



PROPIONIC ACIDEMIA (PROP/PA) CONSUMER SUMMARY

SERN/GMDI Nutrition Management Guidelines

First Edition

Nutrition Guidelines for Individuals with PROP/PA^{1,2,3}

April 2018

This information is intended for individuals with Propionic Acidemia (PROP) and/or their caregivers (hereafter referred to as “you/ your”). This summary includes current management recommendations and suggests topics that you can discuss with your metabolic team to help identify a plan that is best for you.

- Follow your metabolic clinic’s instructions to meet your needs for protein, calories, vitamins and fluid from food and/or medical food (formula). The amounts prescribed are based on individuals age, weight and height, activity level, lab results, and whether you are sick or healthy. Ask your metabolic clinic if you should receive supplements and/or medications such as carnitine, fiber, metronidazole (Flagyl®) or carglumic acid (Carbaglu®).
- Contact your metabolic clinic if you are sick (fever, vomiting, diarrhea, constipation, infection, etc.) or have ketones in your urine. Your care team may want you to change your feeding plan for the next 24-48 hours. Stay in contact with your clinic to report any changes in health status.
- Keep track of the food and medical food you have taken and bring the diet record to clinic. Before or during your clinic visit, discuss with your metabolic team if you should have blood and urine tests. At each clinic visit, your weight, height (and head circumference in young children), should be measured and BMI for (older children, teens and adults) should be calculated and tracked for progress.
- If you are a female, obtain information from your metabolic clinic about the impact of puberty, menstruation, and pregnancy and potential changes to diet, medications and supplements.
- Notify your clinic about any health concerns, as you may need to change your diet or medications. Problems such as pancreatitis, heart issues, vision issues, and a weakened immune system can occur even if you are following your treatment plan.
- Learn about liver transplantation as a possible treatment of PROP. Although liver transplantation does not cure PROP, some individuals may safely add more protein in their diets and experience fewer hospitalizations after transplantation.

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

¹ For children, adolescents, and adults with PROP, or their caregivers

² Based on the 2017 Nutrition Management Guidelines for Propionic Acidemia (PROP) by GMDI/SERN: https://southeastgenetics.org/nap/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/>

