



PROPIONIC ACIDURIA (PROP/PA) FREQUENTLY ASKED QUESTIONS

SERN/GMDI Nutrition Management Guidelines

First Edition

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

February 2020

| NUTRIENT INTAKE | |
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| <i>How much protein can I consume?</i> | The amount depends on individual tolerance, age, weight, and growth (children) or health maintenance (adults). Your dietitian will use your lab results and your clinical status to adjust your protein goals (Rec 1.1). |
| <i>Do I need a PROP medical food (formula)?</i> | PROP formula is needed if your food protein tolerance is less than the amount of protein recommended for your age (Rec 1.2). |
| <i>How do I know if I am getting the right number of calories?</i> | The correct number of calories is based on age, weight, activity level and growth (children) or health maintenance (adults). Your dietitian will recommend the right amount of calories for you. Most individuals with PROP need extra calories when ill (Rec 1.5,2.1). |
| SUPPLEMENTS | |
| <i>Do I need isoleucine or valine supplements?</i> | If your blood levels of isoleucine or valine are too low, your dietitian may increase food (intact) protein and PROP medical formula protein to bring isoleucine or valine blood levels up rather than giving amino acid supplements (Rec 1.1,1.3). |
| <i>Do I need carnitine?</i> | Most individuals with PROP need to take a carnitine supplement to keep their blood carnitine level within the normal range (Rec 3.1). |
| <i>Should I take prebiotics?</i> | More research is needed. Discuss with your physician and dietitian if prebiotics (non-digestible substances in foods, usually fiber) are right for you to support bowel health (Rec 3.6). |
| MONITORING | |
| <i>What are the goals for isoleucine, valine, threonine and methionine blood levels?</i> | For individuals with PROP the goal is to maintain amino acid levels in the blood within the normal range (Rec 1.1, 1.3, 4.4). |
| <i>How often should blood testing be done?</i> | Your physician and dietitian will work with you to establish a lab testing schedule that is best for you based on past lab results. Discuss with your team how often to come to clinic to check your labs as well as your health and nutrition status (Rec 4.4, 4.5). |
| ILLNESS | |
| <i>What should I do if I become ill?</i> | Call your metabolic physician. Refer to your emergency letter (Rec 2.7). |
| <i>How does management change during illness?</i> | Sick day instructions from your metabolic physician and dietitian may include: <ul style="list-style-type: none"> • Reduce or hold protein intake, but for no more than 48 hours • Increase liquids and calories • Stay in contact with your metabolic clinic to report any changes For more severe illnesses, medications such as Carbaglu® or Flagyl® may be recommended (Rec 2.7, 3.3, 3.4). |
| LIVER TRANSPLANT | |
| <i>How does treatment for PROP change after a liver transplant?</i> | After transplant, an individual with PROP may be able to increase food (intact) protein intake to the recommended amount for someone their age without PROP. Most will likely need to continue taking carnitine and stay in contact with their metabolic clinic (Rec 7.4, 7.5). |
| PREGNANCY | |
| <i>Have any women with PROP been able to have children?</i> | With close monitoring and medical management, women with PROP have had children (Rec 5.2, 5.3, 5.4, 5.5). |

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

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

² Based on the Nutrition Management Guidelines for Propionic Acidemia (PROP) by GMDI/SERN: https://southeastgenetics.org/ngp/guidelines_prop.php

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

