

Nutrition in PWS

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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

Adaptive thermo genesis	10-30%	Nutrition intake and composition Surrounding temperature
Body activity	20-40%	Normal activities Sports
Basal metabolic rate	50-70%	Genetics Basic muscle tone Age Gender Hormones Non fat body mass

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

age	Boys with PWS (kcal)	Girls with PWS (kcal)
1-3 y	800	720
4-6 y	950	850
7-9 y	1100	1000
10-12 y	1250	1100
13-15 y	1500	1250

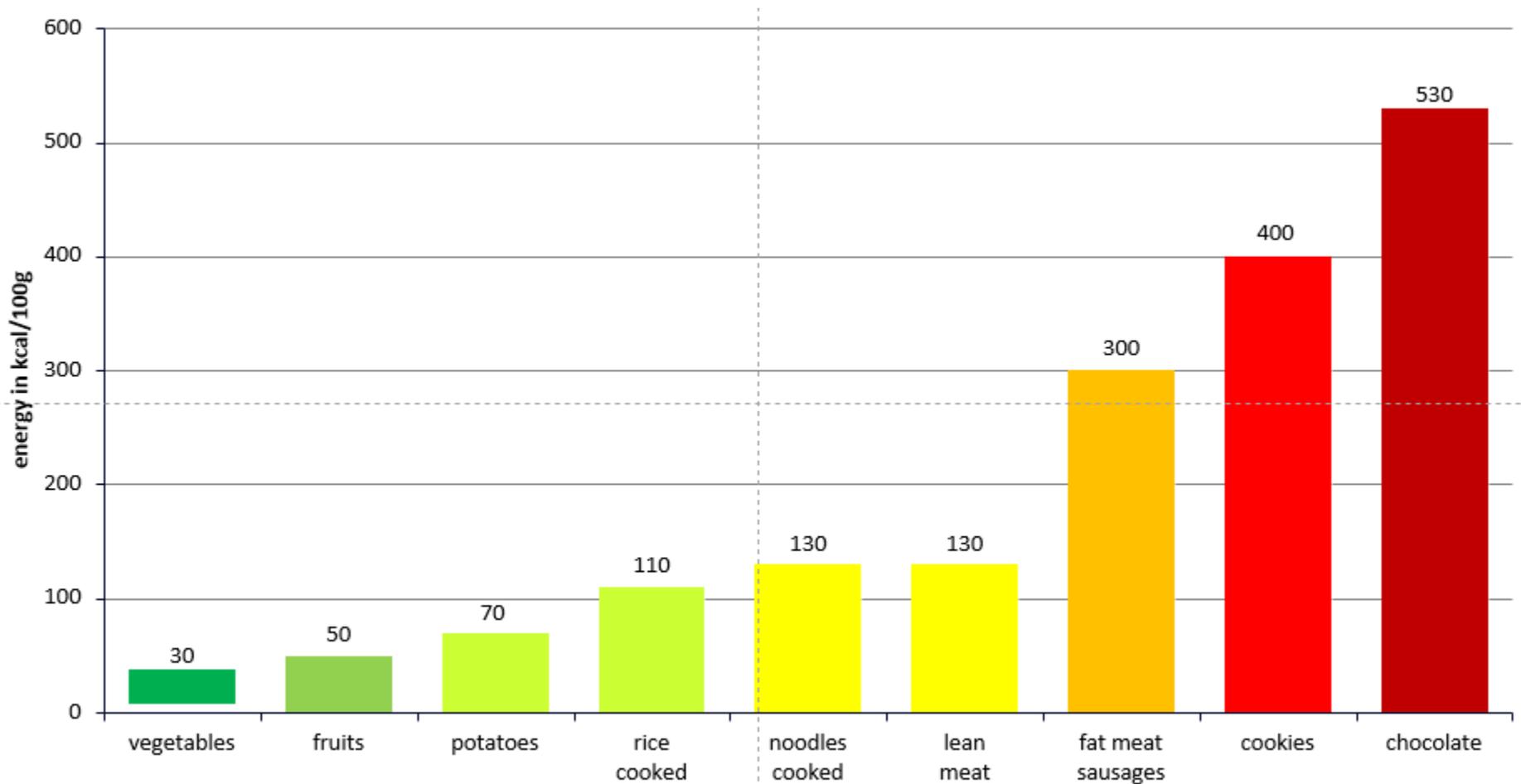
>15 years and for adults:

