



Chapter 5: Ages 3 to 6 Years

A Parent's Perspective

"...Zachary and Rachel...are fine. You would never know they have PKU unless you saw us weighing the French fries or heard us request a happy meal with salad (no cheese, please) instead of a hamburger." - Parent of two children with PKU

What to Expect

Parenting can be stressful for anyone, but parenting a child with a chronic disorder such as phenylketonuria (PKU) can be especially stressful and may negatively impact the parent's "mental health-related quality of life"²². Decreased emotional support – such as what you would get from supportive friends and family – has been reported to decrease a parent's sense of mental health-related quality of life. However, parents also report that as their child with PKU gets older and they feel development has stabilized, there is an increased sense of mental health-related quality of life. Still, there will be times when you will feel stress, such as when you're getting ready for preschool and kindergarten. This is a time of anticipation and, if it is the first time your child will spend long periods of time out of your care, some concern.

A Parent's Quick Tip

Try to network with other PKU families in your region. Even if you have to travel a few hours to get together, it's worth it to socialize with these families. You and your child will both benefit from the interaction.

Your health and sense of well being is important, and may relate to how you parent your child or children. It is important that you keep aware of the emotional support you are receiving, and if you find yourself feeling in need of additional support, vocalize it. Seek support from your friends, family and other parents of children with PKU. Your PKU team can also help with support groups, individual counseling and even match you up with other parents of children with PKU, if you haven't already been introduced¹. Please see Chapter 15 for website information on accessing state and regional support groups.

Development

While most children begin to develop a strong desire for independence at about three years of age, they still have very little self-control²³. In order to maintain proper diet and treatment, expectations for a child's ability to manage PKU need to remain age-appropriate. You play a key role in delivering much of the day-to-day care for managing your child's PKU. However, involving your child in management of his or her PKU will help to empower your child with the ability to make decisions about treatment as he or she gets older and can manage treatment independently. This will hopefully improve long-term adherence to the PKU diet²⁴.

²² ten Hoedt AE, Maurice-Stam H, Boelen CAC, Rubio-Gozalbo ME, van Spronsen FJ, Wijburg FA, Bosch AM, Grootenhuys MA. *Parenting a child with phenylketonuria or galactosemia: implications for health-related quality of life* J Inherit Metab Dis 2011:391-8

²³ Waisbren, S. THE PSYCHOLOGY OF PKU and ALLIED DISORDERS (AND THE BOSTON MARATHON) National PKU Alliance Annual Parent Meeting, November 2009 Presentation

²⁴ van Spronsen, F.J., de Groot, M.J., Hoeksma, M., Reijngoud, D.J., van Rijn, M., J Inherit Metab Dis (2010) 33:671–676

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This is a critical age, as children who maintain proper metabolic control at this age have much better health and developmental outcomes than those who do not. From four to six

years, children have little issue following the PKU diet, as rules around foods and medical formula are clear and concrete. Socially, children with PKU develop normally, although some may experience separation anxiety²⁵. Developmental progress is typically normal and children with well-controlled PKU can be expected to function normally in a kindergarten setting.

Talk to your PKU team if you have any questions about your child's development.

Talking with Your Child About PKU

While your child is not old enough to manage PKU alone, it is valuable for your child to begin to better understand his or her diet and treatment. When speaking with your child about PKU, the following tips may be helpful:

- *Use simple examples to explain ideas.* For example, when explaining to your child why the PKU diet is important, it may be helpful to relate the diet to that of a food allergy. See the resources in Chapter 15 or speak with your PKU team who will also have information about books to read to your child to help him or her understand special diets.
- *Let your child know she can say “no”.* If you haven't already, teach your child to ask you before eating unfamiliar foods, and that it is OK to say no to anyone who offers food that is unfamiliar or off limits.
- *Tell your child it isn't his or her fault.* A child may not understand why he or she has PKU when others do not, and may think that he or she did something to 'deserve' it. Explain to your child that everyone is born with different qualities – such as hair color and eye color – and PKU is something that people are born with, not something that anyone causes. The book *Everybody Has Something* may help show your child that everyone is different with their own unique challenges. It can be ordered at http://www.ucdenver.edu/academics/colleges/medicalschoo/department/pediatrics/subs/genetics/clinical/IMDNutrition/Documents/Everybody_Has_Something_Order_Form.pdf.

With your help, a child of this age usually:

- *Is aware that he or she has a special diet*
- *Begins to learn how to deal with other children's curiosity about PKU*
- *Knows that he or she needs to take medical formula*
- *Begins to watch you prepare the formula ... and by 6, can assist in formula preparation*
- *Knows that he or she needs to have blood tests*
- *Knows to check new foods with parents*
- *Begins to learn 'yes/no', 'red, yellow, green traffic light' or 'low PHE/high PHE' foods*
- *Is aware that the PHE in food is counted*

How do I talk to my child about medical formula?

Treat the medical formula as special – as an 'energy drink' that will make your child 'tall and strong'. Give the medical formula a name, such as 'special milk'. Give lots of positive prompts and praise – but let your child know that drinking the medical formula is something she or he must do.

