

Chapter 7: Ages 13 to 17 Years



A Parent's Perspective

"I wanted to be sure that by high school my daughter did everything herself so by the time she left for college it was old hat and I wouldn't worry because I already stood by her independence through high school and the trials that come with the freedom there."

What to Expect

During adolescence, all parents have to release some control over their teen's life. This is especially true for the parent of a child with PKU.

Teens with PKU report feeling a lack of independence³⁴. Help your teen develop a sense of autonomy by gradually giving up the role of "manager" of your teen's PKU, and move into the role of coach and supervisor. You may find this difficult as you have been continuously told of the important role you have in managing your child's diet, but by helping your child maintain good metabolic control previously, you have already laid the groundwork to ensure that your adolescent will be able to maintain their diet independently³⁵.

By allowing your teen to take charge of PKU, you will give your teen the opportunity to practice PKU self-management and, with your support, prepare to make the transition to adult care while building confidence in his or her abilities. You can help your teen begin the transition to adult care by³⁶:

- Talking to your teen about health care he or she will need in adulthood.
- Trying to find out **how your teen feels** about independently managing the PKU diet and treatment.
- Helping your teen **identify the self-care skills** he or she can do independently
- **Identifying which skills still need to be learned.**
- Talking to your teen about the **warning signs associated with unsafe phenylalanine (Phe) levels.**

Teens, take this quiz to see your level of independence. Strive to achieve checkmarks for all line items - and remember, your parents are always there to help!

- I speak confidently about food restrictions.
- I can tell my friends about PKU and the PKU diet.
- I am comfortable ordering food independently at restaurants and know what kinds of food to look for.
- I can plan my PKU meals independently.
- I can pack a bag of PKU foods to take to school or to a friend's house.
- I know to eat low protein foods and formula before an event to avoid being hungry and overdoing it at a party.
- I can take my blood samples independently.
- I keep track of my diet record independently.
- I speak for myself at my medical appointments.
- I can discuss my feelings with a trusted person.
- If I don't feel like I can manage my feelings, I know I can talk to my PKU team, and they will help me or find someone that can.

³⁴ Jusiene R, Cimbalistiene L, Bieliauskaite R. *Psychological adjustment of children with phenylketonuria* Medicina (Kaunas) 38 (2002) 424-430

³⁵ Gleason, L.A., Michals, K., Matalon, R., Langenberg, P., Kamath, S. *A treatment program for adolescents with phenylketonuria* Clin Pediatr (Phila) 1992;31(6):331-5.

³⁶ University of Washington. Adult Metabolic Transition Project. Available at: <http://depts.washington.edu/transmet/whatis.htm>

Ages 13 to 17 Years

- Helping your teen **create an action plan** to follow when warning signs appear.
- Asking your teen to **take more control** of their medical appointments. This may include creating a list of “things to do” before appointments so your teen can practice getting ready on his or her own.
- Encouraging your teen to **speak for him- or her-self during medical appointments.**
- Working with your clinic team to **find a primary care physician** for your teen as he or she approaches the age of 18.
- **Meeting with the physician** to ensure his or her familiarity with caring for a patient with PKU.
- Talking to your teen about **education and career goals**, and help him or her work through the steps to achieve these goals (for more information about college, see the college section in Chapter 8: 18-Adulthood)
- **Discussing how sticking to diet and treatment will help your teen reach these goals.**

Even as your teen moves into complete adult care, you will still be an important resource. Embrace your new role, and encourage your teen as he or she assumes full responsibility for his or her PKU.

Warning Signs of High Phe Levels

- *Difficulty focusing on tasks (paying attention)*
- *Anxiety*
- *Depression*
- *Inappropriate behavior, mood swings, or confusion about reality*
- *Headaches*
- *Tremors (the shakes)*
- *Eczema*
- *Stiff or weak legs*
- *Agoraphobia (fear of public places)*

A Parent's Perspective

"Allowing her to order in restaurants instead of speaking for her was also tough but once she got the hang of it, we were so proud every time we went out for dinner!"

A Parent's Perspective

"Your PKU child may go from total compliance to throwing caution to the wind at various times in her life. Do not give up on her. Continue to show her that you believe the diet is important by finding ways to demonstrate this. That could mean anything from attending Annual PKU meetings or hosting an event to something as simple as leaving PKU News out on the coffee table. Any little thing you do related to the PKU diet will show her you are continuing to support her even when she seems to be giving up."

