

PKU AND PREGNANCY

A very special time in life

Preconception

Congratulations on taking the first step to having a baby.

The journey to motherhood begins with your metabolic clinic. It is essential to work with your clinic before, during and even after pregnancy. Only your clinic can provide the best guidance to ensure the best outcome for you and your baby. They will work with you to ensure well-controlled blood phenylalanine (PHE) levels prior to pregnancy and throughout pregnancy.

Studies in maternal PKU (MPKU) have shown uncontrolled blood PHE levels during pregnancy can cause the following outcomes for the baby:

- Microcephaly (small head size)
- Intellectual disability
- Behavioral issues
- Congenital heart defects (CHD)
- Low birth weight and reduced length for age

Keeping your PHE levels within recommended range will help protect your baby from these risks.

Speak with your metabolic healthcare professional regarding recommended PHE levels during pregnancy.

This booklet provides tips for your entire pregnancy. However, always follow your clinic's guidance first.

Steps to getting PHE levels in the recommended range:

Re-commit yourself to the low PHE diet

- Find low-PHE recipes at [MedicalFood.com/Recipes](https://www.MedicalFood.com/Recipes)
- Explore vegan recipes, however skip the plant-based protein ingredients such as legumes, soy, tofu, etc.
- Change regular bread, rice and pasta to low protein versions, such as Nutricia's Loprofin.

Find a PKU formula you enjoy

- Nutricia offers the widest range of PKU formula options. You are sure to find one that meets your personal taste and needs* (see page 15).
- Form a plan and stick to it - this will help to make sure you take all your prescribed formula each day.

Make a new routine

- Regular blood samples (blood spots) will let you know when your PHE levels are in the "safe-range" for pregnancy. Set a reminder on your calendar or cellphone to send your blood spots to your clinic.
- Meal planning will help make food shopping and new dining habits easier.

Get Connected

- Join an online Maternal PKU group to share, talk and get to know other women with PKU who are pregnant, planning to become pregnant or have tips to share from their past pregnancy.
- Learn more about the National PKU Alliance (NPKUA) Maternal PKU Mentor program, AdultsWithPKU.org/Maternal-PKU

1st trimester

In this trimester, you will gain anywhere from 1-5 pounds.
Your baby will grow from the size of a tiny poppy seed to a lemon.

Some women report feeling nauseous during their 1st trimester and having difficulty keeping food down. Despite these challenges it is extremely important to drink your formula as it provides PKU friendly protein and nutrition, which is essential for you and your baby.

If nausea and vomiting strikes, here are some tips to help:

- Try eating with just the foods you enjoy
- Cold foods are sometimes better tolerated than hot foods
- Avoid an empty stomach
 - Try small frequent snacks every 2 hours during the day.
- Ask your metabolic dietitian if a lower volume PKU formula is appropriate
- Divide current PKU formula into small frequent portions throughout the day
- Ask your metabolic dietitian if you can try a PKU formula with less of a smell
 - Or drink current PKU formula through a straw from a container with a cover to prevent smelling PKU formula while drinking.
- Avoid taking any vitamin/mineral supplement on an empty stomach

Some women also report constipation in their 1st trimester. Here are some tips to help:

- Focus on fruits and vegetables that are high in fiber and low in PHE
- Be sure you are drinking enough fluid

DO NOT WAIT. Make your clinic aware if nausea, vomiting or constipation does not improve.





2nd trimester

Depending on your pre-pregnancy weight, you will now start gaining about 0.5-1 pound per week. In the 2nd trimester your baby will grow from the size of a peach to an acorn squash.

During this time period, you may notice the return of your appetite and a desire to eat more. It is still extremely important to keep your PHE levels in check, so be sure to work with your metabolic dietitian and have a meal plan in place.

If hunger strikes go for your formula first.

You may also benefit from preparing portioned out low-PHE snacks for the week:

- Make single-serving bags of low protein pretzels, popcorn and cereal
 - Consider gluten-free* items
- Fruit leathers or fresh fruit
- Sliced vegetables such as cucumbers, peppers, zucchini, etc.
- Portioned out cherry or grape tomatoes or baby carrots
- Prepare ahead and freeze bulk items such as low protein breads, biscuits, muffins, cookies and soups

Get Ready!

During this stage you may have an increase in your daily PHE tolerance, which means increased food options. Your dietitian will work closely with you and monitor your blood PHE, but remember PKU formula is still your main source of PKU -friendly protein for both you and your rapidly growing baby.

*Be sure to read labels and check serving sizes when planning a snack. PHE content varies by brand. Not all gluten-free foods are low in protein. For more information on finding the PHE content in common foods, check out www.HowMuchPHE.org (English) or TyrophED.com (French Canadian)

3rd trimester

Depending on your pre-pregnancy weight, you will have gained roughly 11-40 pounds by the end of your pregnancy. In your 3rd trimester your baby will grow from the size of a large butternut squash to a small watermelon

Now you are getting closer to the exciting day. As your baby grows you may experience reflux, heartburn or feel full quickly.