²⁵ Waisbren, S. THE PSYCHOLOGY OF PKU and ALLIED DISORDERS (AND THE BOSTON MARATHON) National PKU Alliance Annual Parent Meeting, November 2009 Presentation

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- *Stay positive.* Sending the right message about foods and treatment is important. It is better to talk about off-limit foods as “high Phe,” “no,” “red” or “stop” foods (see the next section on the Food Traffic Light), rather than “bad” or “naughty” foods²⁶. In the same way, avoid talking about blood sampling in negative ways, such as calling yourself or another parent “mean” for taking a blood sample. Diet and blood sampling are part of life and staying positive will help your child accept and manage PKU as he or she grows. Never say anything negative about the food or formula to your child.
- *Begin to include your child in preparing food.* Empower your child from an early age by including him or her in preparing meals. It may start with something as minor as mixing the formula at 3 and identifying which food should be included in a meal by 6 years.
- *Let your child measure and weigh their food.* Children are naturally curious. Show your child what you’re doing when you’re weighing and measuring the food you are preparing and help her or him use these tools. Explain how it works and slowly, see if he or she can weigh or measure the food for you, with your assistance.
- *Get your child to help count Phe.* As your child begins to learn to count, he or she can begin to help with recording his or her Phe intake. You can make it into a counting game, and even a memory game about what he or she has eaten. You can use a whiteboard or laminated paper to create a chart to count the Phe eaten at each meal. Write the total amount of milligrams/exchanges your child can have each day at the top of the chart, and list meals under this amount. Next to each meal, write the milligrams/exchanges planned for that meal. As the milligrams/exchanges are eaten during the day, your child can cross them off.

A Parent's Quick Tip

"Don't approach the diet as a burden (though some days it is, I know) -- think of it as a special part of the day where you get to teach your child about something very important and build trust in you that you are always there to help!"



Treatment and Diet

Possible Challenges with the PKU treatment

A Parent's Perspective

"PKU has really worked into our life and then it just became a part of it all in a way where I (as a mom and the meal planner/preparer of our house) almost don't even 'think' about it anymore! It gets better, it really does!"

Children at this age are developing a growing sense of independence and sometimes try to gain control over their lives by challenging rules and limits their parents set. For children with PKU, this may include objecting to their diet or treatment.

By allowing your child to have a role in some of the decision making process, he or she will feel like they have more control of their PKU. Above all, it is

important to stay positive and matter-of-fact about your child’s diet and treatment.

²⁶ Kids Health Talking to Your Child About Diabetes. Available at: http://kidshealth.org/parent/diabetes_center/living_diabetes/talking_diabetes.html# Accessed May 17, 2011

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KINDS OF CHOICES YOU CAN OFFER
“Do you want to sit on the chair or on my lap to take your sample?” or “Which finger should we use for your sample?”
“Would you like carrots or apple for your snack?”
“Do you want to drink your special milk in the blue cup or red cup?”

The 3 Kinds of Food: Traffic Lights

Many parents use the Traffic Lights example to teach children about the PKU diet. To create a Traffic Light, draw three circles resembling traffic lights on a large poster board or piece of paper and color them green, yellow, and red. Then cut out pictures of many different types of foods from magazines or websites.

To start the game, explain to your child that there are three kinds of foods: “red”, “yellow”, and “green”. These colors are defined as:

- Green = foods that are low in protein/Phe
- Yellow = foods that are only OK in limited quantities
- Red = foods that are high in protein and not on the PKU diet

Work with your child to organize foods according to each color. Once your child understands these ideas, you can create Traffic Light games to improve or test knowledge. New food pictures can create a fun challenge for your child as he or she figures out where each new food belongs on the traffic light, or you can test your child’s recognition by placing a “red” food on the green light, or vice versa, and asking which of the foods does not belong.

The Traffic Light may also help you talk about diet choices with your child. Children familiar with the Traffic Light will readily understand what a “green” food or “red” food is, and this offers a way for parents to say no to foods without using the word “no” constantly. Some parents may also choose to refer to foods as “low Phe” and “high Phe” foods for this same reason.

Medical Formula

Medical formula is important for growth and development and keeping blood Phe in the safe range. Your child should drink all the medical formula prescribed each day. Talk to your PKU team if your child does not drink the prescribed amount of medical formula. Some trouble-shooting tips for formula drinking are below.

If...	YOU CAN TRY...
Your child seems overwhelmed by the amount of formula she needs to drink at each sitting.	Dividing the amount of medical formula your child needs to drink into three to four smaller servings that will be easier to finish.
Your child does not want to drink the formula during a meal.	Offering medical formula at the beginning of mealtime when your child is hungry.

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Your child does not have an appetite for the medical formula.	Cutting back on other liquids or snacks and offering formula first when your child is hungry or thirsty.
Your child usually likes the medical formula but seems bored with it.	Freezing the formula, then blending it and serving with a spoon for eating. Or serve it in a new cup he or she is excited to use.
Your child doesn't seem to like the taste of the medical formula.	Talking to your PKU team about how to flavor the formula or trying other brands or flavors of formula.

Tips for making mealtime easier

Try to keep family meals and PKU meals similar.

