

## Chapter 8: 18+ to Adulthood



### A Parent's Perspective

*"For individuals with PKU, a controlled diet for life is the only answer to unlock the greatest opportunities."*

### What to Expect

**Take this quiz to see your level of independence. Strive to achieve checkmarks for all line items -- and remember, your PKU clinic team is always there to help!**

- I can explain PKU.
- I speak confidently about food restrictions.
- I am comfortable ordering appropriate foods at restaurants.
- I can plan my PKU meals independently.
- I know how to order medical formula and ensure I have enough . in advance.
- I know to eat low protein foods and formula before an event to avoid being hungry and overdoing . it at a party.
- I take my blood samples independently.
- I keep track of my diet record independently.
- I attend medical appointments with my PKU team regularly.
- I have discussed sexual health with my healthcare team and understand how to use birth control effectively.
- I know the dangers of high Phe levels and know to see my PKU clinic for advice and support.
- I have information about my insurance and manage it independently.
- I am aware of the resources available to me.

Adulthood can be a very difficult time for an individual with phenylketonuria (PKU). Some people with PKU have relaxed their strict diet, but begin to realize that their attention, concentration and relationships have been negatively impacted by high phenylalanine (Phe) levels <sup>46</sup>.

High Phe levels can impair your executive functioning capabilities, such as memory, planning, attention, and organization. As these are the very skills that are needed for managing PKU, it is important for you to maintain control so that you have the ability to plan your diet, remember Phe intake for your records, remember to drink your formula, maintain your PKU supplies and monitor blood Phe<sup>47</sup>. Following your low Phe diet and treatment is the best way to avoid neurocognitive problems and to maintain your ability to organize your life with PKU. Maintaining your diet and treatment will help you to feel healthy both physically and mentally. Diet for life – maintaining the PKU diet throughout your lifetime – is the best way for you to avoid problems associated with excess Phe.

The good news is that metabolic control for life is possible. The low protein food and formula options available today make the PKU diet more manageable than ever, and research continues to uncover new ways to treat PKU. Most people with PKU find they have more success maintaining treatment for life when they are supported, have a positive attitude, and find a way to make the PKU diet manageable for them. This is known as S.A.M.

<sup>46</sup> Waisbren, S. *The Psychology of PKU*. NPKUA annual conference in Dallas, January 2010 Presentation

<sup>47</sup> Waisbren, S. *The Psychology of PKU*. NPKUA annual conference in Dallas, January 2010 Presentation

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## S.A.M: Support, Attitude, Manageability<sup>48</sup>

S.A.M. is an acronym for the key factors that lead to success in staying on diet or returning to the PKU diet. It is natural to sometimes feel like you can't succeed or the diet is too difficult to follow. However, it is important not to lose all hope and give up. Sometimes you just need a reminder of how to handle a certain situation. Just remember S.A.M. is here to help see you through the hard times and assist you in achieving better health.

**S is for Support:** This means you have people around you who believe in the treatment for PKU and support you in following the diet.

You are not alone. Building a base of people who support you in maintaining the diet is critical. Whether it be a friend, parent, significant other or a relative, having someone to confide in, talk to, and yes, even complain to is important. Who you choose to talk to is up to you, but you need people who believe in the benefits of “diet for life.”

Speak with your local PKU team to find out contact information for other people in your area with PKU. You will not only gain support from someone who understands, but you will also have the chance to make a new friend. There are also online support groups and websites listed at the back of this resource that allow you to voice your thoughts, share ideas, and meet others that have PKU and understand the need for diet.

**A is for Attitude:** This means that you have a positive attitude about your formula and following a low protein diet.

The attitude you have toward PKU and following the diet is directly related to your success. A positive outlook and attitude is critical toward gaining and maintaining metabolic control. In terms of the PKU diet, a positive attitude means being open to trying new things, exploring new formula and food options, and accepting PKU as a part of you. Being angry and mad about having PKU will not remove the fact that you do need a special diet. Overcoming a negative attitude toward your situation will remove a major barrier to your future success. Viewing the PKU diet as just a different style of eating can boost your overall acceptance of the diet and foster a positive outlook. You may realize that following a strict diet has taught you skills that will help you excel in your daily life, such as increased creativity, self control and healthy meal planning.

