PROPIONIC ACIDEMIA FOUNDATION

Emergency Preparedness Handbook for Those with Propionic Acidemia/ Methylmalonic Acidemia

Table of Contents

Introduction	01
Things to Learn	02
Things to Do	03
Information to Gather	09
Items to Pack	14
Home Preparations	17
Other Considerations	18
Resupply & Review	19
More Information	20

© 2023 Propionic Acidemia Foundation

Introduction

We hope this guide will be helpful to you in making preparations, so that when an emergency takes place you/your loved one with Propionic Acidemia (PA)/Methylmalonic Acidemia (MMA) will be able to have their needs met and not have any adverse health effects.

An Emergency Plan and an Emergency Supply Bag, often referred to as a 72-hour kit, is recommended for all families and individuals by multiple entities including FEMA and the Red Cross. It is especially important for those with complex medical needs to have extra supplies and an emergency plan in place as organizations giving aid in an emergency will not have the means to support those with highly specific or complex medical needs.

Weather emergencies and supply chain issues can affect anyone. In the United States, we have a wide range of natural disasters that can be impactful to a family: floods, fire, ice, volcano, hurricane, tornado, snow, earthquake, tsunami, or a land/mudslide. Man-made events can also affect your ability to get needed supplies. A trucking or rail strike can affect the distribution of supplies. Contamination or other production issues can affect the production of formula, supplies, or medication.

When natural disasters happen, the Red Cross, FEMA and other organizations typically do not have their response teams in place and ready to give aid until 72 hours after the emergency arises. However, even when they are giving aid they only have the capabilities to meet basic needs of food, water, and some form of shelter. They do not have the ability to provide for the specialized diet and medical needs of a person with PA/MMA. It is recommended that a person with special medical or food needs have a 2-week supply of the items they need to meet their personal and medical needs. In an emergency, individuals are often housed in a congregate setting like a school or church, which may put a person with PA/MMA at a higher risk of infection. A plan for housing outside the immediate area of where you live is also an important part of your plan.

© 2023 Propionic Acidemia Foundation

Things to Learn

Learn about the types of disasters that are common in your area of the country.

The Red Cross has an interactive map indicating what types of emergencies are most common in each area: https://www.redcross.org/get-help/how-to-prepare-for-emergencies/common-natural-disasters-across-us.html#all

FEMA also has an interactive map which indicates areas of flooding. You will want to zoom into your area for the markings to be visible: https://msc.fema.gov/portal/home

Find out from your health insurance plan how you can have a 2-week supply of medication and formula for an emergency supply. Often insurance companies limit how many days of medication, formula, and supplies you can order at a time.

Contact your school district regarding where your child would evacuate to and how they handle lock-down situations, so that you can prepare for what their needs might be in those situations.

Find where your water, gas and shut offs are in your home, so you can turn them off in an emergency.

Notes:

Things to Do

Below are steps you can take to be more prepared should an emergency arise in your area. You can set up ways to be notified about an emergency in your area, prepare your family, and help make sure treatment needs are met.

☐ **Contact Emergency Services in your city/township or county**

Your local government offices of emergency services will often keep a file on a person with special health care needs, so that if emergency services are called to the home some medical information would be known to those who are responding. This can include an emergency protocol letter stating medical conditions and treatment protocols. You can also inform them of communication and other needs, such as if they are an individual who is non-verbal, has behavior issues or has a vision impairment.

Department _____ Phone Number_____

Date Notified _____ Frequency to Update Information _____

☐ **Contact Utility Providers**

Notify providers for water, power, etc. of medical needs, so in the event of utility service being cut-off your home would be on the priority list for repairs or hook-up. Explain what specific needs your family has (i.e. electricity dependent for feeding pump and formula preparation & refrigeration for formula storage), so that the utility companies can prioritize the needs of community members.

Electric Company _____ Date Notified _____

Phone Number _____

Gas Company _____ Date Notified _____

Phone Number _____

Water Supplier _____ Date Notified _____

Phone Number _____

☐ **Sign-up for local emergency alerts**

Sign-up for local emergency alerts with your local government office. Some cities/counties have text, phone, and/or e-mail alerts when there is a local emergency.

