

# Emergency Preparedness Tips For Glut1 Deficiency Patients

Prehospital, Emergency Department, and Family Resources

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# Emergency Preparedness Tips

**This guide will give tips on how to be better prepared for emergency situations with our Glut1 loved ones. Topics to be covered:**

- Prehospital/EMS expectations
- Emergency room expectations and important information to give the healthcare providers
- Useful documents and tools: how and when to present them
- Medical alerts bracelets, Help Belts, and emergency preparedness for family/emergency contacts
- Advocating for yourself and your child: resources that can be of help and communication tactics.

# Pre-Hospital/EMS Expectations

**The role of EMS personnel is to stabilize and bring the patient to the nearest emergency department.**

**What will they need to know?:**

- What happened, why 911 was called, current symptoms, story of what happened leading up to the event.
- Pertinent medical history
- Medications
- Do's and Don't's specific to you or your child's medical care
- What hospital system you typically seek your care
- They will typically Not need your specialist information, extended medical history information, X-ray, CT, MRI reports.
- EMS is generally instructed to bring their patients to the nearest hospital. It may not be your hospital of choice or where you generally seek care. You may ask to be brought to your hospital of choice but they may not be able to grant your request. Rest assured that once things are settled in the emergency department the doctor will be able to consult your specialist if needed.

# Emergency Room Expectations

- No one is ever ready or expecting an emergency situation to happen. The most that can be done is to know what to expect when you present to an emergency department and have resources for you or your child prepared ahead of time.
- You will be asked by multiple people what happened. You will often have to repeat your story a few times to a variety of staff: triage nurse, assigned staff nurse, and perhaps 1-2 doctors.
- The first step will be to make sure you or your child are stable. Emergency personnel (nurses or techs) may or may not start diagnostic tests prior to a doctor coming to evaluate you (blood work, X-rays).

# Emergency Room Expectations continued

- ER personnel have likely never heard of Glut1 Deficiency Syndrome. It will be very important that you tell them about Glut1 and they will likely depend on you for that information. When you are in the ER you will be the expert in Glut1 Deficiency and any treatments that you or your child are on.
- Be prepared with a list of Do's and Dont's. It is ok to communicate this information with staff. For example: If on the ketogenic diet: Do not give any medication with glucose in it. Do not feed my child anything unless it is approved by me. Must use G-tube for all food and medication administration.
- We often want our specialist (i.e. Neurologist) consulted right away. This will happen but will happen at a specific time which will likely not be right away.
- When consultants are called by the ER Physician the ER needs to be prepared with information prior to that call happening. This information will or may include the full evaluation from the treatment team, results of cat scans, lab work, or any other diagnostic studies that were ordered. If the ER team calls a consultant without complete information it could potentially delay care or provide the consultant with incomplete data that may be pertinent to care.

# Useful Documents and Tools

## How and when to present them

- You or your child may or may not have a lengthy medical history. It can often be overwhelming to relay this data in an emergency situation. There is also data that may be pertinent to Us but is not pertinent at the time one presents to the ER. It may be important after everything has been stabilized.
- A useful tool is to have a single sheet of data ready to give to EMS and ER staff.
- Where should I keep this single paper? You should have multiple copies in multiple places.
  - Laminate and keep on the fridge or in an easily seen place in the home.
  - Give to emergency contacts to have on hand.
  - Keep in your wallet or bag (emergency staff will go through personal belongings to acquire information in the event a patient is unconscious)
  - Give to the school nurse or school/work program coordinator.
  - Keep in the car or the glove compartment (If can also be used in the Help Belts which will be discussed in slides to come)
  - Take a picture of it on your phone

