



PHENYLKETONURIA (PKU) FREQUENTLY ASKED QUESTIONS

SERN/GMDI Nutrition Management Guidelines

First Edition

F.A.Q. About Nutrition Management for Individuals with PKU^{1,2,3}

December 2018

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NUTRIENT INTAKE	
<i>How much phenylalanine (PHE) or protein can I eat?</i>	The amount depends on your individual tolerance, age, weight, and needs for growth (in children) or health maintenance (in adults). Your metabolic clinic will use your lab results and your clinical status to recommend PHE or protein intake goals (Rec 1.12).
<i>Do I need a PKU medical food (formula)?</i>	Medical food is needed if your PHE restriction does not allow enough protein from food for appropriate growth and health. This protein will usually be provided by a combination of food and medical food recommended by your metabolic clinic (Rec 1.2, 3.12).
<i>Do I need to take a tyrosine (TYR) supplement?</i>	Medical food contains the extra TYR needed for PKU, if it is consumed through the day and in the amounts your metabolic clinic recommends. If your blood TYR measures within normal range, you should not need a TYR supplement (Rec 1.32).
<i>How do I know if I am getting the right number of calories?</i>	Your calorie needs are based on age, weight, activity level and growth (in children and adults). Your clinic will make recommendations that are right for you (Rec 1.42).
MEDICINE	
<i>Are there medications I can take for PKU?</i>	Two medications are available to help treat PKU. Kuvan® is taken by mouth and Palynziq™ is injected. Your metabolic clinic can discuss these and recommend what is appropriate for your treatment. Talk with your metabolic clinic about what you must do to try one of these medications (Rec 5.1, 5.22).
<i>If I respond to a medication, how can that change my diet?</i>	If your clinic determines that you respond to Kuvan® or Palynziq™, they may ask you to complete a PHE challenge to see how your diet may be liberalized. If you respond to either medication, your clinic will adjust your recommended diet and help you to learn healthy ways to include higher protein foods in your diet (Rec 5.32).
MONITORING	
<i>What are the goals for PHE and TYR blood levels?</i>	The treatment range for blood PHE generally considered safe for brain health is 120-360 µmol/L throughout life. Blood Tyr should be within normal range (Rec 2.12).
<i>How often should monitoring of my PKU be done?</i>	Your metabolic clinic will recommend how often lab testing should be done (based on past lab results and any adjustments needed for growth or activity level) and how often you should come to clinic. They may also recommend additional testing to monitor your nutritional needs and your neurocognitive status (Rec 4.3, 4.4, 4.52).
<i>How should I keep track of how much PHE or protein I eat?</i>	PHE and protein from food can be tracked using mg PHE, g protein, or exchanges (1 exchange=15 mg PHE). Your clinic may also recommend a simplified diet where only higher PHE or protein foods are tracked. Keeping a food diary helps you to monitor what you are eating and helps your clinic to suggest changes (Rec 4.12).

This document is not meant to substitute for the medical advice provided by your doctor.

¹ For the child, teenager, or adult living with PKU and their caregivers.

² Based on the 2015 Nutrition Management Guidelines for Phenylketonuria (PKU) by GMDI/SERN: https://southeastgenetics.org/ngp/guidelines_pku.php

³ The Management Guidelines Advisory Committee used the nationally standardized condition abbreviation of PKU; curated by the US National Library of Medicine for this and related guideline products: <https://newbornscreeningcodes.nlm.nih.gov/>

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PREGNANCY

<i>What should I do before becoming pregnant?</i>	Before becoming pregnant, blood PHE should be consistently in control (120-360 µmol/L). High blood PHE during pregnancy (especially during the first trimester) is known to cause significant birth defects in a baby. If you are already pregnant, this is considered a medical emergency and you should contact your metabolic clinic immediately (Rec 6.12).
<i>How does pregnancy change my PKU treatment?</i>	Your metabolic clinic will adjust your treatment recommendations to provide PHE control and meet the additional nutritional needs of pregnancy. More frequent monitoring and clinic visits will be needed (Rec 6.2, 6.32).

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