Department _____ Date Notified _____

☐ **Sign up with Medic Alert**

Medic Alert is a non-profit organization that provides a centralized location for medical information to be accessed in an emergency. Medical IDs are worn to inform emergency personnel about medical conditions and contain a personal ID # to get more information. Medic Alert has assistance programs if you cannot afford a membership.

www.medicalert.org 1-800-432-5378

☐ **Enable Wireless Emergency Alerts (WEAs) on your Mobile Phone**

Most newer phones have this already enabled. You can verify this in the settings in your phone.

☐ **Download a Weather App to your Mobile Phone**

A weather app like Accuweather or The Weather Channel can send notifications for major government announcements, public health alerts, severe weather warnings/watches and breaking news. Make sure you have notifications turned on.

☐ **Enable severe weather alerts on Smart Home devices**

Echo Devices can be set up to send alerts about severe weather in your area. To enable it say, “Alexa, tell me when there’s a severe weather alert.”

☐ **Contact your Durable Medical Equipment Provider (DME)**

Contact your DME provider regarding how to acquire supplies if out of the area or equipment becomes damaged in an emergency.

Department _____ Phone Number_____

☐ **Discuss with Your Providers Your Desire to Implement Your Preparedness Plan**

Your clinic staff and other providers are key in helping you get prepared for emergencies.

Request that they coordinate with insurance providers to secure a 2 week supply of medication & formula to have on hand, including “sick formula” ingredients.

Discuss also having an emergency supply of formula and medication on hand at school. The school will need physician orders for any emergency medication and formula. A product like Vitaflo SOS that comes in a premeasured sachet (packet) that is mixed with water can be used by individuals with minimal training. Because SOS is carbohydrate only and is a powder, it has a long shelf-life.

SOS by Vitaflo, a carbohydrate based formula module in premeasured packets. Image courtesy of Vitaflo

☐ **Enter Household & Emergency Contact Information into Mobile Phones**

Enter household and emergency contact information into all household members mobile phones and devices. Also, label one contact as “ICE" (In Case of Emergency) on each device. Emergency services often search for ICE on a phone if an individual is not conscious or unable to communicate.

☐ **Make a Family Communication Plan**

Designate a family member or friend outside of the area as a contact person in case the family is separated or you need to communicate with your medical team. If phone service is not consistent it may be easier to call out of the area than call across a city.

Name_____ Phone Number _____

☐ **Medical Release Forms**

Sign medical release forms with your providers, so your out of the area family member/friend contact can communicate health information with the medical team regarding your affected family member.

☐ **Practice Using a Phone**

Have each family member practice sending text messages. Text messages often can go through in an emergency when a phone connection is not possible.

☐ **Designate Emergency Meeting Locations**

Decide on safe, familiar places where your family can go for protection or to reunite. Make sure these locations are accessible for household members with disabilities. Identify the following places:

Indoor: If you live in an area where tornadoes, hurricanes, or other high-wind storms can happen, make sure everyone knows where to go for protection. This could be a small, interior, windowless room, such as a closet or bathroom, on the lowest level of a sturdy building, or a tornado safe room or storm shelter.
Location: _____

In your neighborhood: This is a place in your neighborhood where your household members will meet if there is a fire or other emergency, and you need to leave your home. The meeting place could be a big tree, a mailbox at the end of the driveway, or a neighbor's house.
Location: _____

Outside of your neighborhood: This is a place where your family will meet if a disaster happens when you're not at home and you can't get back to your home. This could be a library, community center, house of worship, or family friend's home.
Location:_____

Outside of your town or city: Having an out-of-town meeting place can help you reunite if a disaster happens and you cannot get home or to your out of neighborhood meeting place or those in your community are instructed to evacuate the area. This meeting place could be the home of a relative or family friend. Make sure everyone knows the address of the meeting place and discuss ways you would get there.
Location:_____

- [ ] **Contact Health Services for Your School District**

Discuss with the health services office regarding having a supply of medication and formula within the classroom or health office in case an emergency happens at the school. A lock-down situation where a child may be within a room for an extended period of time could put a child with PA/MMA at risk for decompensation. Also, inquire as to having similar supplies at the school's evacuation point.