# Useful Documents and Tools

## How and when to present them

- What should be on this single sheet of paper?
  - Name, date of birth, drug allergies
  - Important Medical History or Conditions (Glut1 Deficiency, on ketogenic diet, seizures, movement disorders, cognitive or learning disability, heart or lung conditions) You do not need to list ALL medical history if lengthy, just history that is significant.
  - Brief Description of Glut1 Deficiency
  - Typical manifestations of Glut1 for you or your child (i.e. ataxia, seizures, no seizures, speech delay, PED's, etc)
  - Do's and Dont's for you or your child (for many this is related to treatment on the ketogenic diet). It may include medications that adversely affect you or your child.
  - Medication list with dosages (if you have a long list only include the most important. In an emergency situation medications like vitamin supplements do not need to be listed).
  - Resources and emergency contacts:
    - Emergency contact phone numbers
    - Contact info for your specialist (Name, number, hospital affiliation)
    - If you or your child are on the ketogenic diet: Name and contact info of dietician or if you know someone in the Glut 1 or Keto Community who can be of help in the event of an emergency.

# Useful Documents and Tools

## How and when to present them

- When to present more detailed data
  - Once the dust has settled with the emergency, staff will likely depend on you for more detailed data related to you or your child's history, typical treatments or plan of care, who needs to be consulted.
  - If you have a large file or binder of information regarding medical history and you have it with you that is ok. This may help you stay organized. It is often not a good idea to hand the binder to the ER staff and expect they will be able to go through the entirety it. Again, pertinent information is best so that they can stabilize, consult a specialist if needed, and create a plan of care (discharge from ER or admission to hospital). If admitted to the hospital a larger file of medical records would be handy for the admission team.



# Useful Documents and Tools

## Single Sheet of Data Examples

Name    Date of Birth            No Drug Allergies

\*\*\*\*\* has Glucose Transporter Type 1 Deficiency Syndrome. Glut1 Deficiency is a rare disease in which the body lacks a gene that does not allow glucose to pass through the blood brain barrier. (Low blood sugar of the brain, normal blood sugar in the rest of the body). Because the brain is starved for energy it cause many neurological symptoms.

-major manifestations of disease only occur when out of ketosis and can include movement disorders, ataxia, speech difficulty, dizziness, complex migraines, and behavioral outbursts.

At baseline \*\*\*\*\* walks with a normal gate, can speak clearly, and can perform all activities of daily living for his age. He has learning disabilities, ADHD, and anxiety.

On Ketogenic Diet for Treatment (3:1 ratio)

### Dont's

-Do no give any medications with Glucose

-Do not give anything to eat without asking the parents or emergency contact.

-Blood sugar ranges typically 60-80, Ketone levels large by urine and range in the blood from 2.5-5.5. \*\*\*\*\* may be in slight acidosis depending on ketone level.

### Do's

Consult Glut1 Specialist and Neurologist Dr. \*\*\*\*\*

Daily Medications \*\*\*\*\*

### Resources

-Emergency Contact Names and Numbers

-\*\*\*\*\* Neurology Glut1 Specialist\*\*\*\*\*

-Keto Parents Resources or Dietician -can calculate meals and give guidance if parents not available

# Useful Documents and Tools

## Single Sheet of Data Examples

Name\*\*\*\*\*

Date of Birth

No known drug allergies

Medical Problems:

Glut1 Deficiency Syndrome

-\*\*\*\*\* has a genetic brain disease in which he deficient in the gene that allows glucose to cross the brain barrier.

-This is treated by keeping \*\*\*\*\* in ketosis at all times on the ketogenic diet. (3:1 ratio)

-\*\*\*\*\* blood sugar is considered normal from 55-120. If he should drop below 55 and is symptomatic please give 5cc of apple juice or 8 cheerios and re-asses.

-\*\*\*\*\* betahydroxybutate (serum ketone) should remain at 3.0 and above

-DO NOT give any medications that contain any type of glucose. (ends in -ose)

-If glucose product has to be given this may worsen symptoms. Please consult parent or call Dr.\*\*\*\*, Neurologist at \*\*\*\*\*

-Major manifestations of \*\*\*\*\* disease only occur when out of ketosis and can include movement disorders, seizure like episodes, ataxia, dizziness, migraine, behavioral outbursts.