- 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

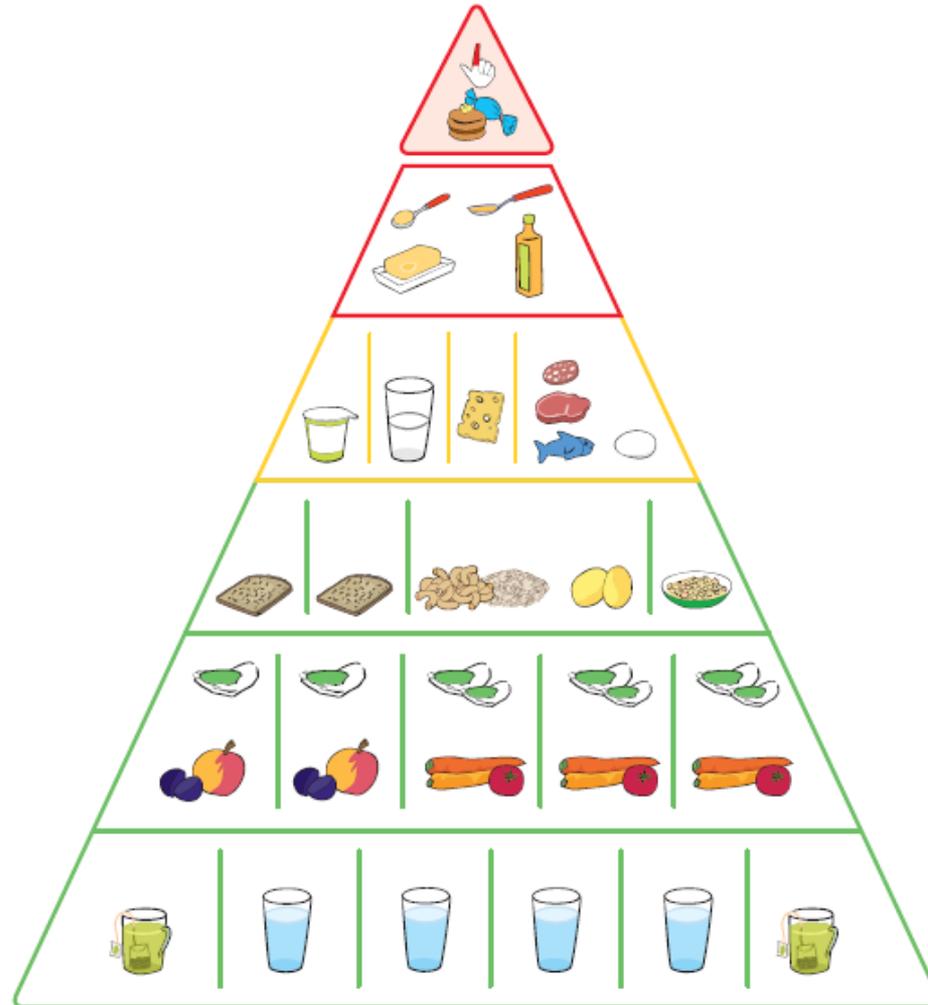
Nutrition for individuals with PWS

- 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

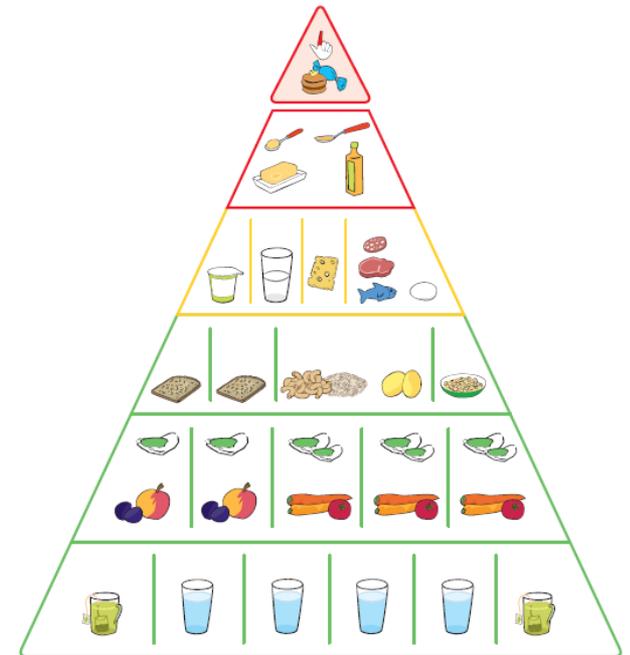


Food pyramid



Vegetables 3 x per day

- Low caloric density
- Less than 30 kcal/100 g (except 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 swalling problems prefer cooked or blanched vegetables