**M is for Manageability:** This means you have found ways to make the diet work for you.

Manageability, as it relates to PKU, is how you are going to implement the diet into your daily life. It can mean anything from obtaining insurance coverage for medical foods to what you do in certain situations, like eating out or social gatherings. Manageability is your own custom-made plan for following the PKU diet and finding one that works best for you. When you have the right tools, anything is possible. Writing

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<sup>48</sup> Waisbren, SE, et al (1995) Psychosocial Factors in Maternal Phenylketonuria: Women's Adherence to Medical Recommendations. *American Journal of Public Health* 85: 1636-1641.  
Finkelson, L. Bailey I, and Waisbren, SE (2001) PKU adult and their return to diet. Predicting diet continuation and maintenance. *J. Inherit Metab. Dis* 24: 515-516.

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a plan that includes the time of day you are going to consume your medical food and what you will do if you are away from home, will help you be prepared.

## Transitioning to Adult Care

### Transitioning

The term transition means change. All people face transitions at various times in their lives. In regards to a person with PKU, transition also means a change in responsibility for your health and medical needs. Now is the time to learn how to rely more on yourself and take charge of your own PKU needs. Transition can also mean<sup>49</sup>:

- Taking on new challenges.
- Gaining a new sense of freedom and independence.
- Having more choices.
- Doing more things on your own.
- More responsibility<sup>50</sup>.



### *Your Transition Checklist*

Get in the practice of doing the following things. They will help make your move to adult health care management easier.

- Know how to explain PKU and communicate your health care needs.
- Keep a record of appointments, medical history and medications prescribed.
- Write down your doctors' names, phone numbers and addresses.
- Begin to make your own medical appointments.
- Write down questions for your doctor before your visit.
- Have parents, friends, or your significant other remain in the waiting room while you spend time with your healthcare provider on your own.
- Learn about your health insurance and type of formula coverage your policy offers.
- Learn new information about the health care needs of an adult with PKU.
- Prepare and educate yourself on personal PKU management. For example: How to obtain formula, monitor your blood PHE levels, count PHE in your diet, etc<sup>51</sup>.
- If you're more comfortable, you can consider having someone with you for support, like a friend, parent or someone else you trust.

## Relationships

### *Your Family*<sup>52</sup>

Up until now, your family or someone in your family has taken care of you and your needs, PKU and otherwise. They have brought you to your healthcare provider and clinic visits, and have taken care of you when you were sick. Now that you are the leader of your own health care and PKU management, your family will have a transition also.

<sup>49</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey: Applied Nutrition Corp. 2007:21

<sup>50</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey: Applied Nutrition Corp. 2007:22.

<sup>51</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey: Applied Nutrition Corp. 2007:22.

<sup>52</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey: Applied Nutrition Corp. 2007:29.

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The maturity associated with adulthood will overflow into your daily life. Your family will have to understand that now you make the important decisions regarding your PKU needs. In order to not be viewed as a child, you must act like an adult and accept responsibility for your actions.

Your family will always be there to support you with the endeavors and choices you make throughout your life. Your transition to managing PKU on your own should be no different. Remember, your family is still a tremendous source of knowledge regarding your dietary needs and you can always go to them for support and advice when needed.

## *Your Friends*<sup>53</sup>

The relationship you have with your friends will also experience a transition as you grow and age. As you get older, the friends you already have and the new friends you make may want to know more about PKU and your diet. Embrace this. Talk openly and share with them what life is like with PKU. Having the support from your peers will only make following the diet easier.

