

Treatment

- How much phe to add
- Phe level Dietary Phe

≤ 10	70 mg/kg
≥10 ≤20	55
≥20 ≤30	45
≥30 ≤40	35
≥ 40	25
- Phe is necessary for growth
- Weekly or biweekly DBS with diet records
- Adjustments are based on levels and growth
- Phenylalanine is an essential amino acid – must gotten in the diet

Home Monitoring

- Weekly or biweekly until 1 yr of age
- Twice a month from 1 yr to 12 yrs
- Monthly after 12 yrs of age
- Weekly to biweekly during pregnancy
- Diet records should be sent with every spot – the type of food and amount are listed as well as the phe/protein content

Adjusting the diet

- Every age group has recommended Phe and protein levels as well as calorie, vitamin and minerals (within any age group their can wide variation in the amount of phe required)
- Diet adjustments are based on Phe levels – either home monitoring or PAA from clinic
- Diet record
- Calorie intake
- Illness – teething, fevers, shots, etc
- Exercise

What's in the formula?

- No phe
- Extra tyrosine (melanin, hormones, neurotransmitters)
- Protein
- Vitamins/Minerals
- Calories

Typical Diet - Infant

- 0 to 4-6 months – formula combination of regular infant formula/breast milk and metabolic formula
- 4-6 months – start baby food and progress normally, continue formula combo, start to decrease infant formula/breast milk
- 6-12 month – continue to progress baby foods (fruits, vegetables, cereal, puffs, depending on infant start table foods), continue formula combo – decreasing infant formula/breast milk

Infant formulas

- Phenyl free 1 – Mead Johnson
- Phenex 1 – Abbott
- Periflex infant – Nutricia
- These contain protein, calories, vitamins and minerals appropriate for infants
- Usually continued until 18 mos – 2 years, amounts are decreased as we increase the next appropriate age formula

Sample Infant diet

- 9 month old
- Phe goal from food and formula – 250 mg
- Formula mix of Similac and Periflex – 18.9 ounces = 127.7 mg phe (6.74 mg phe/oz)
- Food -average was 125 mg phe – rice cereal, strained carrots and strained fruit.

Toddler

- Plain metabolic formula
- Transitioning from infant to older formula that is more appropriate for age and nutrient needs
- All phe is coming from foods
- Fruits, vegetables, low protein foods vs regular grain depending on phe tolerance

Formulas for PKU – 1yr to adulthood

- Mead Johnson – phenyl free 2, phenyl free 2 HP
- Vitaflo – PKU gel, PKU express 15, PKU cooler 10, PKU cooler 15, PKU cooler 20
- Abbott – Phenex 2
- Cambrooke foods – camino pro, bettermilk

PKU formulas cont'd

- Nutricia/Applied Nutrition – merged
- Nutricia - periflex jr, periflex advance, xphe maxamaid, xphe maxamum, lophlex, lophlex LQ 20, phlexy 10 system,
- Applied Nutrition - phenylade RTD, phenylade essentials, phenylade 40 – citrus or unflavored, phenylade 60 – vanilla or unflavored, Phenylade amino acid blends, phenylade amino acid bars

Why different formulas?

- Differing amounts of protein –infant formulas have more protein
- Calories/ nutrients
- Volume
- Convenience
- Taste
- To make adherence to diet easier

Typical diet for a 4 year old

1 slice toast - 100 mg phenylalanine
 2 tbsp margarine – 12 mg
 1 medium banana – 43 mg
 1 lemon pudding - free
 ½ cup blueberries – 18 mg
 ½ cup baked potato (no skin) – 56 mg
 10 strawberries – 14 mg
 12 cherry tomatoes – 31 mg phe

TOTAL – 274 mg phe and 650 calories
 29 ounces of phenyl free 2 - 31.6 mg protein and 589 calories
 Food and formula provide – 274 mg phe, 37 gm protein and 1250 calories

Typical diet for an 11 yr old

1 cup froit loops – 88 mg phenylalanine
 ¼ cup coffee rich – 8 mg
 8 ounces apple juice – 2 mg
 1/3 medium banana – 14 mg
 10 french fries – 76 mg
 1 tbsp ketchup – 7 mg
 1 cup lettuce – 28 mg
 1/2 cup tomato – 22 mg
 2 tbsp mushrooms – 7 mg
 2 tbsp Italian dressing – 0 mg
 1 gel snack cup – free
 10 jelly beans – free
 1 cup lo protein spaghetti, dry – 14 mg
 ¼ cup spaghetti sauce – 24 mg
 ¼ cup green beans – 21 mg
 1 slice low protein bread – 15 mg
 1 tbsp margarine – 6 mg
 ½ cup strawberries – 13 mg

TOTAL- 345 mg phe and 1242 calories
 5 scoops phenylade essential - 50 gm protein and 785 calories
 Food and formula provide 345 mg phe, 56.9 gm protein with 2027 calories

Typical diet for an adult

2 low protein pancakes – 4 mg phenylalanine
 1 tbsp margarine- 6 mg
 Syrup – 0 mg
 1 cup chunky veg. soup RTS – 160 mg phe
 3 saltine crackers – 43 mg
 1 medium banana – 43 mg
 1 cup baked potato – 118 mg
 ¼ cup cooked broccoli – 35 mg
 2 tbsp margarine – 12 mg
 1/3 cup low protein shredded cheese – 42 mg
 ½ cup watermelon – 12

TOTAL – 475 mg phe, 9.5 gm protein, 1043 calories
 3 PKU 20 coolers = 60 gm protein, 372 calories
 Food and formula provide – 475 mg phe, 69.5 gm protein,
 1415 calories

Maternal PKU

- A woman with PKU must be on diet for 3-6 months before trying to conceive
- Prevent maternal PKU syndrome – cardiac issues, microcephaly, intellectual disability
- Diet, formula and calories are critical
- Monitoring 1-2 times a week
- Initially – phe intakes are 100-200 mg a day

Why diet for life?

- Prevent – decreased IQ scores, eczema, behavioral problems, seizures, decreased executive functioning, depression, irritability, headaches, impairment of short term memory, inability to hold a job

Why diet for life?

- Prevent health problems caused by high phe levels
- Have children reach their intellectual potentials
- Have children and adults who are contributing members of society