

# 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 be sure you always follow your clinic's guidance.*

Steps to getting PHE levels in the recommended range:

## Re-commit yourself to the low PHE diet

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

## Find a PKU formula you enjoy

- Nutricia offers the widest selection of great tasting formulas to help meet 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 cell phone to complete your blood spots.
- 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 from a past pregnancy.
- Learn more about the National PKU Alliance (NPKUA) Maternal PKU Mentor program: [AdultsWithPKU.org/Maternal-PKU](https://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; 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. Your baby will grow from the size of a peach to an acorn squash.

In the 2nd trimester, you may notice the return of your appetite and a desire to eat more. It is still extremely important to keep your PHE levels well controlled. Be sure to work with your metabolic dietitian and have a meal plan in place.

**When 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 [HowMuchPHE.org](https://www.howmuchphe.org) (English) or [TyrophED.com](https://www.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.

## High PHE 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 woman with dark hair is lying on a couch, breastfeeding her baby. She is wearing a striped tank top. The baby is wearing a white long-sleeved shirt. The background is a bright, airy room with wooden shelves and a large window. There are decorative watercolor splashes in purple and blue on the right side of the page.

# 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 (be sure to count the additional PHE!)
- 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 extra-large zucchini (400 g)
- 1 tsp salt
- Olive oil spray
- ½ cup (123 g) tomato sauce
- ½ cup (45 g) low protein, dairy-alternative 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. Sprinkle with salt.
4. Bake for 8 minutes on each side or until lightly browned.
5. Spoon tomato sauce onto each slice.
6. Top each slice with dairy-alternative cheese.
7. Return to oven for an additional 2 minutes.

Makes 10 Pizza Bites.

Protein per Bite: **0.7 g**      PHE per Bite: **30 mg**

## Jicama Fries

- 1 medium jicama (660 g or 1 1/2 lbs.), peeled, cut into ½ inch thick fries
- 2 tablespoons 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 baking sheet with foil.
3. Bring 1 quart (950 mL) of water to a boil in medium saucepan.
4. Add jicama and cook for 8 minutes until jicama is less crunchy.
5. Drain jicama, using a colander.
6. In a large bowl, toss the jicama fries with the olive oil and seasonings .
7. Arrange the jicama on the baking sheet.
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 4 servings.

Protein per serving: **1.3 g**      PHE per serving: **32 mg**



Visit [NutriciaMetabolics.com](https://www.nutriciametabolics.com) for pasta, baking mix and other low protein options.

\*Calories and protein may vary by product. Be sure to check the nutrition label.



Find more recipes at [NutriciaMetabolics.com](https://www.nutriciametabolics.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 challenging 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 must be used only under medical supervision and managed by a healthcare professional. Always speak to your metabolic healthcare professional prior to making any changes to your PKU diet plan.





Visit [NutriciaMetabolics.com](https://www.nutriciametabolics.com) for more information.

## Looking for a higher calorie PKU formula?

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


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

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

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; mixing with other drinks may alter calorie and PHE content

## Is drinking PKU formula a problem?

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

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



Looking for a ready-to-drink PKU formula?

	Product Name	Description
	PhenylAde® GMP READY	<ul style="list-style-type: none"><li>• Plain flavor</li><li>• GMP-based</li><li>• Added DHA &amp; EPA</li><li>• 10 g PE (18 mg PHE) / 110 kcal (250 mL, 8.5 fl oz)</li></ul>
	Periflex® LQ	<ul style="list-style-type: none"><li>• 2 flavors (Orange Creme and Berry Creme)</li><li>• Ready-to-drink</li><li>• 10 g PE / 107 kcal (15 g PE &amp; 160 kcal per 250 mL, 8.5 fl oz container)</li></ul>
	Lophlex® LQ	<ul style="list-style-type: none"><li>• 3 flavors (Juicy Orange, Mixed Berry Blast, Juicy Tropical)</li><li>• Juice-based</li><li>• Added DHA</li><li>• 10 g PE / 60 kcal (20 g PE &amp; 120 kcal per 125 mL, 4.2 fl oz pouch)</li></ul>

Looking for a lower volume PKU formula?

	Product Name	Description
	PhenylAde® GMP ULTRA	<ul style="list-style-type: none"><li>• 2 flavors (Vanilla and Lemonade)</li><li>• GMP-based</li><li>• 20 g PE / 98 calories Lemonade (180 mL, 6 fl oz)</li><li>• 20 g PE / 107 calories Vanilla (180 mL, 6 fl oz)</li></ul>
	PhenylAde® 60 Drink Mix	<ul style="list-style-type: none"><li>• 2 flavors (Unflavored and Vanilla)</li><li>• 10 g PE / 49 kcal (90 mL, 3 fl oz)</li></ul>
	XPhe Maxamum®	<ul style="list-style-type: none"><li>• 2 flavors (Unflavored and Orange)</li><li>• 10 g PE / 76 kcal (120 mL, 4 fl oz)</li></ul>



WANT TO TRY SOMETHING NEW?

3 EASY WAYS TO REQUEST A SAMPLE

- 1. Talk to your metabolic clinic
- 2. Request online at [NutriciaMetabolics.com](https://www.NutriciaMetabolics.com)
- 3. Call Nutricia at 800-365-7354





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

Don't face the challenges of formula coverage alone. As part of our Nutricia Connect program, we offer Nutricia Navigator, a free service available to those living in the U.S. to help guide you through the world of formula coverage from start to finish.

Our experts will:

- Complete pre-determinations, prior authorizations, and support medical necessity reviews
- Verify eligibility and benefits for PKU formula through pharmacy or other medical plans
- Help find in-network suppliers to fulfill orders
- Assist in appealing denials for coverage and help resolve billing discrepancies

**NUTRICIA IS HERE TO LEND A HELPING HAND**



**NUTRICIA  
NAVIGATOR**

To learn more visit

**[NutriciaMetabolics.com/Reimbursement](https://NutriciaMetabolics.com/Reimbursement)** or call **1-800-365-7354**

Monday–Friday 8:30a.m. – 5:00p.m. ET.

## Who's who (contact details)

## My dietitian

Name: .....

Phone #:.....

Email: \_\_\_\_\_

## My nurse

Name: \_\_\_\_\_

Phone #: \_\_\_\_\_

Email:.....

## My doctor

Name: \_\_\_\_\_

Phone #: \_\_\_\_\_

Email: 

# Notes

This image shows a single sheet of white paper with horizontal ruling lines. The lines are evenly spaced and extend across the width of the page. There is a light blue decorative border at the top edge and a pink decorative border at the bottom edge. The paper appears to be part of a notebook or a set of stationery.



# 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?

**Visit us at [NutriciaMetabolics.com](https://www.NutriciaMetabolics.com)**

All products shown are medical foods in the U.S. and foods for special medical use in Canada for the dietary management of Phenylketonuria (PKU) and must be used under medical supervision.

