The Mast Cell Disease Society

Emergency Room Response Plan

for **Adult Patients** with Mast Cell Diseases

Patient Name

Date of Birth

Today's Date

Anaphylaxis in a Patient with Mast Cell Disease

*Please note: These recommendations may differ from general guidelines for anaphylaxis in that they may include additional considerations specific for the Mast Cell Disease patient.

PLACE PATIENT IN RECUMBENT POSITION AND ADMINISTER

(Please check all that apply)

- ☐ **Epinephrine** 0.3 mL of 1:1000 IM (auto injector preferred*). Repeat 3x at 5-minute intervals if blood pressure <90 systolic
- ☐ **Oxygen** by mask or nasal cannula
- ☐ If trigger is present, remove trigger from the reaction if possible
- ☐ **Benadryl** (Generic: diphenhydramine) 25-50 mg intravenously **(slow IV push)** every 2-4 hours, or **cetirizine** 10 mg intravenously, or **Hydroxyzine Hydrochloride** 25 mg intramuscular dose every 2-4 hours
- ☐ **IV Fluids** 1-2 L of Normal Saline until SBP is >90
- ☐ **Albuterol** by nebulization / Alternatively, Racemic Epinephrine can be given by nebulization
- ☐ **Solu-Medrol** (Generic: methylprednisolone) 0.5-1 mg/kg X1 and repeat 1-2 hours later if SBP below 90
- ☐ **Glucagon**** for patients on beta-blockers who do not respond to Epinephrine or who have cardiac disease that make continued boluses/treatment of Epinephrine contraindicated
- ☐ **Optional:** Prednisone 1mg/kg orally

Call 911 and take the patient to the closest emergency room.
Please ask for a serum tryptase level to be drawn within 30 minutes of symptom onset.

A special 'Thank you' to Mariana Castells, MD, PhD, Director of the Boston Center of Excellence for Mastocytosis, Brigham and Womens' Hospital, Boston, MA and Professor of Medicine, Harvard University; and Joseph Butterfield, MD, Director of the Mayo Clinic Program for Eosinophilic and Mast Cell Disorders, Mayo Clinic, and Professor of Medicine, Mayo Medical School, Rochester, MN for their contributions to the revision of the protocols.

*Auto injector avoids errors due to similar appearance of Epinephrine and other Ampules

**NOTE: the use of Glucagon is associated with a risk of nausea and vomiting. This can be due to the risk of increased cardiac oxygen demand, arrhythmias, coronary artery vasospasm. Recall that if the patient is on a non-selective beta blocker, administration of epinephrine will give a nearly pure alpha adrenergic effect, resulting in a spike in BP followed by triggering of carotid and aortic baroreceptors and a reflex increase of vagal tone resulting in bradycardia. Selective beta-1 blockers are less of a problem because the beta-2 receptors are not blocked and can offset the alpha receptor effect somewhat, lowering the risk of a spike in BP etc. (Joseph Butterfield, MD)

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Pre-Medication Plan

For major and minor procedures/surgery and for radiology procedures, including ultrasound, with and without dyes.

ADMINISTER

At 12 hours and 1 hour prior to surgery or dye administration give:
(please select all that apply)

- ☐ **Benadryl** (Generic: diphenhydramine) 25 mg orally or IV, or **Atarax** (Generic: hydroxyzine) 25 mg orally, or equivalent nonsedating antihistamine. Examples: Zyrtec (cetirizine) 10 mg IV or PO may be used as a long-acting alternative, Claritin (loratidine), Allegra (fexofenadine)
- ☐ **Pepcid** (Generic: famotidine) 20 mg orally. Another H2 antagonist is tagamet/cimetidine
- ☐ **Singulair** (montelukast) Examples: Accolate (zafirlukast), Zyflo CR (zileuton)

Medications to Be Avoided

AVOID

- Any medication to which the patient has a listed allergy
- Aspirin and nonsteroidal anti-inflammatory medicines if the patient has a known adverse reaction
- Morphine and codeine derivatives (fentanyl is the preferred opioid)
- Vancomycin given IV. Oral route may be tolerated in some patients.
- Quinolones

Please note this is a standardized protocol. Each protocol should be personalized for the patient with the help of a mast cell specialist. Some institutions/medical departments have their own protocols. **Be sure to discuss IN ADVANCE with your physicians and those departments.**

Additional Orders

Physician Signature

Date

A special 'Thank you' to Mariana Castells, MD, PhD, Director of the Boston Center of Excellence for Mastocytosis, Brigham and Womens' Hospital, Boston, MA and Professor of Medicine, Harvard University; and Joseph Butterfield, MD, Director of the Mayo Clinic Program for Eosinophilic and Mast Cell Disorders, Mayo Clinic, and Professor of Medicine, Mayo Medical School, Rochester, MN for their contributions to the revision of the protocols.