## **Treatment and Diet**

### **Returning to Treatment**

As an adult, you may have strayed from the strict, low protein diet and allowed a degree of relaxation to your diet. You may have many questions about how to transition back to the PKU diet and maintain your treatment. If you are an adult who, for whatever reason, has modified or abandoned PKU treatment for a period of time but would now like to return to metabolic control to protect your health and mental functioning, this next section will help you.

### **Medical Formulas**

You may be familiar with traditional powder formulas, which are commonly used as the main source of protein for people with PKU. More recently developed medical formulas come in many new flavors that some may find more palatable than previous medical food, convenient single serving sizes (such as formula packaged in juice box sized containers that are ready to drink) and in powders that can be added to foods and beverages. Some of these formulas are lower in volume than traditional formulas as they are lower in fats and carbohydrates, but most of them have the required vitamins and minerals. You can try out different styles and flavors to find the right formula for you<sup>54</sup>.



<sup>53</sup> Maltzman, S. : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey: Applied Nutrition Corp. 2007:29

<sup>54</sup> Naziri, M. *Meal Planning on a Budget* PKU Bootcamp Children's Memorial Hospital. April, 2011 Presentation

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## Low Protein Foods

Offerings of foods modified to be low in protein have also expanded significantly. Specially formulated breads, pastas, non-meat burgers, cheeses, muffins and cookies are now available, among others, and low protein food companies continue to offer new options for people with PKU.



There are also less expensive foods naturally low in protein that available off the shelves such as a few listed below.

PRODUCT	PRICE	PHE
Sunbelt fruit & grain cereal bars (blueberry/strawberry)	\$2.00 per box	45-55 mg per bar
Sensible Portions Veggie Straws	\$6.00 per 7oz bag	32 mg per serving (38 straws)
Sandwich Mate cheese slices	\$1.50 per pack (16 slices)	24 mg per slice
Sun Luck Rice Sticks	~\$2.00 per bag	35 mg per serving (1/4 of bag)
KAME bean threads	~\$1 per bag	3 mg per 56 gram serving (uncooked)
Pepperidge Farm Very-thin sliced white bread	~\$4.20 per loaf	67 mg per slice
Turtle Mountain So Delicious Coconut Milk Yogurt	~1.80 per 6oz	28 mg per 6oz

Ask your PKU team for more examples of low protein foods you can get off the shelf. Also, the resource section of this resource has information on companies who offer medical foods. Some food companies offer free shipping with a certain amount of food ordered.

### A Parent's Perspective

*"I met David (who is now my husband) and realized that there really could be a future for us. Simply thinking about the future- marriage, children, etc. made me realize that I had to get this under control! This was the first spark in my adjusting back to the regular PKU life and habits."*

## Transitioning to the PKU Diet<sup>55</sup>

Your PKU team is a good resource as you return to diet, and will help you work out a treatment plan. It is important to have good support from family and friends as you make this transition.

When an individual is off their low Phe diet and not drinking any formula, 100% of their protein comes from dietary intake of natural protein. This will lead to high Phe levels, and

<sup>55</sup> Naziri, M. *Adult Outreach*. December, 2009 Presentation

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can lead to health issues previously discussed. When you decide to return to diet, your PKU team will help develop a step-by-step process to slowly reduce the amount of protein coming from natural food items, and increase the protein intake from formula to balance your diet and reduce your Phe levels. This will involve substituting medical formula and low protein foods for high protein foods in your diet over time. An example of how this transition might work is below. As this example shows, you will transition gradually to your diet for life, and your PKU team will work closely with you to ensure that you know how to create a diet that will work for you and your lifestyle. Some people will have been eating a modified low Phe diet, so you may find your diet is more similar to one of the later steps in the diet. You can find where you currently fit, and start from there.

**Some people who have been off diet say that when they return to the PKU diet they:**

- *Feel better*
- *Look better*
- *Are less moody*
- *Find it easier to get along with others*
- *Have more energy*
- *Can concentrate*
- *Can think clearly to study or work*
- *Can complete assignments or work projects more easily*
- *Can think more strategically, such as in team sports or at work*

## 5 Steps to the PKU Diet<sup>56</sup>

*Example of The “Off Diet”:*

100% of protein comes from dietary intake of natural protein

BREAKFAST	SNACK	LUNCH	SNACK	DINNER
Coffee 1 egg 2 slices toast	Donut	2 pizza slices 1 breadstick	1 oz. bag of Lay’s chips	cheeseburger medium fries soda
<b>TOTAL PROTEIN: 54 GRAMS</b>				

### **STEP 1:**

75 % of protein comes from dietary intake of natural protein;

25% comes from formula protein

BREAKFAST	SNACK	LUNCH	SNACK	DINNER
Coffee <del>1 egg</del> <b>+ 1 apple</b> 2 slices toast	Donut <b>+ medical formula</b>	2 pizza slices 1 breadstick	1 oz. bag of Lay’s chips	cheeseburger medium fries soda
<b>TOTAL NATURAL PROTEIN: 49 GRAMS</b>				
<b>TOTAL FORMULA PROTEIN: 15 GRAMS</b>				

<sup>56</sup> Naziri, M. *Adult Outreach*. December, 2009 Presentation

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## STEP 2:

50% of protein comes from dietary intake of natural protein;  
50% comes from formula protein

BREAKFAST	SNACK	LUNCH	SNACK	DINNER
coffee 1 apple 2 slices toast	<del>donut</del> medical formula	2 pizza slices 1 breadstick	1 oz. bag of Lay's chips  + <b>medical formula</b>	<del>cheeseburger</del> + <b>bun only</b> + <b>1 leaf lettuce</b> + <b>2 slices tomato medium fries</b> + <b>small fries</b> soda
<b>TOTAL NATURAL PROTEIN: 36 GRAMS</b> <b>TOTAL FORMULA PROTEIN: 30-35 GRAMS</b>				

## STEP 3:

20% of protein comes from dietary intake of natural protein;  
80% comes from formula protein  
+ medical formula  
Total natural protein: 17 grams  
Total formula protein: 60-70 grams

BREAKFAST	SNACK	LUNCH	SNACK	DINNER
Coffee 2 slices toast 1 apple + medical formula	medical formula	<del>2 pizza slices</del> + <b>low protein pizza crust</b> + <b>2 tbsp sauce</b> + <b>¼ cup low protein cheese</b> <del>1 breadstick</del> + <b>1 cup lettuce</b> + <b>2 tbsp Italian dressing</b>	1 oz. bag of Lay's chips  medical formula	bun only + <b>mayo or mustard</b> 1 leaf lettuce 2 slices tomato small fries <del>Soda</del> + <b>medical formula</b>
<b>TOTAL NATURAL PROTEIN: 17 GRAMS</b> <b>TOTAL FORMULA PROTEIN: 60-70 GRAMS</b>				

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## STEP 4:

15% of protein comes from dietary intake of natural protein;  
85% comes from formula protein

BREAKFAST	SNACK	LUNCH	SNACK	DINNER
<b>coffee</b> <del>2 slices toast</del> <b>2 slices low protein banana bread</b> 1 apple medical formula	medical formula	low protein pizza crust 2 tbsp sauce ¼ cup low protein cheese 1 cup lettuce 2 tbsp Italian dressing	1 oz. bag of Lay's chips medical formula	<del>bun only</del> <b>+ low protein Camburger</b> <b>+ low protein bun</b> <b>+ mayo or mustard</b> 1 leaf lettuce 2 slices tomato small fries medical formula
<b>TOTAL NATURAL PROTEIN: 11 GRAMS</b> <b>TOTAL FORMULA PROTEIN: 60-70 GRAMS</b>				

## STEP 5:

10% of protein comes from dietary intake of natural protein;  
90% comes from formula protein

BREAKFAST	SNACK	LUNCH	SNACK	DINNER
medical formula 2 slices low protein banana bread 1 apple	medical formula	low protein pizza crust 2 tbsp sauce ¼ cup low protein cheese 1 cup lettuce 2 tbsp Italian dressing	medical formula <del>1 oz. bag of Lay's chips</del> <b>1 bag of Wise onion rings or ½ cup carrot sticks</b>	medical formula low protein Camburger low protein bun <b>+ mayo or mustard</b> small fries 1 leaf lettuce 2 slices tomato
<b>TOTAL NATURAL PROTEIN: 9 GRAMS OR APPROXIMATELY 450 MG/PHE PER DAY</b> <b>TOTAL FORMULA PROTEIN: 60-70 GRAMS</b>				

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## Tips for Increasing Medical Formula Consumption

As you transition to a low Phe diet, you will be replacing natural protein with protein from medical formula. It is important that you consume the amount of medical formula your PKU team advises to ensure that you are getting enough protein and other nutrients each day. Many adults try to restrict the protein in their diet without drinking the formula. This may result in health issues such as having low vitamin levels, poor bone health and difficulty maintaining a healthy weight.

The following tips can help you get enough medical formula:<sup>57</sup>

1. Drink a glass of formula with/or before every meal.
2. While grocery shopping or walking around a store, aim to finish a serving of formula before you check out or leave.
3. Drink a glass of formula on the way home from work, school or picking up your children.
4. Use a sports bottle to drink formula on the go or at the gym.
5. Substitute formula for other beverages you may have during the day.
6. Add crushed ice to make formula colder.
7. Spread your formula intake throughout the day. This allows your body to absorb all the nutrients consumed.
8. Change the taste of your formula with flavored extracts.
9. Be creative by adding formula to already made low protein food or flavored drinks.
10. Carry a small bottle of mouthwash or toothpaste to freshen your mouth after you finish your formula.

## Getting Satisfaction from Food

Experiencing satisfaction from food is part of being human. Although many foods may be restricted or limited due to PKU, you do not have to compromise on taste.

Identifying the items and tastes that you crave is important. Knowing what you like and choosing foods that make you feel satisfied is part of enjoyable dining. Eating foods that don't satisfy you may leave you wanting more.

The first step is to identify foods that make you feel satisfied. Is it salty, sweet, creamy, spicy, fatty, juicy, crunchy or mushy that you look for in a meal?

### Got a Craving?

- *Add spicy seasoning, cracked pepper, crushed red pepper, or tabasco sauce to steamed vegetables to give ... them the fiery kick you crave.*
- *End your meal with some pineapple or other fruit to get the sweet and tangy taste that signals your body that the meal is done.*
- *Satisfy your sweet and creamy taste buds with some low protein chocolates or marshmallow treats.*
- *Change the texture of a low protein baked item by using applesauce instead of oil to make products lower in fat, soft, and fluffy.*
- *If crunchy is what you want, make your salads snap. Toast low protein bread to make your own croutons. Season them to meet your savory taste needs and toss over greens. 1*

<sup>57</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey : Applied Nutrition Corp. 2007:47.

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Food and eating includes a variety of factors. Not only are taste and smell large factors in the foods we choose, but texture, sight, and our culture play a large role. So, make the most out of your dining experience and keep the zesty seasonings handy. Be creative!

If you are still hungry at the end of your meal, there are many low Phe or Phe free foods that you can eat to satisfy yourself while not going over your Phe limit for the day. Commonly used low Phe foods include: fruits and vegetables, low protein rice or pasta with a little butter or canola oil and seasonings, low protein egg mix or low protein bread. Ask your dietitian for more ideas!<sup>58</sup>

Tips for successful goal setting:

