

Welcome!

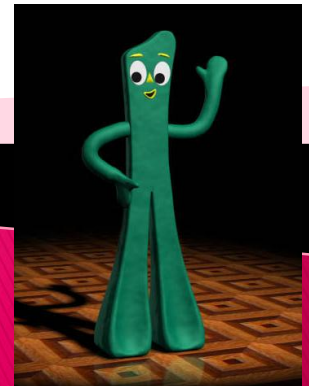


Capital Area
Ehlers–Danlos Syndrome
Support Group

Capital Area Ehlers–Danlos Syndrome Support Group

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Topics

- ▶ Quick tips on healthy teeth and gums
 - ▶ MCAS treatment
 - ▶ Skin issues
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Principles for Treating MCAS

- ▶ Principle #1: Therapeutic goal is to feel significantly better than pre-treatment baseline *most* of the time
- ▶ Principle #2: Identification of the optimal regimen for the individual MCAS patient takes persistence and patience.
- ▶ Principle #3: Whenever possible, try to change only one thing in the patient's regimen at a time.

Principles for Treating MCAS

- ▶ Principle #4: Start trials with least expensive medications first; if they aren't helpful, then escalate to more expensive options (until ability to identify mutations/mediators in the individual as targets)
- ▶ Principle #5: When a stabilized MCAS patient destabilizes, review recent history (new medications, exposures, etc.) carefully to try to identify what changed to cause the worsening.

Principles for Treating MCAS

- ▶ Principle #6: *No* medication is active ingredient only. Inactive ingredients have the potential to trigger a flare, and the same active ingredient in a different formulation may be helpful.
- ▶ Principle #7: “The simpler the better.” Discontinue treatments which have not demonstrated *clear* benefit.

Principles for Treating MCAS

- ▶ Principle #8: Failure to improve on a drug of a certain class doesn't mean all of the medications in that class will not be helpful.
- ▶ Principle #9: Identify and avoid triggers, if possible. Consider desensitization if available.
- ▶ Principle #10: Preventive measures such as handwashing, immunizations (if tolerated), tobacco avoidance, health screening, etc., should be followed.

Principles for Treating MCAS

- ▶ Principle #1 1: Eat a normal, balanced, healthy diet if you can tolerate it.
 - If you wish to try a dietary change as an intervention, change only *one* thing at a time (Principle #3).
 - Keep a diet and symptom diary, including time of food intake and symptoms, so you can more clearly identify triggers or improvement.

T M S



THE MASTOCYTOSIS SOCIETY

Mast Cell Diseases

<https://tmsforacure.org/>

Medications to Treat Mast Cell Disorders

- ▶ **Self-Injectable Epinephrine** (two doses; e.g., EpiPen[®] /EpiPen Jr[®]) should be carried by all patients with a mast cell disorder at all times, even if previous anaphylaxis has not occurred.

Medications to Treat Mast Cell Disorders

- ▶ **Basic Medications for Symptomatic Patients with Mast Cell Disorders¹⁻⁴**
- ▶ **H1 antihistamines:** help with itching, abdominal pain, flushing, headaches, brain fog
- ▶ **H2 antihistamines:** help with gastrointestinal symptoms and overall mast cell stability (all mast cell activation symptoms)
- ▶ *Note: The H1 and H2 antihistamines are necessary to stabilize receptors on the mast cell. Therefore, if additional medication is required for control of gastroesophageal reflux (GERD), a proton pump inhibitor may be added to this protocol, but it cannot replace the H2 antihistamine*


Medications to Treat Mast Cell Disorders

- ▶ **Mast cell stabilizers:** help with stomach and intestinal symptoms and brain fog
- ▶ **Leukotriene inhibitors:** help with respiratory symptoms and overall mast cell stability (all mast cell activation symptoms)
- ▶ **Aspirin therapy (*under direct supervision of a physician*):** if tolerated and if prostaglandins are elevated, helps with flushing, brain fog and bone pain

MCAS

- ▶ Recommended book: *Never Bet Against Occam* by Lawrence Afrin, M.D.
- ▶ Discussion:
 - What symptoms do you experience?
 - What do you use daily, as prevention?
 - What do you use for flares?
- ▶ Slides will be available for review after the meeting.

Skin Issues in EDS

- ▶ Elasticity, laxity
 - ▶ Easy bruising, fragility
 - ▶ Rashes (MCAS-related?)
 - ▶ Striae (stretch marks)
 - ▶ Slow healing
 - ▶ Atrophic scars
 - ▶ Hernias
- 

Thank You!



-and good night!