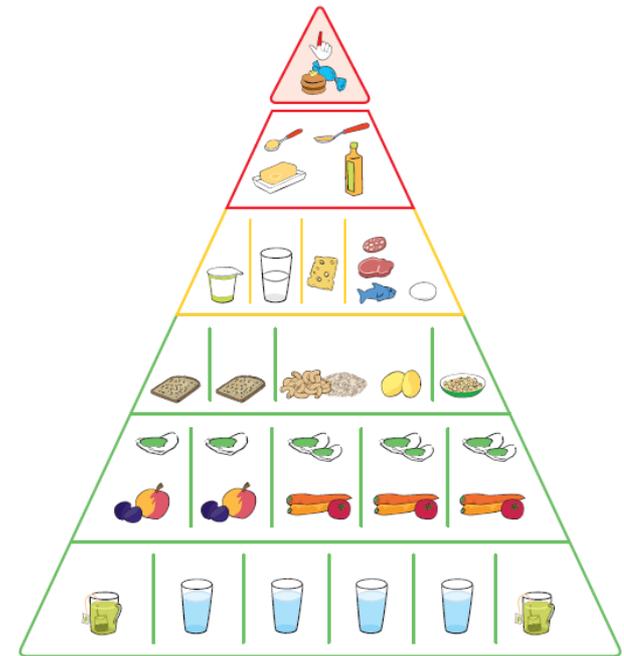


Vegetable helps to reduce energy intake – make them look good



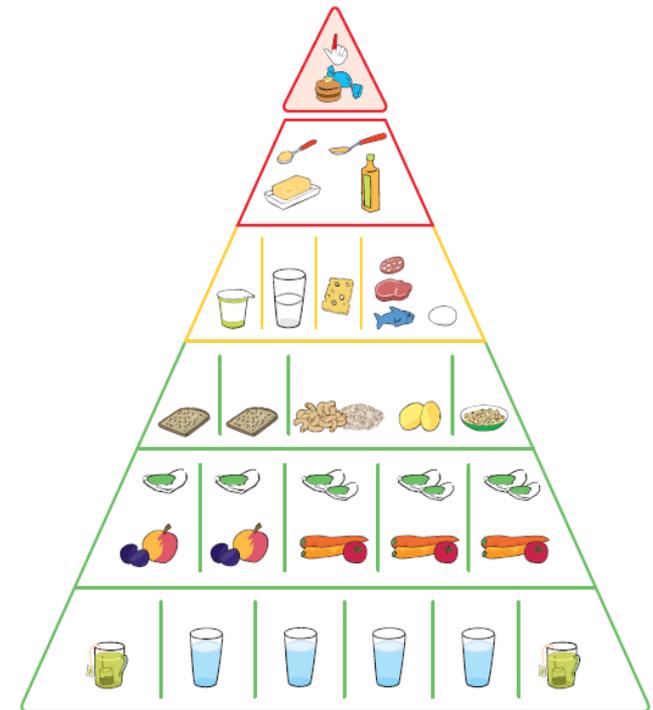
Fruits – 2 x per day

- 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



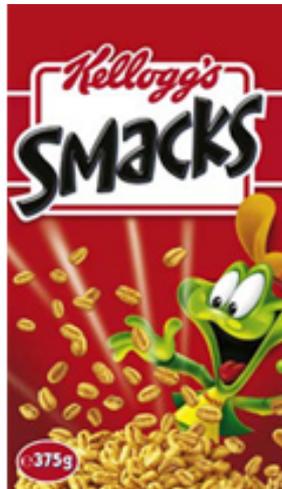
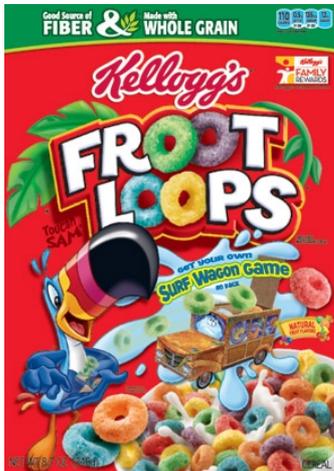
Products from Rice, Grain, Pasta, Potatoes

- 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

In 100g	Fruit Loops	Smacks	Cornflakes	Toppas	Oat müsli
carbohydrate g	80	84	84	72	50
Sugar g	25	43	8	17	3,9



