

# Emergency Preparedness Overview:

## How to Prepare Yourself for an Emergency

When you have HCU or any other special dietary needs, being ready to handle difficult situations requires special planning, in addition to the typical things everyone needs to consider in the event of an emergency or natural disaster. Here are some key things to think about and discuss with your family today.

### Emergencies

People with HCU require specialized care and medical foods, both of which may be unavailable or in limited supply in the event of an emergency.

You may not be able to follow your diet perfectly during an emergency but it is important to adhere to it as closely as possible and continue to take your formula and other medications! Emergencies usually cause confusion and stress; if you take the proper steps, sticking to diet doesn't have to be one of them. Some emergencies allow you to stay at home (such as when you lose power), others require you to evacuate. **It is important to prepare for both of these possibilities before they happen!**

### Hospitalization

- Bring Medical Emergency Letter. **See example below.**
- Notify your metabolic team if you are in the hospital. They can assist the medical team.
- Bring your formula (and anything you mix into your formula) with you.
- For overnight stays, bring low protein food and your betaine (Cystadane).
- You will need to educate the hospital staff on your formula, low protein foods, and betaine.
- If you require surgery that includes general anesthesia, inform your care team and/ or anesthesiologist of the higher risk for blood clots associated with homocystinuria.

### In the Event of an Evacuation

Have a plan for:

- Under what circumstance will you evacuate?
- How will you evacuate?
- Where will you evacuate to?
- Where will you go if the evacuation period is extended?
- What clinic will you go to if you evacuate for an extended period?
- Is your evacuation kit ready to go?

Items to include:

- You may want to consider buying a small hot plate or similar appliance that can be used to prepare food in a hotel room. You may also want to obtain a camp stove (propane) that can be used to prepare food without electricity and a manual can opener.
- Put together an emergency kit. **See check list below**

## **Following Your Emergency Plan**

If an emergency occurs you should be ready to immediately follow the emergency or evacuation plan you have put together.

- If you have warning that there will be an emergency, begin preparing to follow your emergency plan as soon as possible.
- After an emergency situation occurs, make contact with your team as soon as possible.
- Your team may not have access to your diet information, so be prepared to tell them:
  - Your daily protein/ methionine restriction
  - What medical formula you use and how much each day
  - If you take medication, what you use and how much each day
  - How long your supply for each of these things will last
- Contact the companies who supply your medication, medical formula, or low protein foods and tell them your new contact information if you do not have enough supply to last until you can return home or to your usual way of obtaining these things.
- Contact the team and other health care providers in your new location to continue your HCU care.

# Emergency Preparedness Supplies

## for Classical Homocystinuria/ Cystathionine Beta-Synthase (CBS HCU)

- Metabolic geneticist name and contact information \_\_\_\_\_
- Dietitian name and contact information \_\_\_\_\_
- Primary care provider or pediatrician name and contact information \_\_\_\_\_
- Local hospital/ emergency room name and contact information \_\_\_\_\_
- Pharmacy name and contact information \_\_\_\_\_
- Formula manufacturer name and contact information \_\_\_\_\_
- Formula prescription \_\_\_\_\_
- Medication name and dosage \_\_\_\_\_
- Medication prescription # \_\_\_\_\_

- |                                                                                                            |                                                                                                              |
|------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------|
| <input type="checkbox"/> Emergency letter with diagnosis and treatment plan from metabolic/genetics clinic | <input type="checkbox"/> Preferred container for consuming metabolic food/formula                            |
| <input type="checkbox"/> Copy of medical records/most recent lab results                                   | <input type="checkbox"/> Purified or bottled water                                                           |
| <input type="checkbox"/> Paper copy of family contact list                                                 | <input type="checkbox"/> Low protein food list or list of low protein content of foods in your emergency kit |
| <input type="checkbox"/> 2 week supply of metabolic food/formula                                           | <input type="checkbox"/> Low protein food vendor list and their contact information                          |
| <input type="checkbox"/> 2 week supply of low-protein food                                                 | <input type="checkbox"/> Supplements/Prescriptions                                                           |
| ■ Include foods that don't require cooking or refrigeration                                                | <input type="checkbox"/> Daily medications                                                                   |
| <input type="checkbox"/> Food scale with extra batteries                                                   | <input type="checkbox"/> Thermometer and fever reducer                                                       |
| <input type="checkbox"/> Set of household measuring cups & spoons                                          | <input type="checkbox"/> Solar-powered chargers                                                              |
| <input type="checkbox"/> Calculator and preferred method for tracking methionine or protein intake         | <input type="checkbox"/> Adapters for car chargers                                                           |
| <input type="checkbox"/> Container for mixing metabolic food/formula                                       | <input type="checkbox"/> Hand-powered radio                                                                  |

This list was last updated: \_\_\_\_\_

# Other Emergency Preparedness Tools Continued

## Other Medical IDs

### Emergency Card

Patients and families may print out and complete the card provided below and carry it in case of emergency.