"Managing PKU can be difficult but if you are consistent and committed, you can provide your child with the tools he/she needs to manage PKU successfully for a lifetime."

Ages 13 to 17 Years

Development

Developmentally, adolescents continue to gain problem-solving skills and begin to think abstractly. This includes the ability to think through hypothetical situations and imagine outcomes that result from actions, which becomes important as they transition to adult care and long-term planning around PKU management³⁷.

The primary challenge during adolescence is relaxation of the diet due to the perception that there is no immediate harm³⁸. While poor metabolic control is of concern, some research suggests that if PKU has been well-managed earlier in life, the consequences of poorer PKU maintenance at this age are less severe, and deficits can be regained when Phe levels are brought back under control³⁹. However, adolescents with higher Phe levels show reduced abilities in problem solving, reasoning, inhibitory control and other cognitive abilities⁴⁰. For many, going back to diet can be more difficult than remaining on it. Encouraging your child's independent management and dedication to his or her health by pointing out the long-term benefits of sticking with the diet and the long-term consequences of relaxing it is important at this time.

As your teen experiences the changes that come with adolescence, there can be challenges in PKU management. As a parent, your help and guidance is essential to ensure that your teen is ready to make the transition into adult life.

Treatment

For active and growing teens, it is important to consume the prescribed amount of medical formula during the day, as well as eat a balanced diet of low protein fruits and vegetables, and low Phe breads, pastas and rice (find tips on drinking more formula throughout the day in Chapter 8).

Teenagers who plan and prepare their own meals may be more likely to take an interest in eating right. This is especially important for teens with PKU. Keeping his or her own diet

Teens 13-17 years of age, you should work with parents on:

- *Being able to independently prepare formula.*
- *Independently managing total Phe . intake for the day.*
- *Learning menu planning.*
- *Taking responsibility for diet records.*
- *Being able to take blood samples or schedule blood sample at the PKU clinic independently.*
- *Explaining the basics of PKU.*
- *Remembering recent blood Phe levels.*

- University of Washington. PKU and Self Management Timeline Available at: <http://depts.washington.edu/transmet/process/timeline.pdf> Accessed May 18, 2011

³⁷Cherry, K. Formal Operation Stage of Cognitive Development Available at: <http://psychology.about.com/od/piagetstheory/p/formaloperation.htm> Accessed May 18, 2011

³⁸ Bilginsoy, C., Waitzman, N., Leonard, C.O., Ernst, S.L. Living with phenylketonuria : perspectives of patients and their families. *J Inherit Metab Dis.* 2005;28(5) :639-649

³⁹ Waisbren, S. *Neurocognitive Functioning in PKU* Illinois 2010 Presentation

⁴⁰ VanZutphen, K.H., Packman W, Sporri, L., Needham, M.C., Morgan, C., Weisiger, K., Packman, S. *Executive functioning in children and adolescents with phenylketonuria.* *Clin Genet.* 2007; 72(1):13-8.

Ages 13 to 17 Years

record can increase a teen's awareness of eating habits and help him or her take responsibility for diet choices (see chapter 2 for details on how to fill out diet records).

Tips for a Healthy PKU Diet

- **Take the formula** at least three times a day with meals so it's spread throughout the day
- **Eat and count Phe/protein as directed by your PKU team** at meals and snacks
- **Eat plenty of fruit and vegetables** every day, as well as special low protein rice, pasta, bread and crackers based on your tolerance to Phe
- Make sure you're getting **sufficient vitamins and minerals** which comes from your formula. Your team may recommend taking a vitamin or mineral supplement, if needed
- **Drink plenty of water and limit soda, juice and high calorie drinks to 8oz per day**
- Eat the **right sorts of fats such as canola, walnut, olive oil** and limit overall fat intake
- Consume only **moderate amounts of sugar** and of foods containing added sugar

Blood Phe Monitoring

Goal Range: 2-6mg/dL (120-360µmol/L); levels up to 10 mg/dl or 600µmol/L may be permitted in some clinics

Frequency

During the teen years, samples should be taken approximately twice a month, although frequency may vary from time to time. Teens should be responsible for their own blood sampling, although they may still want assistance. This will help develop a sense of self sufficiency and empowerment for a teen with PKU.

Tips for Taking a Blood Sample at this Age

As a parent, all the tips you used for taking samples can now be transitioned to your teenager. Help them to develop good sampling habits with the tips below, and any advice that you may have discovered on your own over the years.

- Have your child set up a reminder on their computer or cell phone with the date marked when blood samples are due
- Encourage your teen to take his or her own sample, but be there to help, especially at first.
- Ask your teen to evaluate if the samples are done properly. If you don't think they are, ask your teen what they think may be wrong with the sample to see if they can identify issues independently.
- Give positive feedback on how well your teen has done.

Teens, here are some tips from other PKU patients that may help you manage your diet independently:

- *Take it in small steps.*
- *Set a goal and work on that for a few weeks. Then add in another goal. Before you know it, it will become a lifestyle and habit and not just checking things off the list.*
- *Use your family and friends to keep you accountable and help you in the process.*
- *Carry good snacks with you to class and sports.*
- *Little things can make a difference . in what you eat and what routine ... you have during the day!*
- *Stiff or weak legs*
- *Agoraphobia (fear of public places)*

Ages 13 to 17 Years

- Have your teen record the results of blood tests when they are available; discuss with him or her any changes in management that might be recommended based on the test result.

Special Considerations for 13-17 Year Olds

Peer Pressure

Adolescence can be a challenging time to maintain treatment due to pressures from peers to conform; social support becomes an essential part of managing PKU treatment⁴¹. Drinking formula and eating a restricted diet may make a teen feel alienated, but it is important for teens not to give in to this pressure to conform. Work with your teen to come up with explanations he or she is comfortable with to explain their diet and formula.

Some teens with PKU start to report psychological disturbances, internalizing disorders (such as depression, anxiety, etc.)⁴² and show slower information processing speeds⁴³. This means that it can take longer for a teen with PKU to comprehend and respond to new information he or she receives. High school academics require higher executive functioning capabilities – including memory, planning, attention and organization – which can also be impacted by PKU⁴⁴. However, there are many strategies that can be introduced to help teens with PKU manage these issues, and teens who maintain metabolic control can reach their full academic potential. Encourage your teen to speak with other teens with PKU and talk to your PKU team if you and your teen need support in maintaining treatment for life.

Social Gatherings

Teens with PKU report feeling socially restricted⁴⁵. Encouraging social interactions may help teens feel less isolated and helping your teen feel comfortable in these social situations may facilitate this.

For teenagers, just like adults, many social gatherings involve food. As they gain independence, teens with PKU begin to navigate more of these situations on their own. As a parent, you can help prepare your teen to handle different situations through encouragement and support of their independence.

A Teen's Patient Perspective

"I was always very open with my classmates, friends, and acquaintances about my diet. When people asked what my formula was, I told them it was my 'milkshake' that I had to drink because I couldn't eat protein. I then rattled off all the foods I couldn't eat and told them that eating protein would cause brain damage. By explaining everything up front, people realized how serious PKU was and respected any special requests I would make. It made it much easier to suggest PKU-friendly restaurants or tell my friends that I had to stop by at my dorm room to take my formula. Every person you meet provides an opportunity to spread awareness for PKU!"

⁴¹Waisbren. S. Neurocognitive Functioning in PKU Illinois 2010 Presentation

⁴²Sullivan, JE. *Emotional outcomes of adolescents and young adults with early and continuously treated phenylketonuria* J. Pediatr. Psychol. 2001; 477-484

⁴³Waisbren. S. Neurocognitive Functioning in PKU Illinois 2010 Presentation

⁴⁴Waisbren. S. Neurocognitive Functioning in PKU Illinois 2010 Presentation

⁴⁵Jusiene R, Cimbalistiene L, Bieliauskaite R. *Psychological adjustment of children with phenylketonuria* Medicina (Kaunas) 38 (2002) 424-430

Ages 13 to 17 Years

- Ask your teen if he or she wants to bring a low Phe food to an event to share with everyone. This ensures that there is at least one “safe” food for your teen to eat.
- Prepare a list of foods that are appropriate for snacks with the Phe content. If your teen is invited somewhere unexpectedly, it will be easy to determine what he or she can have and keep track of how much Phe has been eaten.
- Remind your teen that it is always OK to say no to foods that he or she is offered.
- Ask your teen questions that they may get asked in social situations so they can prepare in advance.
- Let your teen know that they can talk to you about their feelings and how they’re managing their PKU – even if it’s something they think you won’t want to hear.
- Remind your teen that you are a resource for them, and that no matter what, you’re there to help!
- Encourage your teen to share with a friend and find a network of PKU teens to connect with over the internet or through their clinic (see resource list for websites)

Self-Image

All adolescents may feel judged by their peers on how they look. Many feel pressure to be thin or have a “perfect” body. However, it is especially important for teens with PKU to avoid diets for quick weight loss as restricting nutritional intake can force the body to use its own muscle for energy, resulting in unsafe blood Phe levels.