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Emergency Room Response Plan

for **Pediatric Patients** with Mast Cell Diseases

Patient Name

Date of Birth

Today's Date

Anaphylaxis in a Pediatric Patient with Mast Cell Disease

*Please note: These recommendations may differ from general guidelines for anaphylaxis in that they may include additional considerations specific for the Mast Cell Disease patient.

PLACE PATIENT IN RECUMBENT POSITION AND ADMINISTER

(Please check all that apply)

- ☐ **Epinephrine** 0.15 mL of 1:1000 IM (Pediatric Auto injector preferred*). Repeat 3x at 5-minute intervals if blood pressure is <90 systolic
- ☐ **Oxygen** by mask or nasal cannula
- ☐ If trigger is present, remove trigger from the reaction if possible
- ☐ **Benadryl** (Generic: diphenhydramine) 12.5-25 mg intramuscular or intravenously **(slow IV push)** every 2-4 hours, or **cetirizine**: Children over 6 months of age 2.5 mg IV. Children ages 5-10: 5-10 mg IV depending on severity of symptoms. Children over 12 years of age: 10 mg IV push over 1-2 minutes
- ☐ **IV Fluids** 1-2 liters of Normal Saline for 1-2 hrs until Systolic BP is >90
- ☐ **Albuterol** by nebulization / Alternatively, Racemic Epinephrine may be used
- ☐ **Solu-Medrol** (Generic: methylprednisolone) 0.5-1 mg/kg X1 and repeat 1-2 hours later if SBP below 90
- ☐ **Glucagon** for patients on beta-blockers who do not respond to Epinephrine or who have cardiac disease that make continued boluses/treatment of Epinephrine contraindicated
- ☐ **Optional:** Prednisone 1mg/kg orally

Call 911 and take the patient to the closest emergency room.
Please ask for a serum tryptase level to be drawn within 30 minutes of symptom onset.

*Auto injector avoids errors due to similar appearance of Epinephrine and other Ampules

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Pre-Medication Plan

For major and minor procedures/surgery and for radiology procedures, including ultrasound, with and without dyes.

ADMINISTER

At 12 hours and 1 hour prior to surgery or dye administration give:
(please select all that apply)

- ☐ **Benadryl** (Generic: diphenhydramine) 12.5-25 mg orally or IV, or **cetirizine** (Zyrtec) 2.5 mg for 6 months and over; 5 mg for ages 5-10; 10 mg for age 12 and over; or **loratadine** (Claritin) 2.5 mg for age 2-5; 5 mg for ages 6-11; 10 mg for ages 12 and over
- ☐ **Pepcid** (Generic: famotidine) 10 mg orally
- ☐ *Consider* **montelukast** 4 mg orally

Medications to Be Avoided

AVOID

- Any medication to which the patient has a listed allergy
- Aspirin and nonsteroidal anti-inflammatory medicines if the patient has a known adverse reaction
- Morphine and codeine derivatives (fentanyl is the preferred opioid)
- Vancomycin given IV. Oral route may be tolerated in some patients.
- Quinolones

Please note this is a standardized protocol. Each protocol should be personalized to the patient's needs with the help of a mast cell specialist. Some institutions/medical departments have their own protocols. **Be sure to discuss IN ADVANCE with your physicians and those departments.**

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Additional Orders

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______________________________ ______________________________
Physician Signature Date

A special 'Thank you' to Mariana Castells, MD, PhD, Director of the Boston Center of Excellence for Mastocytosis, Brigham and Womens' Hospital, Boston, MA and Professor of Medicine, Harvard University; and Joseph Butterfield, MD, Director of the Mayo Clinic Program for Eosinophilic and Mast Cell Disorders, Mayo Clinic, and Professor of Medicine, Mayo Medical School, Rochester, MN for their contributions to the revision of the protocols.