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

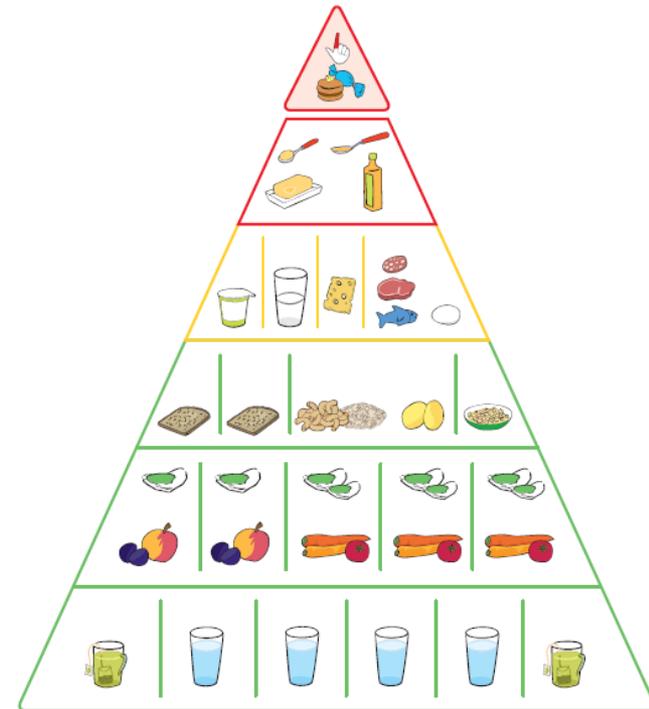
Milk and milk products

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

- Cheese

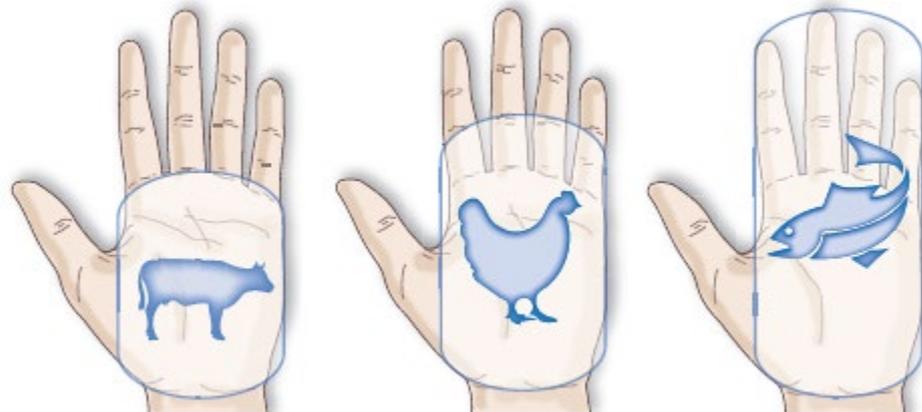
- 70% Fat i.d. 130 kcal/30 g
- 40% Fat i.d. 85 kcal/30 g
- 10% Fat i.d. 35 kcal/30 g