-DO NOT feed unless authorized to do so. All food must be calculated and weighed. If \*\*\*\*\* is hungry \*\*\*\*\* may not eat until food provided by family, keto resources, or directions given by dietician at keto program.

Medications:

-Potassium Citrate 540mg BID

-multiple vitamin supplements

Resources:

Dr \*\*\*\*\* , Division of Neurology at \*\*\*\*\*

Phone Number

Emergency Contacts

Keto Resources-can calculate meals keto meals and give guidance if parents not available

\*\*\*\*\*

# Useful Documents and Tools

## Single Sheet of Data Examples

TB, 99/99/2000

Drug Allergy: None

ALERT: Do not use Lactate or Glucose in IV. Plain Saline is okay. Patient is on a modified atkins diet to help control symptoms associated with Glucose Transporter Deficiency

-Important Medical History or Conditions: Glucose Transporter Deficiency Type 1, Movement Disorders including Dystonia and Chorea, and Intellectual Disability

-Brief Description of Glut 1 Deficiency - Glucose transporter type 1 deficiency syndrome (Glut1DS) is a rare genetic metabolic disorder characterized by deficiency of a protein that is required for glucose (a simple sugar) to cross the blood-brain barrier and other tissue barriers.

-Typical manifestations of Glut 1 for TB: ataxia, speech and motor delays, movement episodes

-Medications:

Diamox 125 mg, 3 tablets 2x/day

Carbidopa-Levodopa 25-100 tables, 2 in morning, 2 at noon and 3 at 5 PM

Doxycycline IR-DR 40 mg - 1 tablet 1x/day

Resources and emergency contacts

AB & phone

GB & phone

Sister & Phone

Sister2 & Phone

-Contact info for your specialist

Pediatrician

Local Neurologist

2nd Neurologist

# Emergency Info for Emergency Contacts

**Emergency contacts and emergency caregivers may need help in the event you as a Glut 1 parent or patient is not available to give guidance or care to your child or self. There are some things you can do to prepare.**

- Have a binder of instructions for an emergency caregiver that has the following information and whatever else you may think pertinent to yourself or child
  - Any diet related instructions:
    - If on the keto diet or MAD-some typical meals and snacks that are pre-calculated, basic instruction on how to use the scale, weigh food, or prepare meals, grocery stores that typical items are bought, foods that are preferred. Beverages that are allowed.
    - If on regular diet: things that one can and can not have. Foods that may trigger symptoms.
    - Special oils or other additives used for or in conjunction with meals
    - G-tube or special feeding methods if any
    - Allergies to foods if any
  - Medication schedules and pharmacy info
  - Ketones and blood sugar norms if applicable
  - Daily routine info and schedules: sleeping habits, bathing, school info-bus times, helpful hints for behavioral challenges if needed.
  - How to get into your home: back up keys, garage codes, etc.
  - Other emergency resources: doctors, dietitians, fellow parent resources, school contact info.
  - It is good to let your emergency contacts know where this info is in your home and go over it with them so they are familiar. Your emergency caregiver may often have questions that prompt you to remember info that would be useful in this binder or packet of information.

# Medical Alert Bracelets

**Medical alert bracelets help emergency personnel or bystanders quickly identify an individual has a medical problem. Medical alert bracelets are not meant to provide a large amount of medical information. There are many different brands of medical alert bracelets. Not one is better than the other but is based on personal preference. Some bracelets have one side for personalized information and some have 2 sides. Some companies have medical alert necklaces. There are no companies, as of yet, that come with standardized terminology for Glut1 Deficiency. You have to customize based on personal preference and need.**

- What should a medical alert bracelet say?
  - Name and emergency contact phone number
  - Simple information: pick one or two important pieces of information you would want relayed. Example: Glut1 Deficiency, Glut1 DS, epilepsy, ketogenic diet, no glucose products. (Abbreviations are fine if you are short on space)