Medication lock boxes can be installed within classroom spaces to prevent access by unauthorized individuals.

A plan for emergencies, including lock down, evacuations, and other emergencies can be included within an Individual Health Plan within an Individual Education Plan (IEP) or 504 plan.

- [ ] **Safety Devices**

Get working smoke alarms, carbon monoxide (CO) detectors and fire extinguishers. There should be a working Smoke Alarm and CO detector outside of each bedroom and on each floor of a house. Smoke alarms and CO detectors can be combined into one unit. Smoke detectors can last 10 years, while CO detectors only last 5-7 years. Combination units only last 7 years Fire extinguishers should be on each level of your home, with one in the kitchen.

☐ **Home Evacuation Plan/Fire Escape Plan**

Make a home evacuation plan by walking through your home and inspecting all possible exits and escape routes. Consider drawing a floor plan of your home and mark two ways out of each room, including windows and doors. Mark the location of each smoke alarm & Carbon Monoxide (CO) detectors.

The National Fire Protection Association has a template for making an escape route drawing, as well as additional fire safety information: https://www.nfpa.org/Public-Education/Staying-safe/Preparedness/Escape-planning

☐ **Out of the Area Evacuation Plan**

Make a plan for evacuating. Decide upon criteria on when you would relocate to and how you would get there.

Evacuation location_____

Primary means of travel_____

Back-up travel method_____

Information to Gather

While we rely heavily on technology, in an emergency we may not have access to information that is needed. Internet access may be unreliable, power may be out in a large area, there may be no way to charge a cell phone battery, and cell service may be out or overburdened, so a paper back-up of important items may be needed.

Medical Contacts

Geneticist

Name _____

Phone_____

Page/cell _____

E-mail_____

Dietician

Name _____

Phone_____

E-mail_____

Primary Care Physician

Name _____

Phone_____

Page/cell _____

E-mail_____

Medical Specialist

Name _____

Phone_____

Page/cell _____

E-mail_____

Medical Specialist

Name _____

Phone_____

E-mail_____

Medical Specialist

Name _____

Phone_____

Page/cell _____

E-mail_____

Medical Specialist

Name _____

Phone_____

Page/cell _____

E-mail_____

Local Hospital

Name _____

Address_____

Phone_____

Contact_____

Pharmacy

Name _____

Location_____

Phone_____

Specialty Pharmacy

Name _____

Location_____

Phone_____

DME Provider

Name _____

Contact_____

Phone_____

E-mail_____

Formula Manufacturer

Name _____ Emergency Assistance Number _____

Name _____ Emergency Assistance Number _____

Printed Orders

- [ ] Formula Prescription
- [ ] Equipment/DME prescription
- [ ] Medication Prescriptions
- [ ] Lab Orders (CBC, CMP, ammonia, quantitative amino acids, urine organic acids, etc.)

Medical Documents

- [ ] Formula Recipe
- [ ] Sick Day Recipe
- [ ] Medication List (If you use myChart, print out a list from a recent After Visit Summary
- [ ] Emergency Protocol Letter
- [ ] Recent Medical Reports & Labs or Up-to-date Care Notebook

Personal Documents

Place in Water Proof Container

- [ ] Insurance Policies
- [ ] Identification (passports, birth certificates, marriage certificates)
- [ ] Social Security Cards
- [ ] Copy of Driver's License
- [ ] Bank Account Records
- [ ] Deed to Your Home
- [ ] Car Title
- [ ] Copy of Will
- [ ] Copy of Guardianship
- [ ] Copy of DNR/Living Will

Personal Contacts

Family members/Friends who are both local to you and a distance away who likely would not be impacted by the same emergency.

Name_____ Phone_____

Name_____ Phone_____

Name_____ Phone_____

Name_____ Phone_____

Name_____ Phone_____

Items to Pack

This section has several parts, specific medical related items that you should gather for the person affected by PA/MMA, items for the caregiver/family member, and general emergency related items. Disregarding any type of supply that would not apply to your situation or location.

