



# UREA CYCLE DISORDERS (UCD) CONSUMER SUMMARY

MNT4P/GMDI Nutrition Management Guidelines

First Edition

Nutrition Guidelines for Individuals with UCDS<sup>1,2</sup>

April 2026

This information is intended for individuals with Urea Cycle Disorders (UCDs) 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.

- Individualized treatment plans are based on your UCD type, disease severity, age, growth, clinical status, and laboratory results.
- Follow your metabolic clinic’s instructions to meet your needs for protein, calories, fluids, and other nutrients. Your prescribed protein intake should provide enough for growth and health while helping prevent high ammonia levels.
- Essential amino acid-based medical food may be prescribed to help meet protein and nutrient needs while limiting nitrogen load. Your metabolic team will determine whether medical food is needed and how much is right for you.
- Work with your metabolic team to always have a sick-day plan and clear instructions for when to call the on-call metabolic specialist and when to seek emergency care. Seek medical attention promptly if you are unable to tolerate food or fluids.
- Avoid prolonged fasting. If you need surgery, a medical procedure, or another reason for not eating for a long period of time, work with your metabolic team in advance so fasting time can be minimized and metabolic support provided.
- For infants with UCD, breast milk (direct breastfeeding or expressed breast milk) may be used with close guidance from the metabolic team. Standard infant formula and/or protein-free formula may also be recommended to help meet protein and energy needs safely.
- If oral intake is not enough to meet your needs, especially in severe UCD, your team may recommend [feeding support](#), a feeding specialist, or a feeding tube to help provide nutrition and medications safely and consistently.
- Ask your metabolic team whether you need medications or supplements such as nitrogen scavengers, L-citrulline, L-arginine, carnitine, vitamins, minerals, or essential fatty acids.
- During clinic visits, your growth, development, physical activity, feeding tolerance, and nutrient intake will be assessed, and blood tests will be used to monitor nutrition status and metabolic control.
- If you are female, ask your metabolic team about the effects of menstruation, pregnancy, delivery, postpartum recovery, and breastfeeding, and whether temporary changes in diet, medications, or monitoring may be needed during these times.
- Some individuals with UCD may be candidates for liver transplantation. This can improve survival and may allow an unrestricted protein diet, but nutrition monitoring remains important before and after transplant.

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

<sup>1</sup> For children, adolescents, and adults with UCDS, or their caregivers

<sup>2</sup> Based on the 2026 Nutrition Management Guidelines for Urea Cycle Disorders (UCD) by GMDI / MNT4P:

[https://managementguidelines.net/guidelines\\_ucd.php](https://managementguidelines.net/guidelines_ucd.php)

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