- Cheese with 30% Fat i.d. and 1,5% milk are ok

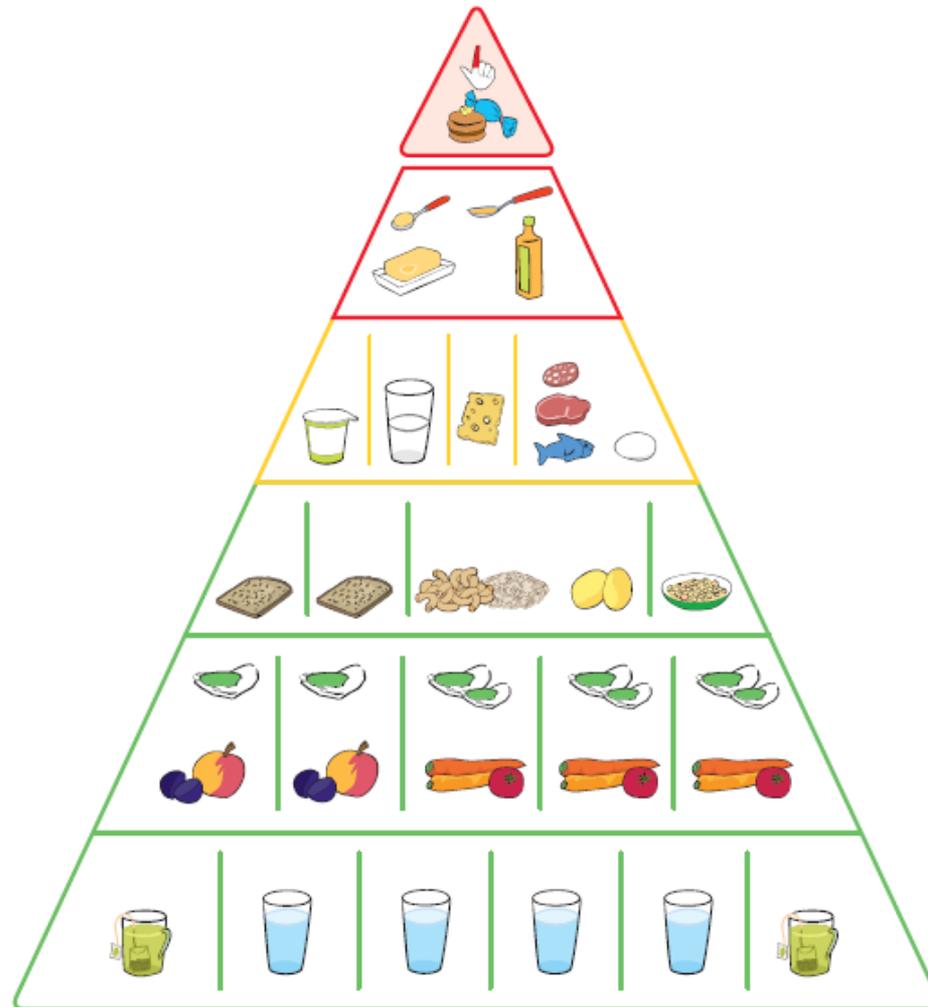


Meat, sausage, fish and eggs

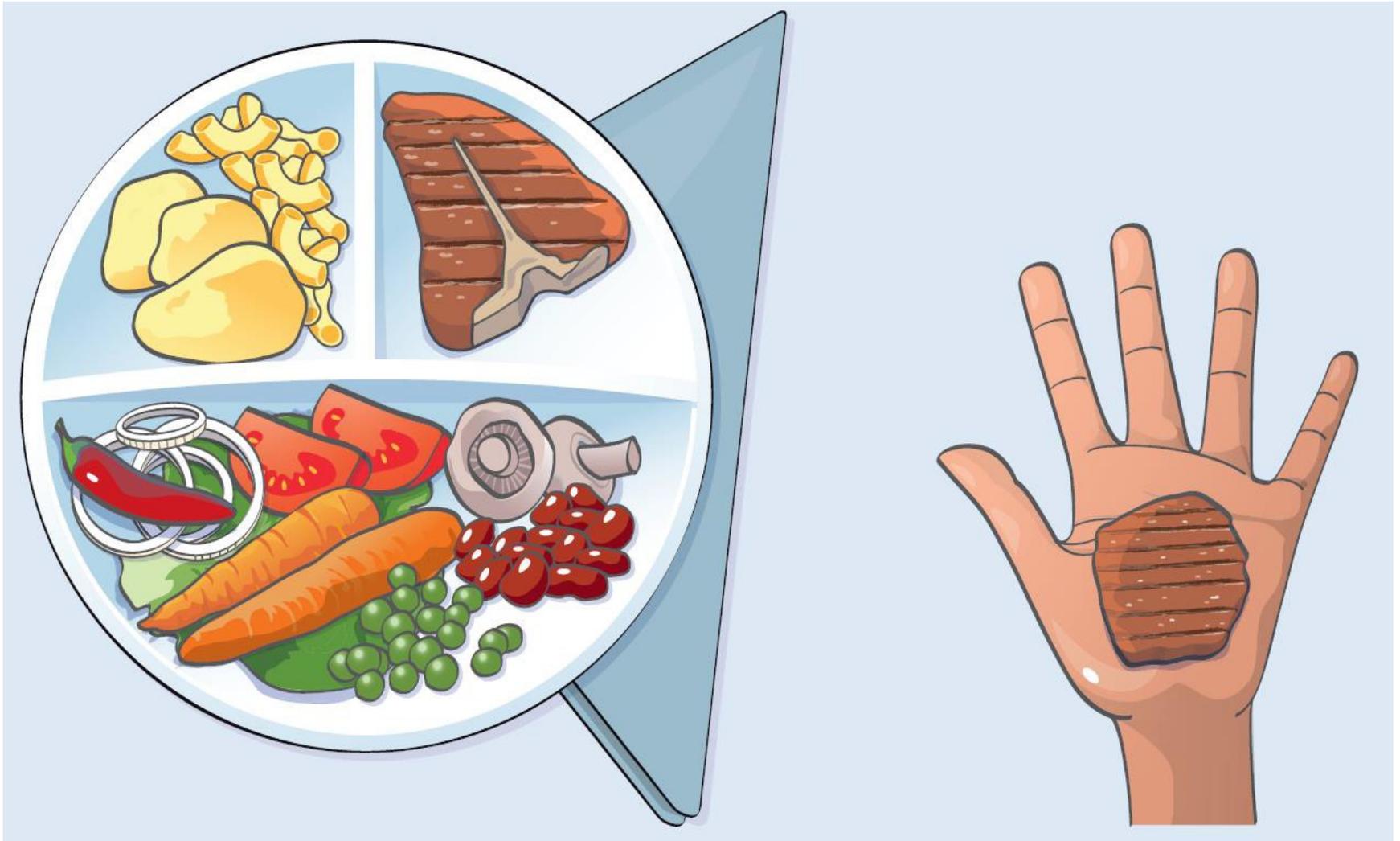
- Important for animal protein
- Rich on iron
- 2- (3) times a week meat, 3 times a week 2 slices of sausages or lean ham
- Once a week fish
- 2-3 eggs per week



