Diet

- Genetic testing to indicate genetic deletions on chromosome 15
- Weight gain
- Overweight, obesity associated health conditions

c. Rationale for Diet

- Low calorie diet needed to compensate for the body's decreased caloric needs
- Caloric restriction is necessary to prevent additional weight gain and overweight/obesity associated health conditions

2. Population

a. Overview

- 1 in 12,000 to 15,000 males and females of all races and ethnicities is diagnosed with PWS
- There is no cure for PWS
- The only known treatment includes caloric restriction, a strict physical activity regimen, and possible treatment with growth hormone

b. Disease Process and Diagnostic Criteria

Disease Progression and Diagnostic Criteria for Prader-Willi Syndrome	
Birth - 2 Years	1. Hypotonia (low muscle tone/function) with poor suck
2 Years – 6 Years	1. Hypotonia with history of poor suck 2. Global developmental delay
6 Years – 12 Years	1. Hypotonia with history of poor suck 2. Global developmental delay 3. Excessive overeating (hyperphagia, preoccupation with food); central obesity if not controlled
13 Years – Adulthood	1. Cognitive disabilities, mild mental retardation 2. Excessive overeating (hyperphagia, preoccupation with food); central obesity if not controlled 3. Behavior problems; hypothalamic hypogonadism

	% Affected
Major criteria	
Neonatal hypotonia	88
Feeding problems in infancy	79
Excessive weight gain	67
Facial features	88
Hypogonadism	51
Developmental delay	99

Hyperphagia	84
Minor criteria	
Decreased fetal activity	62
Behavior problems	87
Sleep disturbance/sleep apnea	76
Short stature	63
Hypopigmentation	73
Small hands and/or feet	88
Narrow hands/straight ulnar borders	82
Eye abnormalities	68
Thick viscous saliva	89
Articulation defects	80
Skin-picking	83

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Reference:

Meral Gunay-Aygun, Stuart Schwartz, Shauna Heeger, Mary Ann O'Riordan and Suzanne B. Cassidy
 Pediatrics 2001;108;92-DOI: 10.1542/peds.108.5.e92
 Cassidy SB. Prader-Willi syndrome. J [Med Genet.](#) 1997;34:917-923