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Lab Tests to Be Drawn in ER

During Mast Cell Activation Event

Please see note under tests about having your own physician sign this form for follow-up

1. Serum Tryptase—upon arrival in the ER and three hours later. If hospital lab is outfitted with the Immunocap system, serum tryptase results are obtained in 4 hours or less.
2. 24-hour or spot urines for:
   - n-methyl histamine
   - prostaglandin D2(PGD2)
   - 11-beta prostaglandin F2 alpha
   - Leukotriene E4
3. Complete chemistry panel
4. CBC with differential

You MUST have your allergist or primary care provider sign the bottom of this form stating that **he or she will be responsible** for the follow-up on the 24-hour urine collections. Otherwise, the ER physicians will be reluctant to order them since they cannot be sure of follow-up care. Remember to contact your physician for follow-up after discharge.

I agree to provide follow-up care for my patient, ______________________________
and will obtain the results of the 24 hour or spot urine collections that were initiated in the emergency room setting to provide appropriate care based on the results.

____
Printed Name of Physician

______________________________ ______________
Signature of Physician Date

____
Contact Address

______________________________ ______________________________
Phone Number Fax Number

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Medications to Use and Avoid

Quick Reference Guide

Medications _to avoid_ or _use with caution_ in patients with mast cell disease in emergency situations

Please note: *Some of the Medications to Avoid may be given if absolutely necessary, if given with a prep to stabilize mast cells. Please refer to one of our mast cell experts for instructions.*

Medication Type	Avoid or Use With Caution	Medications That Are Typically Tolerated
General Medications	• alcohol • amphotericin b • dextran • dextromethorphan • polymyxin B • quinine • vancomycin IV • alpha-adrenergic blockers • beta-adrenergic blockers	• calcium channel blockers • centrally acting alpha 2 adrenergic stimulants • aldosterone antagonists • Oral doses of Vancomycin may be tolerated in some cases.
Pain Medications	• opioid narcotics (may be tolerated by some individuals) • Toradol (ketorolac) • Non-steroidal anti-inflammatory drugs (unless the patient is already taking a drug from this class)	• fentanyl [may require adjunct treatment with Zofran (ondansetron)] • tramadol
General Anesthetics	• atracurium • doxacurium • rocuronium • mivacurium	• pancuronium • vecuronium
Local Anesthetics	• benzocaine • chloroprocaine • procaine • tetracaine	• bupivacaine • lidocaine • mepivacaine • prilocaine • levobupivacaine • ropivacaine
Intraoperative Induction Medications		• ketamine • midazolam • propofol
Inhaled Anesthetics		• sevoflurane

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Helpful Links

To Contact TMS Nurses (Non-Emergency Only):

nurses@tmsforacure.org

For Additional Patient Resources Please Visit:

tmsforacure.org/er

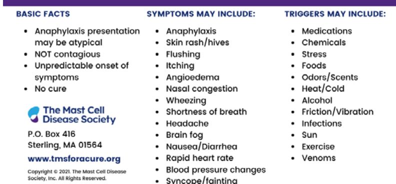
For the AAAAI Anaphylaxis Action Plan:

https://www.aaaai.org/Aaaai/media/MediaLibrary/PDF%20Documents/Libraries/Anaphylaxis-Emergency-Action-Plan.pdf

Medic Alert Bracelet/Jewelry

When deciding what to put on your medical jewelry, the first word should always be: **Anaphylaxis!**

1. Systemic mastocytosis, systemic mast cell disease, mast cell activation syndrome or hereditary alpha tryptasemia.
2. If, and only if, you are on a beta blocker, add the following:
   a. After Epinephrine, give Glucagon.
3. Medication Allergies: if you have 1 allergy, then list it. If you have multiple, then state “drug allergies”.
4. Food Allergies: if you have 1 food allergy, then list it. If you have multiple, then state “multiple food allergies”.
5. Latex Allergy, if you have one.
6. Medication, food, and latex allergies can be combined.
7. Next, add other illnesses: diabetes, dysautonomia, EDS, angina, thyroiditis, etc.



**EMERGENCY INFORMATION
MAST CELL DISEASES**

BASIC FACTS

- Anaphylaxis presentation may be atypical
- NOT contagious
- Unpredictable onset of symptoms
- No cure

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SYMPTOMS MAY INCLUDE:

- Anaphylaxis
- Skin rash/hives
- Flushing
- Itching
- Angioedema
- Nasal congestion
- Wheezing
- Shortness of breath
- Headache
- Brain fog
- Nausea/Diarrhea
- Rapid heart rate
- Blood pressure changes
- Syncope/fainting

TRIGGERS MAY INCLUDE:

- Medications
- Chemicals
- Stress
- Foods
- Odors/Scents
- Heat/Cold
- Alcohol
- Friction/Vibration
- Infections
- Sun
- Exercise
- Venoms

Visit **www.tmsforacure.org/er** to download additional resources

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