If heartburn strikes, here are some tips to help:

- Avoid – caffeine, peppermint or spearmint, sodas, spicy foods and acidic foods
- Avoid large meals
 - Try small frequent meals spread throughout the day
- Limit meals close to bedtime
- Sleep with your head at elevated angle
- Consider ginger candy or tea

You have been working really hard on your diet. Set a plan to help you stay on diet once the baby arrives. **Here are some suggestions:**

- Teach your significant other, family and friends how to prepare your formula so they can help in the first few weeks.
- Share your favorite recipes with family and friends so they can prepare some meals for you.
- As you begin “nesting” cook some meals low in protein in advance and freeze for easy reheating.

Almost there! Keep sending in your blood spots.



Now your baby is here!

You now have your new wonderful bundle of joy. You worked really hard on your diet to get to this point. Don't give up, you have another person to take care of now, and being your best means staying on diet.

As you know, going off diet could mean an increase in your PHE and a decrease in your tyrosine (TYR) blood levels.

PHE build-up (high levels) can cause:

- Difficulty concentrating
- Feeling tired, moody or in a fog
- Tremors in hands, fingers, feet or legs
- Poor memory, short term recall or general forgetfulness
- Trouble focusing at work or school
- Difficulty following directions and problem solving
- Anxiety, phobias or depression

Taking care of a newborn is tough work. Be sure to keep formula as part of your daily routine. Here are some tips:

- Set an alarm for several times each day to drink your formula
- Have your significant other, family or friends be in-charge of preparing 24 hour batches of your formula
- Make feeding time an opportunity for both of you to have your formula (or feeding)
- Turn your formula into a fruit smoothie to start your day



Staying on diet is important for both you and your baby!

Please seek help from a healthcare professional if you start to feel down or overwhelmed.

A close-up photograph of a woman with long brown hair, smiling and looking down at her baby. She is breastfeeding the baby, who is wearing a white onesie. The background is softly blurred, showing what appears to be a hospital room setting. The image is partially covered by a large, light blue and purple watercolor-style graphic on the right side.

Breastfeeding

You can breastfeed your baby just like any other mother. For every nursing mom, calories play an important role in the ability to produce breastmilk. Your dietitian will review how your PHE intake and your diet overall may change with breastfeeding.

Here are some tips to help when more calories are needed:

- Mix oil or non-dairy margarine into low protein pasta or rice
- If your dietitian feels it's appropriate, change from a low calorie PKU formula to a higher calorie PKU formula containing fat
- Dip low protein bread in olive oil
- Add avocado to salads or sandwiches
- Add sweets to teas, coffees, muffins and smoothies
 - Maple syrup, honey, agave nectar, sugar

Be sure to continue checking your blood levels regularly!

Low protein cooking

Planning ahead, making grocery lists and exploring vegetable-based recipes will help you keep control of your PHE levels.

Here are two quick and delicious recipes low in protein.

Zucchini Pizza Bites

1 XL zucchini (400 g)
1 tsp salt
Olive oil spray
¼ cup tomato sauce
½ cup imitation low protein cheese

Directions:

1. Preheat oven to 450° F (230° C)
2. Cut zucchini into ½ inch slices
3. Grease baking sheet with olive oil and place zucchini slices on sheet
4. Bake for 8 minutes on each side or until lightly browned
5. Spoon tomato sauce onto each slice
6. Top each slice with imitation cheese
7. Return to oven for an additional 2 minutes

1 pizza bite (70 g)

Protein **0.8 g** PHE **30 mg**



Shop **MedicalFood.com**
for low protein pasta,
baking mix and more

Jicama Fries

1 jicama (1 lb), peeled, cut into ½ inch thick fries
1 tablespoon olive oil
½ teaspoon coarse salt
½ teaspoon garlic powder
½ teaspoon paprika
¼ teaspoon ground black pepper

Directions:

1. Preheat oven to 400° F (205° C).
2. Line a cookie sheet with foil and fit it with a cooling rack
3. Bring water to a boil in medium saucepan on medium heat
4. Add jicama and cook for 8 minutes until jicama is less crunchy
5. Drain water, using a colander
6. In a large bowl, toss the jicama fries with the olive oil and the spices.
7. Arrange on the cooling/booking rack.
8. Bake 45 minutes, until browned. Serve immediately

Prep Tip: To microwave instead of boiling on stove-top, place the jicama strips in a microwave-safe bowl. Add 2 tablespoons of water. Cover and microwave for 15 minutes, stopping once to gently stir.

Tip: Blend with your personal favorite flavors, herbs and spices for variety.

Makes 2 servings (170 g)

Protein **1 g** PHE **29 mg**



Find more recipes at **MedicalFood.com**

Nutricia products

Find the Right PKU Product for You




Every person is different and has different needs – especially during pregnancy. Taste, smell and volume may be a challenge at times. Nutricia provides an extensive selection of PKU formulas to help during this critical time. We also provide a selection of low protein foods.

All PKU formulas should be used only under medical supervision and managed by a healthcare professional. Always speak to your metabolic healthcare professional prior to making any changes in your PKU diet plan.