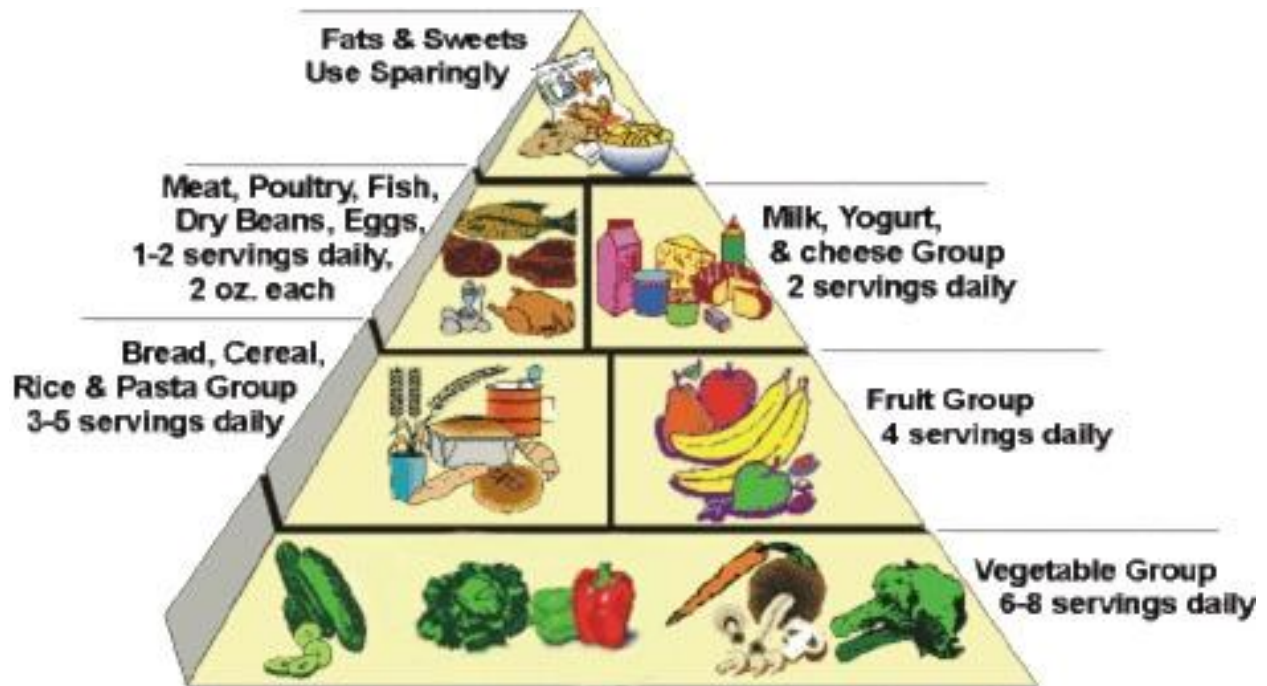
d. Biochemical and Nutrient Needs

- Decreased caloric needs
- Vitamin and calcium supplementation are necessary to provide additional nutrients that the patient may not receive on a reduced calorie diet

3. General Guidelines

a. Nutrition Rx

- Typically 1,000-1,200 kcal/day for adults
- PWS Food Pyramid
 - Decreased servings of meat, poultry, fish, dry beans, eggs→results in decreased fat consumption (1-2 servings, 2oz each)
 - Decreased bread, cereal, rice, and pasta group (3-5 servings)
 - Decreased servings of dairy (2/day)
 - Increased consumption of fruit and vegetables to provide more bulk in the diet with a lower caloric intake (4 servings fruit/day; 6-8 servings vegetables/day)
 - Fats and sweets should only be consumed once per month on an 800 kcal diet or once weekly on a 1200 kcal diet because they provide added calories and little nutrition
- Patients should engage in at least 30 minutes of physical activity daily
- Availability of food should be limited
- It may be necessary to put locks on the refrigerators and cupboards, especially to limit the intake of children
- Strict supervision is necessary



b. Adequacy of Nutrition Rx

- Provides sufficient calories to prevent weight gain
- Sufficient nutrients are obtained through increased intake of produce and vitamin/calcium supplementation

c. Goals

- Prevent additional weight gain
- Prevent the development of weight-related complications

d. Does it Meet DRI

- With supplementation, the reduced calorie diet provides sufficient nutrients

4. Education Material

a. Nutrition Therapy

- Nutrition consultation
- Physical training and exercise
- Reduced calorie diet using above food pyramid

b. Ideas for Compliance

- Increased consumption of high volume, low calorie foods
- Limited consumption of empty calories and beverages with calories
- Restriction of foods brought into the house/careful grocery shopping

5. Sample Menu

a. Foods Recommended

- Low-fat, low-caloric, nutrient-rich foods
- Produce, whole grains, fat-free dairy, lean meat

b. Foods to Avoid

- High caloric items
- High fat foods, fried foods

- Sweets and desserts
- c. Example of a meal plan
- Breakfast:* ½ c oatmeal prepared with ½ c milk, 1 packet Splenda, and cinnamon; 1 small banana
- Snack:* 1 medium apple or orange
- Lunch:* Black bean enchiladas (2 small corn tortillas, 2 oz black beans, ½ c red pepper strips, 1 oz low-fat Mexican cheese, topped with 2 T salsa); ½ c baby carrots dipped in salsa
- Snack:* 6 whole grain crackers, 1 oz sliced turkey, ½ c cherry tomatoes sliced in half, mustard (if desired)
- Dinner:* Vegetarian stir-fry (1 oz tofu, ½ c broccoli, ½ c shredded carrots, ¼ c onions, ¼ c mushrooms, ½ c pineapple canned in own juice) served on 1 c. brown rice with a low-sodium stir-fry sauce
- Snack:* 1 c. strawberries with 1 T fat-free, sugar-free chocolate syrup and 1 c fat-free milk

6. Websites

- a. Organizations with Websites
- Prader-Willi Syndrome Association: <http://www.pwsausa.org/>
 - Prader-Willi Syndrome Association of Ohio: <http://www.pwsaohio.org/>
- b. Government Websites
- MedLine Plus:
<http://www.nlm.nih.gov/medlineplus/praderwillisyndrome.html>

7. References

- a. Journal articles
- <http://www.eatright.org/WorkArea/DownloadAsset.aspx?id=8424>
 - <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2422866/>