

Worksheet 26-1: Calculation of Nutrition Needs for a Child with PKU

Using the information in Table 26.7, calculate the nutrition needs for P.H., a female with PKU, during the various stages of her life.

Age	Weight	PHE	TYR _{mg}	Protein	Energy	Fluid
1 month	10# 4.5	1125-315	1350-1575	15.9-13.6	544.6	720-606
4 months	12# 5.5	1595-3025	1650-1925	19.1-16.4	659	664-702
6 months	14# 6.4	96-224	1920-2240	19.1-15.9	699	413-787
10 months	18# 8.2	82-267	2050-2460	24.5-20.9	858	984-1100
2 years	24# 10.9	200-400	1972-300	≥ 30	1300	900-1600
5 years	35# 15.9	210-450	225-3.50	≥ 35	1700	1300-2300
9 years	52# 23.6	220-500	255-4.00	≥ 40	2400	1650-3300
12 years	75# 34.1	250-750	345-7.00	≥ 50	2200	1650-3300
16 years	105# 47.7	230-700	345-5.00	≥ 55	2100	1650-3300
21 years	115# 52.3	220-700	3.75-5.0	≥ 60	2100	1650-3300

1. When is the need for protein the highest for P.H. (grams/kg)? Why?

birth → 6 months

→ accommodating growth needs

2. When is the need for phenylalanine highest for P.H. (milligrams/kg)? Why?

birth → 6 months

→ accommodating growth needs

3. Why are the needs for protein and phenylalanine higher in males than in females after age 11?

Men have higher lean muscle mass

Worksheet 26-2: Diet Prescription—Meal Plan for Child with PKU

P.H.'s mother is seeing an outpatient dietitian to determine the phenylalanine content of the current meal plan for P.H. Use the USDA National Nutrient Database (available at <http://www.nal.usda.gov/fnic/foodcomp/search/>), Table 26.8, and Box 26.1 to search for the phenylalanine and calorie content of the sample menu you have planned based on her preferences and tolerances. Check to make sure the meal plan is meeting the nutrition needs for 5-year-old P.H. (210-450 mg phenylalanine and 1700 kcal). Note that the current phenylalanine-free formula they are using provides 22 kcalories per ounce.

Food/Beverage	Phe (mg)	Kcal
Breakfast		
½ cup Cinnamon Toast Crunch Cereal	30	90
1 medium apple	8	12
8 ounces phenylalanine-free formula	0	176
Lunch		
2 slices low-protein bread	30	200
1 slice low-protein cheese	30	60
8 medium baby carrots, raw	21	26
1 tablespoon low-fat mayonnaise	6	120
8 ounces phenylalanine-free formula	0	176
½ cup grapes	6	31
PM Snack		
4 ounces phenylalanine-free formula	0	88
1 cup popcorn	35	35
Dinner		
½ cup medium-grain brown rice, cooked	116	109
½ cup stir-fried broccoli	7	29
1 teaspoon butter	2	34
½ cup applesauce	6	97
8 ounces phenylalanine-free formula	0	176
Bedtime Snack		
8 ounces phenylalanine-free formula	0	176
2 graham cracker squares	48	59
Total:	345	1746

BOX 28.1 CLINICAL APPLICATIONS

Case Example for Phenylketonuria

Mary is a 4-year-old girl with PKU. Weight = 17 kg, Height = 100 cm. She is currently taking 150 grams of Phenex1, a metabolic formula powder, which is mixed with water to make 32 oz. of liquid formula. This provides her with 22.5 g of (phenylalanine-free) protein and 720 kcal. The remainder of her protein and kcal are provided by her diet. Mom reports that Mary is often not hungry because her formula fills her up. She also reports difficulty getting Mary to eat the recommended amount of phenylalanine.

Flow Sheet

Month	Phe Level	Caloric Intake	Protein Intake	Phe Intake
January	4.1 mg/dL	1,400	30 g	325 mg
February	3.8 mg/dL	1,450	29.5 g	315 mg
March	5.3 mg/dL	1,350	29.5 g	315 mg
April	7.2 mg/dL	1,400	29.5 g	320 mg

Estimated needs (based on Table 28.4):

- 30–35 g protein
- 1,400–1,700 calories
- 310–320 mg phenylalanine

Discussion:

Mary's phenylalanine intake has been generally within the prescribed range, but blood phenylalanine levels have increased over the last few months. Mary's weight is between the 50–75th percentile; her height is at the 25–50th percentile growth curves. Kcal and protein intakes are on the lower end of recommended range.

The increase in blood levels could be related to increasing protein needs due to growth. Mary would benefit from an increased protein intake, but if she consumed more protein from her diet, she would also receive more phenylalanine. Consuming more formula would likely further decrease her appetite and daily food intake. This could lead to compromised metabolic control since phenylalanine is an essential amino acid, and a lower than required intake could lead to catabolism and cause an increase in her blood level.