Having a positive self-image will help your teen focus on what is most important – taking care of his or her body to stay strong and healthy. Teenagers who have a positive self-image are more likely to manage their PKU properly, as well as avoid other risky behaviors. You can help your teen work on a positive self-image by reinforcing the tips for teens. If you are concerned that your teen needs additional support, ask your PKU team for suggestions and support group information.

During your teen years, it's normal to feel uncomfortable in your own skin! Developing self esteem is a never ending process. Here are some tips to help you develop and maintain a positive body image from an expert!

- **Be active:** *Regular exercise puts you in a better mood and reduces anxiety. This gives you more confidence in yourself and the way you feel about your body.*
- **Focus on what you do for your body each day, not on how others respond to it:** *You can't control others' responses to you, but you can make healthy choices each day that will leave you feeling good about yourself.*
- **Engage in positive activities:** *Find time every day for a rewarding activity - do some exercise, go for a swim, play a sport, have a walk with a friend, listen to music. . .*
- **Avoid people who give body shape or weight too much importance.**
- **Examine your own self talk and challenge distorted thinking about your body.**
- **Develop many sources of self-esteem:** *So much makes you unique and special beyond your appearance. Develop listening .. skills to be a good friend, practice a skill in sport or the arts or enjoy a good book...*
- **Develop perspective:** *The older you get the less importance people place on judging people by their appearance. It gets better!*
- **Judge yourself as a whole person, not just a body:** *Create a list of people you admire who have contributed to your life, school, community, and the world. Was their appearance important to their success and accomplishments? If their appearance was not important what was?*

-Kearney-Cooke, A. (2003). 'Helping Adolescents Become Strong Adults'. A Project of the Partnership for Gender-Specific Medicine at Columbia University, U.S.A.

Ages 13 to 17 Years

Risky Behaviors

Alcohol

There are many risky behaviors that a teen will be confronted with. While they are not legally able to drink alcohol yet, teens may encounter situations where their friends are drinking during parties. It is important to advise your teen that alcohol contains Phe, and should be avoided. If he or she intends to have a drink, the amount of Phe consumed from alcohol should be accounted for in the overall Phe intake for the day. See chapter 8 for more information about alcohol consumption.

Becoming Sexually Active

Speaking to your teen about the risk of becoming sexually active is important for all parents. This is especially true for parents of young women with PKU. If women with PKU become pregnant while blood PHE levels are not under strict control, serious damage may occur to the developing fetus, resulting in heart and brain defects associated with Maternal PKU Syndrome (see Chapter 9 for more on pregnancy and Maternal PKU Syndrome).

You may not feel ready for your teen to be sexually active - you may not even feel ready to talk to your teen about being sexually active – but it is essential for any young woman with PKU to use birth control if she becomes sexually active.

Schedule an appointment for your teen with a gynecologist to discuss her PKU condition and birth control options. Let your daughter know that you are always available to talk to her about anything, even sexual questions she may have. Some teens won't be comfortable speaking with their parents about sexual topics. To ensure that this doesn't limit the information they are accessing, assure your daughter that she can go by herself to her gynecologist appointment if that makes her more comfortable. Also, reassure her that you are always there to talk to her without judgment.



Teens, test your knowledge.

Did you know:

- Girls can get pregnant before their first period.*
- Birth control pills do not protect you from sexually transmitted disease (STDs).*
- Some medications (like antibiotics) can make birth control pills not work effectively.*
- Condoms do not protect against pregnancy and STDs 100% of the time.*
- Abstaining from sex is the most effective method of birth control and STD prevention.*
- If a woman with PKU gets pregnant with high PHE levels, the baby can be born with serious damage, including brain and heart defects (see Chapter 9 for more information about this and how a PKU woman can have a healthy baby).*