



COVID-19 Information for Patients with Urea Cycle Disorders and their Families

COVID-19, the coronavirus currently causing global concern, is of particular concern for the elderly and those with underlying health issues. We acknowledge that this is a difficult time with much uncertainty, and recommend that taking precautions is currently the best strategy. In the following **FAQs**, we have attempted to provide answers to some of the questions that are relevant to those affected with urea cycle disorders (UCD) and their families.

Coronavirus (COVID-19) is a respiratory illness caused by a novel (new form) of coronavirus. Individuals affected with COVID-19 typically develop fever, cough, and shortness of breath. There may be mild cases that may not have all of these three key features. Others may have general body aches or flu-like symptoms. Complications of COVID-19 can include development of pneumonia in the lungs, respiratory failure, multi-organ failure and, in some cases, death.

In order to avoid complications that may trigger instability in UCD such as elevated ammonia levels, we recommend that individuals with urea cycle disorders and their families be particularly careful about exposure, and contact their physicians immediately if they suspect they are symptomatic. This is the time to be in close contact with your metabolic specialist about any concerns that you may have about risks of exposure, and to have a plan in place in the event that there is an COVID-19 exposure in your family.

COVID-19 is highly contagious and can spread from person to person through coughing, sneezing, or touching surfaces. The World Health Organization (WHO) has declared the coronavirus outbreak a pandemic, meaning a worldwide epidemic. COVID-19 has been reported in countries on all continents except Antarctica. In the US and Europe, clusters of COVID-19 outbreaks through community transmission have been identified. This is an evolving situation and there are still many unanswered questions. We encourage you to refer to sources that provide updated and reliable information like the ones listed below:

<https://www.cdc.gov/coronavirus/2019-nCoV/index.html>

<https://www.who.int/emergencies/diseases/novel-coronavirus-2019>

Please also check for area-specific resources available on the official websites of your Local and State Government. Additionally, hospitals that provide care for you or your UCD-affected family member should have information on their preparedness and policies regarding COVID-19. Please make sure to get your information from official and reliable sites, as there are many “websites” and Facebook pages that are spreading misinformation.

A. Risks of infection with COVID-19 and precautions that may help reduce the risk of infection

Are individuals with UCDs at higher risk of complications from COVID-19?

1) Currently, it is not known whether individuals with UCDs or other inborn errors of metabolism are at increased risk for developing COVID-19 or its respiratory complications. Individuals of any age or health status can become infected and develop complications. Experts believe that although healthy children are generally less severely affected by the virus than adults, they can be carriers of COVID-19 and infect vulnerable UCD children and adults.

2) **An acute viral (infectious) illness in individuals with UCDs can trigger hyperammonemic crisis (elevated ammonia levels).** Therefore, individuals with UCDs and their caregivers/families should take extra precautions. This is especially important in UCD individuals who have a history of recurrent episodes of hyperammonemia. We advise that you contact your metabolic physician and have a plan in place in the event that the UCD-affected individual needs emergency care during the pandemic.

3) Individuals with UCDs who received a liver transplant typically take medications that suppress their immune system. Individuals with liver transplant also need to take extra precautions and should seek the advice of the medical professionals managing their posttransplant care.

What steps can we take to prevent infection in individuals with UCDs?

The virus is thought to spread mainly between people who are in close contact with one another (within about 6 feet) through respiratory droplets produced when an infected person coughs or sneezes. These viral particles may also stay suspended in the air. An infected person without symptoms can also carry the virus and infect others. A person could get COVID-19 by touching a surface or object that has the virus on it and then touching their own mouth, nose, or eyes. COVID-19 has been reported to live on surfaces, like plastic and metal, for several days.

The following precautions may help decrease the chance of infection. The most effective preventions include hand hygiene and social distancing:

- Wash your and your child's hands with soap for at least 20 seconds frequently. Scrub the palms, fingernails, between the fingers and the backs of the hands. If soap and water are not available, use hand sanitizers that contain at least 70% alcohol. These measures are especially important before eating, touching your face, or after touching shared surfaces such as doorknobs, faucets, keyboards, etc. Frequently disinfect surfaces with Lysol spray or disinfecting wipes.
- Teach children to avoid touching their face as much as possible.
- Avoid (and teach your child) touching surfaces outside your home that carry viruses, such as doorknobs, hand railings, etc. Use hand sanitizer or wash hands immediately.
- Avoid large crowds. If you need to go out, try to maintain a distance of at least 6 feet from others. Don't shake hands or hug others. Avoid using public transportation.
- Ask everyone entering your home to wash their hands. Do NOT be shy about asking individuals whether they have viral symptoms or have been exposed and, if so, telling them to stay away.
- If someone in your household develops signs of viral illness, have them wear an earloop mask and isolate them to avoid exposing the individual with UCD and other family members.

Should my UCD-affected child stay home from school/day care and other activities?

The Centers for Disease Control (CDC) is recommending social distancing as a measure to curb the spread of the

virus. Please discuss this with your metabolic doctor, and work with your school. Schools are working hand-in-hand with their local departments of health to determine if schools need to be closed. Most schools are developing online curriculums and programs that can be accessed from home.

We are planning travel, is it safe to travel with a child or adult with UCD?

Please discuss any travel plans with their metabolic doctor. The CDC advises refraining from non-essential travel. We recommend NOT taking a UCD-affected person on a cruise and to avoid airports and air travel. Read the CDC's travel advisory at: <https://www.cdc.gov/coronavirus/2019-ncov/travelers/index.html>

If you or a family member have traveled in the last 2 weeks, call your doctor to determine if a quarantine is needed.

B. Management of UCDs during the COVID-19 pandemic

Is it safe to take my child for a clinic appointment in the hospital?

1) The healthcare facility or region where you receive care may have specific policies regarding infection control and screening for COVID-19. Healthcare facilities may also have limitations on the number of caregivers who will be allowed to accompany children during these clinic appointments. Please refer to these policies before you go take your child to the appointments. Some UCD clinics are planning for telemedicine visits. Ask about the availability and appropriateness of a telemedicine visit.

What additional measures should we take for management during these times?

- 1) Contact your metabolic team and have a plan in place for evaluation and management of a potential hyperammonemic crisis. Emergency department visits may not be recommended during the pandemic and an alternative plan should be in place.
- 2) The COVID-19 outbreak could potentially affect supply chains. Work with your physician's office and pharmacy to maintain an adequate supply of medications, supplements, and metabolic formula that are critical for management of a child or adult with UCD. Don't forget gloves, saline and other supplies needed for tube feed care, ports, etc.
- 3) Ask your UCD physician what type of fever-reducing medicine the UCD-affected individual can take, and have a supply on hand, along with oral rehydration solutions to use as recommended.
- 4) Have a plan for where and how family members with symptoms of COVID-19 will be isolated in the home, and how you would prevent their contact with the UCD-affected family member.
- 5) In the event the primary caregiver has symptoms of COVID-19, have a plan for who will take over the care of the UCD-affected person, measure and administer their UCD medications, formula and supplements, and monitor their UCD diet and protein intake. Make sure to post emergency contact information for the UCD metabolic physician in a conspicuous place (like on the refrigerator).

What should I do if a UCD-affected child or adult develops symptoms of COVID-19?

Call your primary healthcare provider and your metabolic physician's office to inform them of the symptoms so that they can advise you regarding the next steps for evaluation and treatment.