A child with PKU may find it difficult to understand why she can't eat the same foods as others; as a result, she may refuse to eat her food. If possible, make your child's meals similar to the family meal. For example, if the family is having beef stir-fry and rice, serve your child stir-fried vegetables with low protein rice. Your child may want to eat foods that are not appropriate for her PKU diet; educate your child about which foods she can have and support positive food choices.

A Parent's Quick Tip

Order pasta from more than one company and offer your child different brands regularly. Sometimes a certain brand will be on back order when you need pasta, so you want your child to have a taste for more than one brand.

Create a pleasant mealtime environment.

Parents and siblings can be good role models for young children. Eat meals together as often as possible. Try to limit distractions such as TV during mealtime. A relaxed atmosphere during mealtimes can help make this time enjoyable for the family.

Avoid battles over meals.

Parents often become anxious when their children don't eat, and children quickly pick up on this. It can happen especially when you have made a lot of effort to prepare meals for your child with PKU. Some children refuse to eat, knowing it is an effective way to gain attention. It is never a good idea to force-feed a child. This often leads to fear of mealtimes and further refusal of food. Continue to offer new foods multiple times; children may need to see a new food many times before they will try it. Praise your child for trying new foods.

Educate your child about which foods he or she can have.

Your child may want to eat foods that are not appropriate for her PKU diet; educate your child about which foods he or she can have and support positive food choices. Another way to set a good example is to encourage family members to eat plenty of fruits and vegetables.

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Blood Phe Monitoring

Goal Phe Range: 2-6 mg/dl (12-360 umol/L)

Frequency

At this age you should check your child's Phe level every 2 weeks, unless directed otherwise by your clinic.

Tips for Taking a Blood Sample at this Age

Preschoolers and early school-age children are old enough to become more involved in taking blood samples. For example, you can let your child choose where he or she wants to sit when a blood sample is being taken or to choose which finger will be used to take the sample. As with earlier ages, following a routine for blood sample taking can help children know what to expect, which can reduce fear around blood sampling. Other tips for taking blood samples at this age:

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"[My daughter] has made up a little dance she does to shake the blood down and she gets all the supplies ready herself. I wouldn't say this is something she necessarily likes but she accepts it and is a part of it and on we go!"

- Have a calendar with the date marked when blood samples are taken
- Let your child choose which finger to use for the blood sample.
- Encourage your child to help you press the button on the lancet or diabetic pen. Your child can put a finger over your finger and press on the count of three.
- Ask your child to help you count out the blood drops onto the filter paper.
- Ask the child to help "finger paint" the circle(s).
- Explain what a correct sample looks like, and encourage your child to see how well he or she has done in producing a correct sample.
- Give lots of positive feedback on how well your child has done.
- When your child is old enough to write his or her name, have your child help you fill it out on the filter paper.

A Quick Tip

Topical 'numbing agents' such as Pain Ease[®] may help reduce the pain of the finger stick. Ask your PKU team if this is an option to use.

Overall, try to maintain a consistent schedule and matter-of-fact manner about blood sample testing. Blood tests are a way of life for children with PKU, and the patterns you set when your child is young will help create the foundation for how he or she manages PKU throughout life.

Special Considerations

Explaining PKU to Friends

Your child's friends can be a source of support. Although his or her friends cannot tell that your child has PKU at first glance, they may notice his or her special diet and ask questions. Helping your child find ways to explain PKU to friends can help him or her adjust to new situations and help build his or her acceptance for this condition. Your child can share simple information or give a more detailed explanation, but being direct and honest may help other children clearly understand PKU. The book *Everybody Has Something* may also help

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show your child's friends that everyone is different with their own unique challenges. It can be ordered at http://www.ucdenver.edu/academics/colleges/medicalschoo/department/pediatrics/subs/genetics/clinical/IMDNutrition/Documents/Everybody_Has_Something_Order_Form.pdf

Listed below are some helpful explanations of PKU:

- No one can “catch” PKU. People are born with PKU, like I was.
- Having PKU means I have to eat foods that keep me healthy.
- I can't eat some kinds of foods, like meat or cheese, or they will make me sick. I won't get sick right away, but if I eat these foods, my body and brain won't grow the way they are supposed to.
- I drink a special milk which is like a vitamin drink. It gives my body good things that come from foods I can't eat.

Offer encouragement to your child by:

- Giving positive messages about the foods he or she can have
- Reinforcing to your child that he or she is special, and that this special way of eating is to keep him or her stay healthy.
- Talking to your child about other people you know who are on a special diet, even if they are adults, so your child knows that he or she is not the only one on a special diet.
- Discouraging negative comments about the taste of the medical formula and the low protein foods from any family members.

See the resources in Chapter 15 for examples of questions your child may be asked and how he or she can respond.

What should I do if my child doesn't want to tell friends?

It is important that friends know about your child's PKU so that they understand or can help offer support in social situations. If your child doesn't want to tell friends herself, you can discuss with her the idea of inviting friends over so that you can help explain PKU. Some parents also provide a presentation to their child's class, offering classmates the chance to ask questions and see the 'tools' of PKU treatment, such as formula and blood sampling material.