Plan:

Change Mary's metabolic formula to one that is more concentrated in protein and provides fewer kcal. This type of formula is designed for older children and adults and contains greater concentrations of protein along with more vitamins and minerals to better meet her needs. The plan would be to transition Mary to the more concentrated formula to provide her more protein in less volume. The new formula Phenex 2 will contain 350 kcal and 25 grams of protein in 85 grams of powder. Mixed with water, this amount of powder will make

16 oz. of formula. If Mary keeps her dietary protein intake restricted at 7 grams, she will now be receiving 32 grams of protein each day. A decrease in her phenylalanine intake, to 300–310 mg each day will also promote improved blood levels. The decreased volume and kcal from formula will help to stimulate Mary's appetite, but since she has to also slightly decrease her dietary phenylalanine intake, she will have to include more special low-protein foods in her diet. These specially designed foods often are fairly high in kcal and will help ensure she meets her caloric needs while not exceeding her phenylalanine prescription. See the adjusted meal plan shown in the table.

Original Meal Plan			Adjusted Meal Plan		
Food/beverage	Phe (mg)	kcal	Food/beverage	Phe (mg)	kcal
Breakfast			Breakfast		
1/2 cup Froot Loops	56	82	1/2 cup Froot Loops	37	55
1 banana	43	105	1/2 cup low-protein cereal loops	1	52
8 oz. formula	0	180	1 banana	43	105
			4 oz. formula	0	88
Lunch			Lunch		
1 slice low-protein bread	15	100	2 slices low-protein bread	30	200
1 slice low-protein cheese	30	60	1 slice low-protein cheese	30	60
20 goldfish crackers	36	52	20 low-protein pretzels	34	112
1/2 cup canned peaches	17	29	1/2 cup canned peaches	17	29
1 tsp. mayonnaise	2	40	1 tsp. mayonnaise	2	40
8 oz. formula	0	180	4 oz. formula	0	88
			Juice box	0	100
Dinner			Dinner		
1 cup low-protein pasta	8	150	1 cup low-protein pasta	8	150
1/2 cup spaghetti sauce	27	25	1/2 cup spaghetti sauce	27	25
1/4 cup broccoli	49	16	1/2 cup broccoli	33	11
8 oz. formula	0	180	4 oz. applesauce	6	97
			4 oz. formula	0	88
Snack			Snack		
8 oz. formula	0	180	4 oz. formula	0	88
1 cup popcorn	35	35	1 cup popcorn	35	35
Total	318	1414	Total	303	1418

metabolic formula (e.g., Phenex, Phenylfree). The metabolic formula is usually a powder that contains all of the amino acids except for the one(s) that are not able to be metabolized appropriately. The powder formula can contain a carbohydrate source, a fat source that includes essential fatty acids, and vitamins and minerals. Many of the metabolic formulas also contain carnitine, utilized to scavenge toxic metabolites in certain amino acid disorders.

Formulas containing little carbohydrate or fat have recently been developed in an attempt to minimize the caloric contribution of the formulas. It is important to determine whether extra kcal are needed to promote satiety and to decrease the intake of foods containing the offending amino acid(s). On the other hand, as more individuals are successfully treated and reaching adulthood, lower caloric needs are indicated due to decreasing rates of growth. Higher-kcal formulas can result in excessive weight gain in these individuals. Newer formulas that contain little or no vitamins and minerals have also been developed in order to improve the palatability of the product. These products need to be used in conjunction with a vitamin/mineral supplement. Additional

calcium may be needed if using an unfortified product, given the restricted use of dairy products and inadequate amount of calcium in a general multiple vitamin/mineral supplement. Formula bars and tablets are now available for certain disorders in order to meet the demands of the more active, older patients who do not want to prepare a liquid formula each day.

Intake of small amounts of metabolic formula throughout the day provides for the best utilization of the synthetic amino acids. Studies have shown increased nitrogen loss when the formula is consumed 1 to 2 times per day versus 4 to 5 times per day (MacDonald et al. 2003). Although an essential part of the treatment, the coverage of metabolic formulas by various health insurance plans can vary considerably between states.

Calculated protein needs are determined by weight in a growing child and are higher than the standard recommended dietary intakes (RDI) for age, due to the synthetic nature of the protein in the formula (Kindt and Halvorsen 1980). Although guidelines are available to help estimate protein and amino acid requirements, needs are best determined

TABLE 28.5

Phenylalanine Content of Selected Foods

Food	Weight (g)	Approximate Measure	PHE (mg)	TYR (mg)	Protein (g)	Energy (kcal)
<i>Cereals, Cooked, measure after cooking</i>						
Corn grits						
instant						
cheese flavor	36	¼ packet	36	29	0.7	27
plain	34	¼ packet	27	22	0.5	20
regular, quick (plain)	45	3 Tbsp	33	28	0.6	28
Cream of rice	81	½ cup	30	40	0.7	42
<i>Cream of Wheat</i>						
instant	30	2 Tbsp	30	18	0.6	19
Mix'n Eat						
flavored	38	¼ packet	33	20	0.6	33
plain	36	¼ packet	37	22	0.7	26
quick	30	2 Tbsp	25	14	0.4	16
regular	30	2 Tbsp	30	18	0.5	19
Oats, regular, quick, and instant	20	1 Tbsp + 1 tsp	28	18	0.5	12
<i>Cereals, Ready to Eat</i>						
All Bran®	5	1 Tbsp	29	22	0.8	13
Apple Jacks®	9	½ cup	28	18	0.5	36
Cap'n Crunch®	12	½ cup	33	25	0.6	51
Cheerios®	4	3 Tbsp	35	23	0.6	17
Cinnamon Toast Crunch®	19	½ cup	30	18	0.7	80