- **Aim for a realistic goal that is right for you.**
  - *For example*, if you are not currently drinking any medical formula it might be best to start with 1/3 or 1/4 of the goal recipe and work your way towards a 100%. Starting small will make sure that you get there!
- **Put in place a goal that is specific.**
  - *For example*, drink 2 cups of formula with breakfast and dinner each day of the week. That is a more specific goal than saying “I will start drinking my formula”.
- **Plan for potential setbacks & reassess/adjust your goals as needed.**
  - *For example*, if you set a goal to completely cut out all high protein foods from your diet and find that the sudden restriction is too drastic it might be more realistic to first start by cutting back on the number servings of high protein food per day.
- **Think short term and long term.**
  - *For example*, a short term goal would be to decrease the amount of Phe you are consuming in your diet. This will get you closer to the ideal Phe intake and may take only a few weeks to reach. A long term goal would be to keep your Phe levels within treatment range. This long term goal may take several months or longer to achieve and maintain.
- **Focus on the process.**
  - *For example*, the long-term outcomes of the goals you’ll be setting are to ultimately help you decrease your Phe levels. During this process you will set short-term goals like drinking the prescribed amount of formula every day. This process will help create behaviors and habits that will help you reach the long-term goal.

## Kuvan® Treatment

If you are an individual that has been on diet since childhood, you may have already been tested for responsiveness to Kuvan® (sapropterin dihydrochloride). If you are responsive, you may have incorporated this into the management of your PKU. If Kuvan is part of your treatment, don’t forget to take it every day. You can use your phone or computer to set up reminders.

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<sup>58</sup>Maltzman, S. : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey : Applied Nutrition Corp. 2007:38.

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For individuals who are returning to diet after an extended time, you may or may not have been tested for responsiveness to Kuvan. See Chapter 2 for information about treatment with Kuvan to support managing your PKU.

## Blood Phe Monitoring

*Goal Phe range: Although a Phe level under 6 mg/dl (360 µmol/L is ideal, levels up to 10 mg/dl or 600 µmol/L are often viewed as acceptable for adults*

Once an individual with PKU is an adult, blood Phe is usually be monitored once or twice per month. Described below are some factors that may alter how frequently you should monitor your blood Phe levels.

AGE AND OTHER FACTORS	RECOMMENDED FREQUENCY OF SAMPLES
Pre-pregnancy and during pregnancy	Levels should be monitored 1-2 times per week. (see pregnancy section for more information)
Illness	Recommendations vary; frequency may be increased during and after illness to ensure consistent Phe levels
Returning to diet	Levels should be obtained after each adjustment in treatment
Physical training	Training may affect Phe levels. If you are concerned, discuss Phe level monitoring with your PKU team.
Dieting for weight control	Dieting may affect Phe levels. If you are concerned, discuss Phe level monitoring with your PKU team.

## Special Considerations in Adulthood

### College<sup>59</sup>

The first few weeks of college will be exciting and full of change. You will meet a variety of new people and have many new experiences. The one thing that remains constant is managing your PKU. Keeping levels in control when you don't have family watching over you may be more difficult than anticipated. Focus! Committing yourself to your PKU treatment and keeping your PHE levels in control is more important than ever.

Long lectures, late nights studying, and cramming for exams all require you to be at your best. Adhering to your treatment regimen, continuing your low protein diet and consuming your formula daily should be a top priority. If you are on Kuvan, you will need to develop a routine that helps you remember to take it daily. Elevated blood Phe levels result in

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<sup>59</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey : Applied Nutrition Corp. 2007:47.

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compromised executive functioning skills such as memory, recall and concentration. Keeping your levels in control, especially before an exam, will help ensure your success in school.

## *Dining on Campus*

Some schools require all first year students to be on a meal plan. Although you may not consume as much as your fellow students, meal time is a great social experience and can lead to many new friendships. If the meal plan is required, speak with the school nutritionist and general manager in the dining department. They may be able to stock and prepare some low protein foods for you.

