Nutrition in PWS

Dr. med. Constanze Lämmer
Pediatric Endocrinologist, Diabetologist and Medical Nutrition Specialist
Childrens Hospital St. Bernward Krankenhaus Hildesheim, Germany
Overview

• Reduced energy intake
• Special metabolic situation of the individuals with PWS
• Keystones of a healthy balance diet
• Food pyramid
• Role of Insulin
• Everyday nutrition - Sunday nutrition
• Psychological aspects of nutritional management
## What define the energy expenditure

<table>
<thead>
<tr>
<th>Factor</th>
<th>Range</th>
<th>Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adaptive thermo genesis</td>
<td>10-30%</td>
<td>Nutrition intake and composition</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Surrounding temperature</td>
</tr>
<tr>
<td>Body activity</td>
<td>20-40%</td>
<td>Normal activities</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sports</td>
</tr>
<tr>
<td>Basal metabolic rate</td>
<td>50-70%</td>
<td>Genetics</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Basic muscle tone</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Age</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Gender</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hormones</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Non fat body mass</td>
</tr>
</tbody>
</table>
Energy intake and Metabolism in PWS

- Reduce metabolic rate due to muscular hypotonia and less muscle mass
- Greater fat storage, gain fat readily
- Reduced active energy expenditure due to reduced movement and lack of exercise
- 60-80% daily recommended intake, of peers

- Disturbance in the glucose metabolism
- Insulin resistance
- Reduced thermo genetic effect
- Disturbance of the oxidative energy production
- Less Growth hormone
Recommendations for Energy intake for individuals with PWS

- Children with PWS: start with 2/3 of the caloric intake of peers because of the lower energy expenditure
- Calculate the daily caloric intake with

<table>
<thead>
<tr>
<th>age</th>
<th>Boys with PWS (kcal)</th>
<th>Girls with PWS (kcal)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-3 y</td>
<td>800</td>
<td>720</td>
</tr>
<tr>
<td>4-6 y</td>
<td>950</td>
<td>850</td>
</tr>
<tr>
<td>7-9 y</td>
<td>1100</td>
<td>1000</td>
</tr>
<tr>
<td>10-12 y</td>
<td>1250</td>
<td>1100</td>
</tr>
<tr>
<td>13-15 y</td>
<td>1500</td>
<td>1250</td>
</tr>
</tbody>
</table>

>15 years and for adults:
- To maintain weight: 8-9 kcal/cm body height/day
- To reduce weight 7 kcal/cm body height/day
- To gain weight 10-11 kcal/cm body height/day
Nutrition for individuals with PWS

• Balanced diet
  – Energy relations:
  – 45-50% carbohydrates, 25-20% proteins, 30% fat
  – Restricted use of sugar and high-glycemic-index carbohydrates
  – Prefer unsaturated fatty acids (oil)
  – Enough free-caloric drinks
• Add vitamins and minerals if necessary: usually only
• Vit D has to be added
Energy of Food Groups

- Vegetables: 30 kcal/100g
- Fruits: 50 kcal/100g
- Potatoes: 70 kcal/100g
- Rice cooked: 110 kcal/100g
- Noodles cooked: 130 kcal/100g
- Lean meat: 130 kcal/100g
- Fat meat sausages: 300 kcal/100g
- Cookies: 400 kcal/100g
- Chocolate: 530 kcal/100g
Food pyramid
Vegetables 3 x per day

- Low caloric density
- Less than 30 kcal/100 g (expect corn, kidney beans)
- Rich on vitamins and minerals
- Rare vegetable as a snack
- Gives nice colors to meals
- Can be given as finger food
- Training for chewing muscles
- Have to be offered positive
- In case of swallowing problems prefer cooked or blanched vegetables
Vegetable helps to reduce energy intake – make they look good
Fruits – 2 x per day

- Have vitamins and minerals like vegetables
- Main energy source: carbohydrates mostly with less glycemic index
- Free of fat
- Suitable as a snack
- Avoid bananas and grapes because of the higher sugar content
- Prefer apples, cherries, berries, orange, kiwis, grapefruits, pears
Products from Rice, Grain, Pasta, Potatoes

