COMMON TREATMENTS

H1 antihistamines: Cetirizine, Fexofenadine, Loratadine, Levocetirizine

H2 antihistamines: Famotidine, Cimetidine

Leukotriene inhibitors: Montelukast

NSAIDs *(can cause mast cell activation reaction for some patients; use with caution)*

Mast Cell Stabilizers and Other Treatments:

- Cromolyn sodium (oral and/or inhaled)
- Ketotifen
- Quercetin
- Biologics
- Glucocorticoids: Prednisone (Brief Burst), etc.
- Diphenhydramine/Hydroxyzine (1st generation H1 Antihistamine-Rescue Medication)
- Benzodiazepines

TREATMENTS FOR SPECIFIC SITUATIONS

- Epinephrine
- Pentosan
- Cladribine (used for Mastocytosis)
- Avapritinib (used for Mastocytosis)
- Intron (Interferon Alfa-2B used for Mastocytosis)
- PPI's
- Antiemetics
- Electrolytes

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MAST CELL DISEASE IS COSTLY.

Oftentimes, patients are faced with deciding between treatment needs versus living needs. Super T's provides support through our grant program, which helps to offset financial burdens for patients. Please refer your patients to our website for further information.

supertmastcell.org
supertsmcfoundation@gmail.com
610.265.4373

SUPER T'S MAST CELL FOUNDATION

MAST CELL DISEASE

Mast cells are a normal part of our immune system. They release chemicals such as histamine, prostaglandins, and more in a process called degranulation. In mast cell disease, these chemicals often become excessive and can cause multi-system inflammatory reactions, ranging from mild to life-threatening. MCAD is an umbrella term for all Mast Cell Activation Diseases. In mast cell disease, one can have too many mast cells (Mastocytosis) or have mast cells erupt at inappropriate times (Mast Cell Activation Syndrome (MCAS)). Triggers can vary and are often different from traditional allergens. There are numerous supportive treatments for patients, but there is no cure.

Super T's Mast Cell Foundation is here to help guide you on your journey!

TYPES OF MCAD

Mast Cell Activation Syndrome (MCAS)

Monoclonal Mast Cell Activation Syndrome (MMAS)

Hereditary Alpha Tryptasemia (HAT)
(due to extra copies of the alpha-tryptase gene)

Cutaneous Mastocytosis (CM)

- Maculopapular Cutaneous Mastocytosis (MPCM) also called Urticaria Pigmentosa or Telangiectasia Macularis Eruptiva Perstans (TMEP)
- Diffuse Cutaneous Mastocytosis (DCM)
- Cutaneous Mastocytoma

Systemic Mastocytosis (SM) and variants:

- Indolent Systemic Mastocytosis (ISM)
- Systemic Mastocytosis with Associated Hematologic Disease (SM-AHD)
- Mast Cell Sarcoma
- Aggressive Systemic Mastocytosis (ASM)
- Smoldering Systemic Mastocytosis (SSM)
- Mast Cell Leukemia (MCL)

COMMON TRIGGERS*

- Heat, cold, or rapid temperature changes
- Scents, fragrances, and chemicals
- Skin friction
- Vibration
- Ultraviolet Light (Sunlight or Fluorescent)
- Medications (especially "inactive" ingredients, a.k.a excipients, such as fillers, binders, preservatives, and dyes.)
- Infections
- Exercise (often specific exercise(s) and sometimes only beyond certain extents)
- Food (usually just specific foods, possibly high histamine foods)
- Medical Procedures (usually just certain procedures in the individual patient)
- Stress (physical/phycological/emotional)

**Not an exhaustive list.*
A patients full set of triggers is usually unique to that patient.

COMMON SYMPTOMS*

**Each patient may present symptoms.*

Skin: flushing, rashes, hives, itching, angioedema, dermographism

Gastrointestinal: abdominal pain and bloating, reflux/heartburn, diarrhea, constipation, nausea, vomiting, cramping

Cardiovascular: high or low blood pressure, abnormal heart rhythm, rapid or slow heart rate, palpitations, dizziness, fainting (syncope)

Respiratory: sinus nasal congestion, mouth/throat irritation, discomfort with breathing, wheezing

Neuropsychiatric: headache/migraine, tingling/numbness, tremors, neuropathy, insomnia and other sleep disturbance(s), concentration and memory difficulties, anxiety, depression, and other cognitive and dysautonomic symptoms

Constitutional: Fatigue, malaise, cold/heat intolerance, flushing, sweats, weight loss or gain, chemical/physical environment sensitivities

Musculoskeletal section: joint laxity/hypermobility, osteopenia/osteoporosis, arthritis, joint pain, bone pain

Anaphylaxis (not required for diagnosis or present in every patient)

Each patient may present with symptoms that may be seen in any body system and are driven by the inappropriately released mast cell mediators.

- **Ophthalmologic:** irritated eyes, episodes of vision problems
- **Otologic/osmic:** tinnitus, odor reactions
- **Oral/oropharyngeal:** burning mouth, sores, throat irritation, post nasal drip
- **Lymphatic:** swelling (episodic swelling of lymph nodes)
- **Genitourinary:** endometriosis, interstitial cystitis, various problems with pregnancy, genital tract pain, abnormal uterine bleeding
- **Endocrinologic/metabolic:** thyroid dysfunction, electrolyte abnormalities, dysmenorrhea, elevated liver function tests
- **Hematologic:** easy bruising, abnormalities in red cell, white cell and platelet counts, anemia
- **Immunologic:** hypersensitivity reactions, autoimmunity, impaired healing, increased susceptibility to infections, abnormal immunoglobulins

TESTING

USED FOR DIAGNOSIS AND TO GUIDE TREATMENT+

24-hour and random urine testing[1]:

- Histamine, N-methylhistamine
- Prostaglandin D2, 2,3-dinor-11-beta-prostaglandin-F2-alpha
- Leukotriene E4

Blood testing:

- Serum tryptase[2]
- Plasma Heparin[1]
- Chromogranin A (CgA)
- Tryptase CNV[2,3]
- KIT D816V Mutation Analysis[4]
- Prostaglandin D2
- Histamine

Biopsies

(For Mastocytosis):

- Bone marrow
- Skin punch

(For MCAS):

- Gastrointestinal[5]

Skin prick test for allergies in MCAD patients can be unreliable.

+There is some disagreement about preferred testing among specialists

[1]Sample must be kept cold at all times for accurate results

[2]A normal tryptase level does NOT rule out a MCAD diagnosis

[3]Gene by Gene test for HAT

[4]Common mutation in SM

[5]Gastroenterology can be helpful for diagnosing.