Where to Store

You should store your Handbook & Emergency Kit in your home where it is accessible. You should consider the circumstances of the area in which you live.

For example, in Illinois you may want to keep yours in your basement, but elevated since you are at risk of tornado and would shelter in the basement in that situation, but are also prone to flooding. On the West Coast of the US, you may want yours near the front door of the house in order to evacuate quickly if there is an earthquake or fire.

Supplies for the Person with PA/MMA

Water
Consider where you store your water in case heat is lost in your home. It is recommended that you store 1 gallon of water per person per day. However, you will need consider any protentional illnesses that may cause fluid loss, extra fluid needs with tube flushing, etc., and additional water for hand hygiene.

If feasible, purchase a Portable Power Station for the feeding pump

- [ ] 2 week supply of metabolic formula
- [ ] 2 week supply of sick formula ingredients
- [ ] 2 week supply of non-perishable low protein foods
- [ ] 2 week supply of all medications
- [ ] 3-14+ gallons of clean water
- [ ] Food scale with extra batteries
- [ ] Preferred container for metabolic formula/food
- [ ] Cooler to keep formula cold
- [ ] Oral Syringes & supplies
- [ ] Headlamp, for making formula in the dark
- [ ] Anti-nausea medication
- [ ] G-tube supplies- tubing, syringes, pump bags
- [ ] Pump, extra charging cord, backpack
- [ ] Preferred items to clean stoma
- [ ] Port supplies

Gravity feeding in a pinch

If you find that you need to do a pump feeding, but it is not available or you cannot use an open syringe to feed, you can set up a gravity feeding with a pump bag.

Hang a Command Hook above where you will be feeding.

Hang a gravity feeding bag that has an adjustable roller for flow rate (they can be purchased for a few dollars) or you can use a MOOG Infinity bag and use a clothes pin on the part that fits into the pump and it will open the tubing for it to flow.

Check the flow often to make sure it is not too fast or too slow.

Family Member Supplies

- [ ] 3+ gallons of water per person
- [ ] Pedialyte/gatorade
- [ ] 3+ day supply of non-perishable food per person
- [ ] 14+ days supply of medications
- [ ] Emergency health information for each person
- [ ] Change of clothes for each person
- [ ] Jacket, gloves, hat, rain poncho
- [ ] Flashlight
- [ ] Diaper/incontinence supplies
- [ ] Hand powered radio
- [ ] Large first-aid kit
- [ ] Solar powered cell phone charger
- [ ] Spare chargers/adapters for the car
- [ ] Toiletries
- [ ] Ear Plugs
- [ ] Cash & extra copy of credit card

Have small denominations of cash. In an emergency electronic payment methods may not be working and vendors may not be able to make change.

Family Member Supplies continued

- [ ] Notebook & pen
- [ ] Chemical hand/foot warmers
- [ ] Blanket for each person

Pet Supplies

- [ ] Water Bowl & Food Bowl
- [ ] Water (A dog needs 1oz/lb/day)
- [ ] 3+ day supply of food per animal
- [ ] Supply of medications
- [ ] Leash
- [ ] Pet chews/toys
- [ ] Pet bedding/blanket

Home Preparations

Food Safety

Since formula is so critical for those with PA/MMA having the ability to keep formula cold is very important. A refrigerator only stays cold for up to 4 hours with the power off, a full freezer 48 hours, and a half full freezer 24 hours.

To help maintain the temperature in the event of a power outage, store partially full water bottles (water, soda) in the freezer to keep it at capacity and help maintain the temperature. They can be moved to the fridge or a cooler to keep formula cold.

To help to maintain the temperature, do not open the freezer or fridge unless necessary.

Disruption to water supply

If there is a possibility that there will be a disruption to the water supply during an emergency, fill any bathtubs with tap water, which can be scooped into a toilet bowl to flush the toilet. If there are small children in the home, LOCK THE BATHROOM DOOR to prevent accidental drowning.