### ***A Patient's Perspective***

*"My freshman year of college, I think I missed out on a lot of things because I was stressed all the time and so overly emotional [from being off diet]."*

### ***How much Phe is in beer?***

*One 12 fl oz can of beer = about 40mg of Phe!*

*(The darker the beer the higher the Phe.)*

*For Phe content in other alcoholic beverages, look in the low protein food list for PKU.*

Group dining usually offers a wide array of options, including a salad bar stocked with fresh fruits and vegetables. Be sure the people who prepare your food understand the true nature of your PKU and what accommodations you will need if you are not exempt from the meal plan. Maybe a discounted rate is possible so you can still enjoy the social aspect of group dining your first year.

If you are granted an exemption or live off campus, planning your meals is up to you. Choose a day to make a few different items so you spend less time cooking and more time learning. Once again, the key is to be creative and plan ahead.

## *Tips for Drinking Formula on Campus*

- Add powder to an empty water bottle and fill with water later so you can drink your formula when you need it.
- Mix formula in advance and freeze it. It will defrost while you are on campus and you can drink it when you need a cold boost of energy.
- Is your dorm room far from campus? Use a locker at the school gym or student union to keep formula closer to your classes!
- Add formula straight to the bottle of popular store-bought beverages and drink while you are on campus.
- Use the ready to drink formula pouches

## **Alcohol<sup>60</sup>**

There are many decisions you will make as you figure out how to make diet for life manageable for you. One of these choices includes whether or not to consume alcohol, once you are of legal age. Like all other choices involved with becoming an adult, consuming alcohol brings additional responsibility. You should already be aware of the dangers of drinking too much. If you have PKU and you choose to drink alcohol you need to be aware of additional issues involved with drinking.

<sup>59</sup> Maltzman, S, : *My PKU Toolkit A Transition Guide to Adult PKU Management*. New Jersey : Applied Nutrition Corp. 2007:37.

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Some beer and mixers contain significant amounts of phenylalanine. If you choose to consume these types of beverages you need to account for this in your total allotment of phenylalanine for the day. Both alcohol consumption and high phenylalanine levels impair your judgment. Even if you choose alcoholic beverages that are low in phenylalanine, impaired judgment can lead to poor food choices and overeating resulting in elevated blood phenylalanine levels. Moderation is the key.

## Family Planning and Maternal PKU

If you are female and sexually active, you need to use birth control to prevent an unplanned pregnancy. Becoming pregnant while your blood Phe levels are not under strict control could cause serious damage to a developing fetus, causing a variety of heart and brain defects known as Maternal PKU Syndrome. Schedule an appointment with a gynecologist (a doctor who specializes in women's reproductive health) to discuss your PKU condition and birth control options. It is essential to use your birth control exactly as prescribed to ensure that you do not become pregnant until you are ready and able to plan for a safe pregnancy. See Chapter 9 for more on pregnancy and Maternal PKU Syndrome.

## Weight Management

Adjusting eating habits for weight management is not unusual for adults. For an individual with PKU, changing your diet may affect your blood Phe levels. When an individual with PKU goes without food or medical formula, the body starts to break down some of its own stores of protein, resulting in an increase in the individual's blood Phe level.

Before altering your diet, it's very important that you contact your PKU team for information about how you can manage your weight without losing control of your Phe levels. Advice may include:

1. Reduce calorie containing beverages (soda, juice, sports drinks, energy drinks)
2. Increase fruit and vegetable consumption (limit starchy vegetables such as corn, potatoes, peas)
3. Make sure you are getting plenty of formula - protein from formula helps make you feel full. Talk to your dietitian about finding the right formula to limit calories.
4. Eat small, frequent meals
5. Drink plenty of water
6. Limit added sugar (candy, cookies) and fats

Increasing physical activity is a great way to help maintain a healthy weight. Physical activity will help you feel better, give you energy and may help reduce Phe levels.

Here are a few tips to increase physical activity:

1. Start with realistic goals
2. Increase walking by taking the stairs or walking places, get a pedometer
3. Join a gym
4. Have an exercise partner to help keep you on track
5. Join a class like dancing, pilates, yoga, local sports league