Visit www.MedicalFood.com for more information.



Looking for a higher calorie PKU formula?

	Product Name	Description
	PhenylAde® GMP Drink Mix	<ul style="list-style-type: none"> • 2 flavors (Original and Vanilla) • GMP-based • Added DHA • 10 g PE (15.3 mg PHE) / 132 kcal (180 mL, 6 fl oz)
	PhenylAde® Essential Drink Mix	<ul style="list-style-type: none"> • 5 flavors (Unflavored, Vanilla, Chocolate, Orange Creme and Strawberry) • 10 g PE / 157 kcal (240 mL, 8 fl oz)
	Periflex® LQ	<ul style="list-style-type: none"> • 2 flavors (Orange Creme and Berry Creme) • Ready-to-drink • 10 g PE / 107 kcal (15 g PE & 160 kcal per 250 mL, 8.5 fl oz container)
	Periflex® Advance	<ul style="list-style-type: none"> • 2 flavors (Unflavored and Orange) • 10 g PE / 110 kcal (120 mL, 4 fl oz)

Looking for a lower volume PKU formula?

	Product Name	Description
	PhenylAde® 60 Drink Mix	<ul style="list-style-type: none"> • 2 flavors (Unflavored and Vanilla) • 10 g PE / 49 kcal (90 mL, 3 fl oz)
	Lophlex® LQ	<ul style="list-style-type: none"> • 3 flavors (Juicy Orange, Mixed Berry Blast, Juicy Tropical) • Juice-based • Added DHA • 10 g PE / 60 kcal (20 g PE & 120 kcal per 125 mL, 4.2 fl oz pouch)
	XPhe Maxamum®	<ul style="list-style-type: none"> • 2 flavors (Unflavored and Orange) • 10 g PE / 76 kcal (120 mL, 4 fl oz)


Is the smell or taste of the PKU formula a problem?

	Product Name	Description
	PhenylAde® GMP Mix-In	<ul style="list-style-type: none">• Mix into any liquid or semi-solid food without significant flavor change• GMP-based• 10 g PE (15.3 mg PHE) / 42 kcal*
	PhenylAde® Amino Acid Blends (MTE)	<ul style="list-style-type: none">• Mixes into any flavored liquid (except water)• 10 g PE / 40 kcal*

Please note when using these products sufficient fat and energy intake from other sources are essential; these products must be combined with an appropriate vitamin & mineral supplement when taken as the sole source of PKU formula.

*When mixed with water, other drinks may alter caloric and PHE content

Is drinking PKU formula a problem?

	Product Name	Description
	Phlexy-10® Tablets*	<ul style="list-style-type: none">• 12 tablets = 10 g PE / 49 kcal

Please note when using this product sufficient fat and energy intake from other sources are essential; this product must be combined with an appropriate vitamin & mineral supplement when taken as the sole source of PKU medical food.

*Available in the US only



WANT TO TRY
SOMETHING NEW?

3 EASY WAYS TO REQUEST A SAMPLE

1. Talk to your metabolic clinic
2. Request online at [MedicalFood.com](https://www.MedicalFood.com)
3. Call Nutricia at 800-605-0410



*One-on-one support to obtain formula coverage at no cost to you!**

Don't face the challenges of formula coverage alone. Through the Nutricia Connect program, our formula coverage specialists are here to help you through each step to get formula coverage. Our experts will:

- Complete pre-determinations, prior authorizations, medical necessity reviews and help find in-network suppliers to fulfill orders
- Verify eligibility and benefits for PKU formula through pharmacy or other medical plans
- Work with families and their employers to find a coverage exception in order to accommodate Nutricia PKU formulas
- Assist in appealing denials for coverage and help resolve billing discrepancies

NUTRICIA IS HERE
TO LEND A HELPING HAND

Talk to a Nutricia Formula Coverage Specialist
Coverage@Nutricia.com

9 out of 10
people
successfully
obtain formula
coverage

To learn more visit
MedicalFood.com/Reimbursement or call **1-800-605-0410**
Monday–Friday 8:30a.m. – 5:00p.m. ET.



**NUTRICIA
CONNECT™**
Formula Coverage • Education • Support

Who's who (contact details)

My dietitian

Name:

Phone #:

Email:

My nurse

Name:

Phone #:

Email:.....

My doctor

Name:

Phone #:

Email:.....

Notes

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Your family journey is about to begin... Nutricia is here to support you along the way.

"This is the hardest and most rewarding journey in a PKU woman's life and worth every second of all the hard work we put into our pregnancies."

Lisa Marie , Classical PKU, Age 36, Northern NJ

"Finally gave birth to my beautiful and healthy baby girl Claire Marie (7 lbs 2 oz). I am so proud of myself and what I've accomplished with my diet and pregnancy. Now time to enjoy this little lucky charm of mine! – feeling blessed."

Leeanne, Classical PKU, Age 28, Tampa, FL

Need additional information or support
about returning to the PKU diet?

Email us at DietForLife@Nutricia.com