Lighting & Heating Safety

Do not not plan for candles for light or outdoor or camping stoves/heaters for heat in an emergency. LED lanterns or flashlights are a much safer option than an open flame, especially with children or a person with developmental disabilities.

If your heating system is not working, then gather all household members, including pets into a smaller room with a door, preferably on the south side of the house. Bundle in layers and under blankets. An outdoor grill can be used out of doors to heat water. The water can be placed in heavy duty water bottles as a warmer.

Other Considerations

Small supply bag

You may want to keep a small amount of medication and formula at a location near to your house in case there is a crisis situation that occurs when you are not at home or in case you are not able to get your emergency supplies from your own home.

Communication tools

If your loved one has a disability that impacts the ability to communicate, you may want extra communication tools in with your emergency supplies.

Resupply & Review

It is a good practice to review and resupply your emergency supplies at least every 6 months. The time of year is determined by your area. For example, May 1st & November 1st for those prone to hurricanes. Put a reminder on your calendar to perform a quick “check and restock”.

6 mo	Check batteries in smoke alarms & CO detectors
6 mo	Check charge levels of fire extinguisher
6 mo	Check expirations on formula, food, and medication
12 mo	Replace water, it can develop an "off" taste with storage
12 mo	Get new prescription & lab orders
12 mo	Replace insurance information at plan renewal time
PRN	Update medical information, history, and supplies as needs change.

Moving

When you move to a new location update your evacuation plan & locations, contacts, and communicate with the school about emergency planning.

Aging out of Pediatrics

When you age out of pediatrics, update your medical contact's information AND legal documentation such as guardianship paperwork or HIPPA release forms for the affected individuals.

Information on How You Can Better Prepare for an Emergency

These are additional resources on Emergency Preparedness

FEMA Federal Emergency Management Agency: www.fema.gov
Preparedness Specific Resource Site: Ready.gov

The American Red Cross: How to Prepare for Emergencies: www.redcross.org/get-help/how-to-prepare-for-emergencies.html

A guide to for Preparing for Disaster for People with Disabilities: www.redcross.org/prepare/location/home-family/disabilities

The National Association of Realtors: Information on what to do and take when evacuating your home. www.houselogic.com/finances-taxes/home-insurance/home-evacuation-checklist

The National Fire Protection Association: Escape Route Planning Drawing & Fire Safety Information. https://www.nfpa.org/Public-Education/Staying-safe/Preparedness/Escape-planning

Notes:

How Your Community Can Better Prepare for an Emergency

Community Alerts

- Designate a community member to utilize a NOAA emergency radio for weather emergencies. (Midland Radio Company is developing a NOAA radio with only weather alert capabilities and no other features.) Have the radio on during all storms.
- Create a contact tree to alert community members about an emergency.
- Weather forecasts for Ohio area (216) 416-2900

Supply storage

Keep a supply of formula, L-carnitine and other medications on hand. Maintain a supply of fuel for refrigerators used for formula, medicine, and medical foods. Keep additional fuel for lamps for lighting.

Community Gathering place

Designate a community gathering place, such as a schoolhouse, where families can gather during an emergency. Maintain a list of all community members at that location. Utilize a bulletin board or chalkboard for community members to report in that they are safe or specify any needs that they have.

Schoolhouse

Maintain a supply of warm clothing, food, water, and fuel at the schoolhouse for the teacher and students in case of severe weather or other emergency during school hours.

DON'T FORGET

Place this sign in a prominent place like the fridge to remind you to get the needed items that are not in your Emergency Kit.

Examples: Scale, refrigerated medication, pump, Care Notebook

_____

_____

_____

_____

_____

_____

_____

_____

_____

_____

_____

_____

© 2023 Propionic Acidemia Foundation

DON'T FORGET

Place this page on the inside cover of your Handbook to remind you to get the needed items that are not in your Emergency Kit.

Examples: Scale, refrigerated medication, pump, Care Notebook

_____

_____

_____

_____

_____

_____

_____

_____

_____

_____

_____

_____

© 2023 Propionic Acidemia Foundation