- Main component: starch, beside proteins, Vitamins, minerals
- Use whole grain products because they are rich on fibre
- Less fat, no cholesterol
- 3 slice of bread (handsize) and 1 portion of rize potatos/pasta (2 handsize)
- Instead of one slice of bread:
  - 30 -50 g cereals = individual portion
  - Please weigh the cereals!
Choose cereals carefully: avoid sugar

<table>
<thead>
<tr>
<th>In 100g</th>
<th>Fruit Loops</th>
<th>Smacks</th>
<th>Cornflakes</th>
<th>Toppas</th>
<th>Oat müsli</th>
</tr>
</thead>
<tbody>
<tr>
<td>carbohydrate g</td>
<td>80</td>
<td>84</td>
<td>84</td>
<td>72</td>
<td>50</td>
</tr>
<tr>
<td>Sugar g</td>
<td>25</td>
<td>43</td>
<td>8</td>
<td>17</td>
<td>3,9</td>
</tr>
</tbody>
</table>
Insuline has a key role in metabolism

- Insulin has the task to transport glucose in the muscles
- Fast carbohydrates need a lot of insulin in a short time
- Stops Gluconeogenesis in liver
- Builds up fat reserves
- Stops fat reduction
- High insulin levels in the blood will be followed by insulin resistance and later diabetes
- Complex carbohydrates prevent hyperinsulinemia and make weight reduction possible
- Low-glycemic-index-carbohydrates prevent hyperinsulinemia and make weight reduction possible
Milk and milk products

- 3 times daily
- Important source for Calcium
- Energy from protein, milk fat and milk sugar

- Cheese
  - 70% Fat i.d.  130 kcal/30 g
  - 40% Fat i.d.  85 kcal/30 g
  - 10% Fat i.d.  35 kcal/30 g

- Cheese with 30% Fat i.d. and 1.5% milk are ok
Meat, sausage, fish and eggs

- Important for animal protein
- Rich on iron
- 2- (3) times a week meat, 3 times a week 2 slices of sausages or lean ham
- Once a week fish
- 2-3 eggs per week
Food pyramid
Lunch plate rule
Write a Nutrition Plan

- Calculate the individual necessary energy intake

- 5 meals
  - breakfast and morning snack 1/3 each
  - lunch 1/3
  - Afternoon snack and dinner (incl “bed hop”) 1/3 each
One healthy day
Rules for Healthy Food

- Varied, but not too much food
- Cook yourself with fresh ingredients, avoid ready to serve meals
- Cook tasty and with care on the vitamins
- Enough grain and potatoes
- Plenty of vegetables
- Daily low fat milk and milk products
- Less fat and no high caloric food
- Use herbs and spices for a better taste
- Accept the natural taste of food
- Avoid sugar and high-glycemic-index-carbohydrates
- Drink enough of caloric free drinks per day
- Avoid fried food
- Take your time when you are eating
Psychological Aspects of Nutrition

- “My plate – my meal”
- Write individual food plan
- Plans have to be given also to grandparents, nursery school/kindergarten/care giver
- Cut the bread in pieces - looks more
- Do without butter to reduce calories
- Add vegetables - looks more
- Drink one glass of water or one cup of tea before every meal
- Be sure that there is no uncontrolled access to food, if verbal rules are not accepted locking the kitchen is necessary
- Involve the individual with PWS in decisions if possible
- Be prepared for the possible change in behavior
Clients with PWS like their daily routine - it gives them safety

- Clearly defined eating rules
  - Where are places to eat?
  - When is the right time to eat?
  - What is to be eaten?
  - No eating in front of the TV

- Changes irritating them and may trigger behavioral problems
- Nutrition is the most sensitive point
- be aware of extra energy intake - from unplanned food access
- Explain the exceptions hardly in advance
- Nutritional control has to be in the hands of the parents and caregivers
Nutrition Management in PWS needs control

- Control weight on a regular basis: daily or weekly
- Nutrition and Exercise together can prevent loss of muscle mass under low caloric diet
- Biological impedance analysis is helpful to control body composition