Precautions for mastocytosis

1. Mastocytosis patients may have individualized drug sensitivities and symptoms of anaphylaxis that are atypical.
2. Mast cell activation symptoms can quickly disintegrate into anaphylaxis, and simple symptoms should never be overlooked nor should treatment be delayed.
3. Additional doses of H1 and H2 antihistamines may be needed to control itching, abdominal pain, nausea, diarrhea and bloating even after acute anaphylaxis has responded to treatment.
4. Control the patient’s environmental temperature and stress to avoid setting off a mast cell reaction again.
5. If the patient’s symptoms flare after the initial reaction is treated, check all parenteral medications to make sure that none of them contain alcohol or preservatives.
6. Any blockers or drugs that interfere with epinephrine or that contribute to histamine release should be withheld.
7. If emergency surgery is needed, mastocytosis patients should receive H1 and H2 antihistamines, steroids, mast cell stabilizers and leukotriene blockers preoperatively.

What else should I know?

Triggers are unique to each patient. If a patient tells you that a certain drug, substance or environmental factor is a mast cell trigger for them, believe the patient even if it does not seem plausible.

References


Castells, MC. Anaphylaxis and Hypersensitivity Reactions. Springer; 2011


Norred, CL. Anesthetic-induced anaphylaxis. AANA J 2012:April, in press.


What are mast cell diseases?

Mast cell diseases are disorders of mast cell proliferation (mastocytosis) and activation (MCAD) affecting children and adults. Mastocytosis can affect skin and internal organs such as the bone marrow, GI tract, liver and spleen. Most patients with mastocytosis have cutaneous or indolent (benign) systemic forms, but aggressive disease (similar to malignancy) may have hematologic disorders. Mast cell patients may have unpredictable symptoms that require anti-mediator therapy. Diagnosis of mastocytosis is confirmed by a bone marrow or skin biopsy. MCAD patients do not fulfill all criteria for mastocytosis but exhibit symptoms, may or may not have increased measurable mast cell mediators (commonly tryptase, histamine or its metabolites) during or shortly after an attack and do respond to anti-mediator therapy.

Patients with mast cell disorders are more likely to experience anaphylaxis than the general population (approximately 33%) which can be unprovoked or triggered by varied stimuli including hymenoptera venom, medication, physical or emotional stress. Additionally, 65% of their anaphylactic episodes are severe.
Drugs to Administer with Caution

- Analgesics: Nonsteroidal anti-inflammatory drugs. AVOID morphine or codeine derivatives.
- Anesthetics: Ester local anesthetics.
- Antibiotics: Amphoteracin or polymyxin B. AVOID vancomycin.
- Cardiovascular: α- and β-adrenergic receptor blockers.
- Muscle Relaxants: Atracurium, rocuronium or succinylcholine.
- Preservatives: Ethanol or other alcohol solutions or metabisulfites.
- Other: Dextran, dextromethorphan, quinine.

Anaphylaxis Severity

Anaphylaxis symptoms can occur on a continuum:
- Grade I: Cutaneous signs such as hives or rash.
- Grade II: Cutaneous signs, and hypotension, tachycardia, presyncope, dyspnea or GI distress.
- Grade III: Profound hypotension, bradycardia or tachycardia, cardiovascular collapse, confusion, bronchospasm, hypoxia (SaO2 <92) and GI distress.
- Grade IV: Pulseless electrical activity (PEA) or cardiac arrest.

Call for Help

Anaphylaxis treatment requires teamwork to start 2 IVs (16-18G) to administer fluids rapidly. Place the patient in Trendelenberg position continuously.

Reassess the diagnosis and monitor EKG, SaO2 and ABGs. CPR/ACLS may be necessary. Draw a serum tryptase level at 30-120" after symptom onset.

Epinephrine

Epinephrine may be titrated according to the severity of anaphylaxis symptoms:
- Grade I: None.
- Grade II: 10-20 mcg IV.
- Grade III: 100-200 mcg IV Q 1-2 min; 1-4 mcg/min infused.
- Grade IV: 1-3 mg to 3-5 mg IV Q 3 min; 4-10 mcg/min infused.

If no IV has been started, epinephrine should be given rapidly by IM injection (vastus lateralis) or endotracheal tube. The dose is 0.3ml of 1 mg/ml solution repeat X3 at 5 min intervals for systolic BP <90 mmHg.

Bronchospasm and Angioedema

If the patient has hypoxia, myocardial ischemia, or needs epinephrine or β agonists he or she could need 100% O2 or intubation and ventilation.

Drugs for bronchospasm or angioedema include:
- Albuterol: 2.5-5 mg in 3 ml NS nebulized or 100-200 mcg IV.
- Ipratropium: 500 mcg in 2.5 ml NS nebulized.
- Terbutaline: 250-500 mcg subcutaneous.
- H2 blockers: Diphenhydramine 1-2 mg/kg or 25-50 mg IV/IM/PO (under age 12: 12.5-25 mg) or Hydroxyzine 25-100 mg IV/IM or 25 mg PO every 2-4 hours (pediatric: 12.5-25 mg).
- H1 blockers: Ranitidine 1 mg/kg IV/IM (pediatric IV: 12.5-50 mg).
- Corticosteroids: Hydrocortisone 0.5-1 mg/kg IV initially, then 2.5 mg/kg every 4-6 hours.

Cardiac Arrhythmias

- Tachycardia should be treated first with IV fluids then ACLS protocols should be followed.
- Bradycardia should also be treated with IV fluids prior to ACLS protocols with atropine 0.4-1 mg IV, followed by dopamine 400 mg in 500 cc NS at 2-20 mg/kg/min IV, or epinephrine 1:1000, 0.1-0.3 mg (in 10ml NS IV). Cardiac pacing may be indicated for severe unresponsive bradycardia.
- PEA or arrest should be treated with IV fluids and CPR/ACLS with epinephrine up to 1-3 mg/ 3-5 min. IV/IM, vasopressin 40 U IV, or atropine 0.4-1 mg IV.

Hypotension

Because anaphylaxis can cause rapid profound vasodilation, hypotension should be treated with:
- NS 5-10 cc/kg in 5 min, up to 30cc/kg IV.
- Hetastarch* 500cc IV.
- Vasopressin 1-2 U IV.
- Dopamine 400 mg/500 cc NS at 2-20 mg/kg/min IV.
- Norepinephrine 0.05 mcg/kg/min IV.
- Glucagon 1-5 mg or 20-30 mcg/kg IV for unresponsiveness or β blockade; infusion: 5-15 mcg/min IV.

Continuation of Care

Anaphylaxis can reoccur for 72 hours. An ICU bed & ventilator and continued treatment with vasopressors, H1 & H2 blockers, or corticosteroids may be needed. Laboratory tests including serum tryptase, specific IgE assay and urine N-methyl histamine should be monitored after anaphylaxis. Record events and consult an immunologist.

TMS is a Lay Organization member of the American Academy of Allergy, Asthma, and Immunology and a 2011 member of the National Organization of Rare Disorders.