Food pyramid



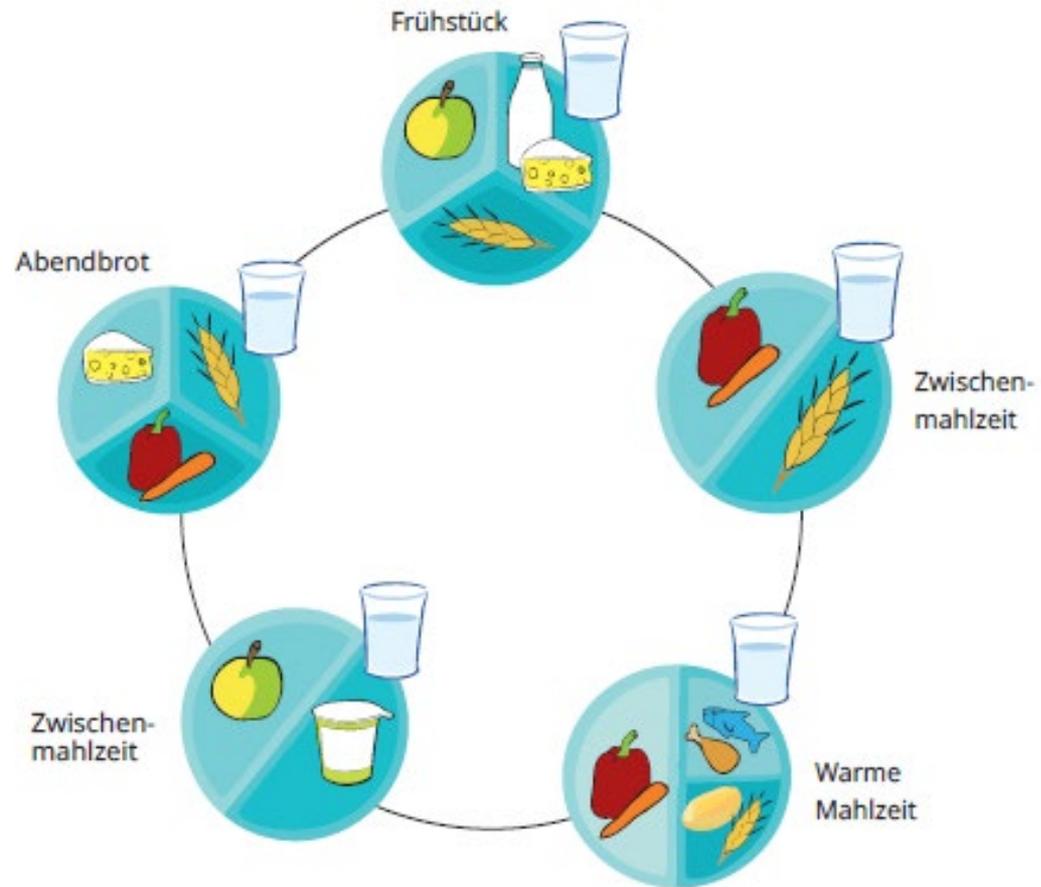
Lunch plate rule



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

Everyday nutrition- Sunday nutrition

- 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