# Help Belts

**Help Belts are a seat belt cover that can be used with any age child or adult. You would place the Help Belt around the seat belt where the individual with Glut1 typically sits. You can have one for each car that the individual typically travels in. This can be used on back packs, strollers, purse handles, baby seats, or anything it will fit on. Inside of the Help Belt would be the perfect place to put the “One sheet of Important Information” as discussed in previous slides.**



# Help Belts Continued

## Where can I order a Help Belt?

- ETSY- <https://www.etsy.com/shop/HELPBelts>
- <https://www.helpbelts.com>

They do create custom Help Belts. You would need to contact them to place the order at [helpbelts@gmail.com](mailto:helpbelts@gmail.com)

- I am not sure if this company ships internationally. There may be similar companies in other countries. This is also something someone may be able to make on their own with supplies from a fabric or craft store.

# MediPal

## MediPal is another company with a similar product to Help Belt

- <https://medipal.com>
- Amazon carries this product as well.



The flyer features the MediPal logo and tagline 'Saving Time Saves Lives'. It includes two testimonials: one from Mathew Price, a paramedic in San Diego, and another from Cande Stone, a parent of a child with special needs. Below the testimonials is a list of four key features: the form is a free PDF that can be updated anytime; it wraps around seatbelts with velcro; it has a reflective trim for night visibility; and it provides peace of mind for family members. The flyer also contains two QR codes, the website URL 'www.medipal.com', and the Amazon logo. At the bottom, it mentions 'bulk &amp; nonprofit pricing available', a BBB logo, and copyright information for MediPal Inc. (2020-2021) with Patent No. D681940.



# Yellow Dot Program

**Some states in the United States have adopted an initiative called the Yellow Dot Program. This program is intended to help first responders identify individuals with medical problems when they are in a car accident and not able to relay medical information on their own. The “yellow dot” or sticker would be placed in the outside of the vehicle and notify EMS or paramedics to look in the glove box for an envelope of medical information.**

- Please refer to your state website or your local police to see if your state participates in this initiative. In many states, only certain counties have implemented the program. Here are the states that have known programs in place.
- Pennsylvania, Tennessee, New York, Kansas, New Jersey (County based), Maine, Virginia.

# Communication Tactics in the ER

**You are your best advocate for yourself and your loved ones. It is always ok to ask questions and provide the necessary information relevant to yourself or your loved one.**

- Always ask questions. It is ok to ask what is being done, what medication is being given, what is in a medication, or what the plan of treatment or care is. These are all normal questions that every patient should ask.
- You are the expert in Glut1 Deficiency. You will have a knowledge base about Glut1 that no one else has in the ER. Most medical professionals will be thankful for the knowledge you can provide and use you as a resource.
- Unfortunately, you may occasionally run into staff who don't take you or your knowledge seriously. Stand your ground calmly and perhaps try to explain in a different way. Use resources available on the web if needed. In this instance it might be a good time to ask the ER physician staff to consult with your specialist for assistance.
- If you don't have an advocate with you and you are having trouble with communication or medical care in the ER, you can ask for your nurse, a social worker, or last resort the charge nurse. There is always someone who will listen to your concerns.

# Communication Tactics in the ER

## Continued...

- If you or your child will be in the emergency room for a lengthy amount of time for extensive testing or admission is required, it will now be a good idea to present the larger files of information if you have it on hand.
- The admission team will likely sit with you and/or your child for a more lengthy amount of time getting a full history and diving deeper into the medical history, previous testing, daily medications, etc.
- Not many people remember in emergency situations but having your medication bottles or a complete list will be very helpful. Many people use their smart phones to keep pictures of medication bottles and lists.

**Thank you and please feel free to reach out if  
you have any questions or need of support.  
[mrebbecchi@g1dfoundation.org](mailto:mrebbecchi@g1dfoundation.org)**