#### Set up your Medical ID

1. Open the Health app comes with phone and tap the Medical ID tab.
2. Tap Edit. If asked, tap Edit Medical ID.
3. To make your Medical ID available from the lock screen on your iPhone, turn on Show When Locked. In an emergency, this gives information to people who want to help.
4. Enter health information like your birth date, height, and blood type.
5. Tap Done.

#### Edit emergency contacts

1. Open the Health app and tap the Medical ID tab.
2. Tap Edit, then scroll to Emergency Contacts.
3. To add an emergency contact, tap  under emergency contacts. Tap a contact, then add their relationship.
4. To remove an emergency contact, tap  next to the contact, then tap Delete.
5. Tap Done.

### Android Users

Android phones don't have a built in health app that allows you to create a Medical ID. We recommend the app: *Medical ID (Free): In Case of Emergency* from your phones app Store.

1. First, download the app. Go to the app store and download *Medical ID (Free): In Case of Emergency*
2. Open the app to create a profile. Fill out the profile with as much information as you prefer. The more information you include the more helpful it will be to Emergency Medical Services when they access it.
3. When done, tap the check mark in the upper right corner to finish the profile. Your Medical ID is ready.
4. One last step - In the top right corner of the app you will see three stacking dots, click the dots, then click **settings**.
5. Then click Floating Icon, this will add a universal Medical ID symbol to the lock screen of your phone. Anyone, even if your phone is locked can see your medical emergency information now.

# Other Emergency Preparedness Tools Continued

## Other Medical IDs

### Emergency Card

Patients and families may print out and complete the card provided below and carry it in case of emergency



#### Diagnosis: Homocystinuria (HCU)

HCU is an inherited metabolic disorder where methionine and homocysteine levels are elevated. If not treated, homocysteine and methionine can build up to harmful levels causing lens dislocation, strokes, heart attacks and other thromboembolic events. HCU symptoms are triggered by high homocysteine, which is made from proteins you ingest.

If hospitalized, do not provide foods containing protein. Special low-protein foods and formula must be provided. General anesthesia should be avoided.



#### Diagnosis: Homocystinuria (HCU)

In case of an emergency: call the Metabolic Specialist

Patient Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_ State: \_\_\_\_\_ Zip code: \_\_\_\_\_

Phone: \_\_\_\_\_ Birthdate: \_\_\_\_\_

Allergies: \_\_\_\_\_

Metabolic Specialist: \_\_\_\_\_

Phone: \_\_\_\_\_ Fax: \_\_\_\_\_

Primary Physician \_\_\_\_\_

Phone: \_\_\_\_\_ Fax: \_\_\_\_\_

### Medical ID Tag



A medical ID tag is a small tag worn as a bracelet or neck chain that includes a message that the person wearing it has an important medical condition that may require immediate attention.

We recommend including your full name, emergency contact number, your diagnosis (homocystinuria), allergies, and notation if you require a low protein diet and/or formula.

# Medical Emergency Letter

**[Institution Information]**

**[Date]**

**RE: [Name]**

**DOB: [ ]**

**MR: [ ]**

To whom it may concern:

**[Name]** is a patient followed in the Metabolism Clinic at **[Hospital Name]** due to **[his/her]** diagnosis of Homocystinuria. **[Name]** is at increased risk for blood clots due to increased homocysteine. **[S/He and his/her]** family are educated about the signs and symptoms and of the risk of severe illness, and may present to your emergency room if **[s/ he]** does not respond to home management.

Complications may be prevented or ameliorated by immediate and appropriate management.

If **[Name]** should present with a history of prolonged decreased oral intake and/or vomiting for any reason, and if home management of sugar and fluid has not reversed the symptoms or if **[s/he]** presents with signs of infection or blood clot; we suggest the following:

1. Evaluate immediately.
2. Speak to the doctor on call for Genetics at **[Hospital Name]** calling **[Genetics number]** and asking for the "geneticist on call" Immediately upon **[Name]'s** arrival for additional instructions
3. Start IV fluids with glucose (at least 6-8 mg/kg/min), and appropriate salts (usually D10 normal saline with 10 mEq/L potassium chloride at 200 mL/h). A normal saline bolus may be Y-ed into the D10, but continue dextrose delivery is important.
4. Check electrolytes, liver function, CBC, urine ketones, total homocysteine and appropriate cultures or infection screens. Depending on **[his/her]** condition, a blood gas is warranted.
5. Diagnose and treat the precipitating event.
6. **[Name]** is at increased risk for blood clots and strokes so respond accordingly.

Please call with any questions and prior to discharge or admission.

Sincerely,

**[Doctor Signature]**