

Emergency Preparedness Overview:

How to Prepare Yourself for an Emergency

When you have Homocystinuria (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.**

Hospitalization

- **Have your genetics team complete the appropriate emergency letter. See example below.**
- Bring Medical Emergency Letter.
- 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:

- **Put together an emergency kit. See check list below**
- 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.

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 information, so be prepared to tell them:
 - Your daily protein/methionine restriction (if applicable)
 - What medical formula you use and how much each day (if applicable)
 - If you take medication, what you use and how much each day
 - How long your supply for each of these things will last
 - What risk or allergies do you have
- 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 HCU/ Cobalamin Disorders and Severe MTHFR

- ◆ 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 _____

Must-haves

- Emergency Letter with diagnosis and treatment plan from metabolic/genetics clinic
- Copy of medical records and most recent lab results
- Paper Copy of family contact list
- 2 weeks supply of:
 - Formula (if applicable)
 - Cystadane/Betaine
 - Hydroxocobalamin and syringes (if applicable)
 - Low protein food (if applicable)
 - Include foods that don't require cooking or refrigeration
- Set of household measuring cups and spoons
- Container for mixing metabolic food or formula
- Preferred container for consuming metabolic food/ formula/ tube feeding supplies
- Purified or bottled water
- Supplements/Prescriptions/ other medications
- Thermometer and fever reducer

Nice to Haves

- Food Scale with extra batteries
- Calculator and preferred method for tracking methionine or protein intake (if applicable)
- Low Protein food list or list of low protein content of foods in your emergency kit (if applicable)
- Low protein food vendor list and their contact information (if applicable)
- Solar-powered chargers
- Adapters for car chargers
- Hand powered radio

This list was last updated: _____

Other Emergency Preparedness Tools Continued

Medical IDs

Medical ID helps first responders access your critical medical information from the lock screen, without needing your passcode. They can see information like allergies and medical conditions as well as whom to contact in case of an emergency.

iPhone Users

Set up your Medical ID

1. Open the Health App: This is the app with a white icon that has a pink heart.
2. Tap on your Profile Picture: On iOS 13 and later, you'll find your profile picture or icon in the top-right corner of the screen. On earlier versions of iOS, tap "Medical ID" at the bottom of the screen.
3. Tap on "Medical ID": This will take you to the Medical ID setup.
4. Tap "Edit": Located usually at the top-right corner.
5. Fill in Your Information: You can add various details like:
 - Name
 - Medical notes
 - Emergency contact
 - Date of birth
 - Allergies & reactions
 - Blood Type
 - Medical conditions
 - Medications
 - Weight and height
6. Enable "Show When Locked": This ensures that your Medical ID can be accessed even when your phone is locked, which can be crucial in emergencies.
7. Tap "Done": Once you've entered all your desired information.

Other Emergency Preparedness Tools Continued

Android Users

Adding Medical ID information on Android can vary by phone brand and Android version, but most devices include it in the emergency information settings. Here is a guide for adding Medical ID details to your Android device:

1. Unlock your Android Device: Start at your home screen.
2. Open the Settings App: This is typically represented by a gear icon.
3. Search for Emergency Information: Depending on your version and brand, you might find it under:
 - o About phone > Emergency information, or
 - o Users & accounts > Emergency information.
4. Enter the Emergency Information Page: Once you've found the emergency information setting, tap on it.
5. Edit or Add Information:
 - o Tap on "Edit" or the pencil/edit icon.
 - o You'll have fields to input medical details such as allergies, medications, blood type, and any other medical notes.
 - o You can also add emergency contacts. This will typically pull from contacts you've saved in your phone's Contacts app.
6. Save the Information: Once you've entered all the necessary details, make sure to save or confirm the input, if required.
7. Test the Setup: Lock your phone. On the lock screen, there should be an "Emergency" option. Tap on it, and you should be able to view your emergency information without unlocking the phone. This ensures that first responders can see your medical information even if your phone is locked.

Optional 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.

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. Nitrous oxide 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: _____



Diagnosis: Cobalamin (Cbl)_____

Cobalamin _____ is an inherited B-12 responsive metabolic disorder where methionine is low and homocysteine levels and methylmalonic acid (methylmalonic acid, will not be high in CblE and CblG patients) are elevated. If not treated, harmful levels can cause eyesight degradation, strokes, heart attacks and other thromboembolic events. HCU symptoms are triggered by high homocysteine and methylmalonic acid, and low methionine.

If hospitalized, B-12 should be administered IM or IV and nitrous oxide should always be avoided.



Diagnosis: Cobalamin _____

In case of an emergency: call the Metabolic Specialist

Patient Name: _____

Address: _____

City: _____ State: _____ Zip code: _____

Phone: _____ Birthdate: _____

Allergies: _____

Metabolic Specialist: _____

Phone: _____ Fax: _____



Diagnosis: Severe MTHFR (NOT the common variants)

Severe MTHFR is an inherited metabolic disorder where methionine is low and homocysteine levels are elevated. If not treated, harmful levels can cause eyesight degradation, strokes, heart attacks and other thromboembolic events. HCU symptoms are triggered by high homocysteine, and low methionine.

If hospitalized, nitrous oxide should always be avoided.



Diagnosis: Severe MTHFR

In case of an emergency: call the Metabolic Specialist

Patient Name: _____

Address: _____

City: _____ State: _____ Zip code: _____

Phone: _____ Birthdate: _____

Allergies: _____

Metabolic Specialist: _____

Phone: _____ Fax: _____

Medical Emergency Letter: Classical HCU

Please take this to your genetics team and have them complete BEFORE an emergency arises

[Institution Information—Put on Institution Letterhead]

[Date]

RE: [Patient First and Last 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] by calling [Genetics Emergency On Call 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}

Medical Emergency Letter:

Cobalamin Disorders and Severe MTHFR

Please take this to your genetics team and have them complete BEFORE an emergency arises

[Institution Information—Put on Institution Letterhead]

RE: [Patient First and Last Name]

[DOB:]

[MR:]

To whom it may concern:

[Date]

[Name] is a patient followed in the Metabolism Clinic at [Hospital name] due to his/her diagnosis of [Cobalamin __, deficiency, Methylene Tetrahydrofolate Reductase deficiency (MTHFR)]. [Name] is at high risk of metabolic decompensation with metabolic acidosis from his/her disorder (Only in cobalamin C, D, F, J, and X patients). In addition, he/she is at increased risk for blood clots due to increased homocysteine. His/ Her family has been educated about the signs and symptoms and of the risk of metabolic decompensation during ordinary childhood illnesses, and may present to your emergency room if the patient 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 she 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] by calling [Genetics Emergency On Call Number and asking for the "geneticist on call" immediately upon [Name]'s arrival for additional instructions**
3. If not given today already by family, please give one mg IM/SC hydroxocobalamin immediately. Cyanocobalamin is contraindicated, especially in patients with cbIC.
4. Start IV fluids with glucose (at least 6-8 mg/kg/min), and appropriate salts (usually D10 half-normal saline with 10-20 mEq/L potassium chloride at 1 ½ maintenance rate is adequate).
5. Check blood glucose, electrolytes, liver function, CBC, urine ketones, total homocysteine and appropriate cultures or infection screens. Depending on [his/her] condition, a blood gas is warranted.
6. Diagnose and treat the precipitating event.
7. Individuals are also at increased risk for blood clots and strokes so respond accordingly.
8. Nitrous oxide should be avoided as anesthetic agent. Please call with any questions and prior to discharge or admission.

Sincerely,

Issuing Provider

Date
