Emergency Room Response Plan
for Adult Patients with Mast Cell Diseases

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)
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 non-sedating 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

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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

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**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.

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**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.

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

Additional Orders

__________________________

Physician Signature

__________________________

Date

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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 ____________________________
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.

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

References:
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.

For the AAAAI Anaphylaxis Action Plan:

To Contact TMS Nurses (Non-Emergency Only):
nurses@tmsforacure.org

For Additional Patient Resources Please Visit:
tmsforacure.org/er

Visit www.tmsforacure.org/er
to download additional resources
References:


