**MAST CELL ACTIVATION SYNDROME (MCAS)**

**WHAT ARE MAST CELLS?**

Mast cells (MC) are immune system cells that live in the bone marrow and in body tissues, internal and external, such as the gastrointestinal tract, the lining of the airway, and the skin. Everyone has mast cells in their body, and they play many complex and critical roles in keeping us healthy. The positive roles that they play include protecting us from infection, and helping our body by participating in the inflammatory process. However, mast cells are also involved in allergic reactions, from the tiny swelling that appears after a mosquito bite to life threatening, full-blown anaphylaxis.

Mast cells have within them small sacs surrounded by membranes. The sacs contain many different kinds of substances called mediators, which participate in all of the roles above, including allergic response and anaphylaxis. The mediators are selectively released when there is an allergic or mast cell based reaction.

There is a difference between someone who is healthy, with mast cells that are functioning normally, and someone with a mast cell disorder, whose mast cells may be activating inappropriately in response to triggers, or may also be proliferating and accumulating in organ tissues.

**WHAT ARE MAST CELL DISORDERS?**

Mast cell disorders are caused by the proliferation and accumulation of genetically altered mast cells and/or the inappropriate release of mast cell mediators, creating symptoms in multiple organ systems.The two major forms of mast cell disorders are **mastocytosis** and **mast cell activation syndromes (MCAS)**. While systemic mastocytosis (SM) is a rare disease, those suffering with MCAS have recently been increasingly recognized and diagnosed. It is important to note that the *process* of mast cell activation can occur in anyone, even without a mast cell disorder, as well as in patients with both mastocytosis and MCAS.

**WHAT is MAST CELL ACTIVATION SYNDROME (mcas)?**

This describes a subset of mast cell disorder patients who experience episodes of mast cell activation without detectable evidence of a proliferative mast cell disorder.

Symptoms may involve different organ systems:

* Skin: itching, flushing, hives, sweating
* Eyes: itching, watering
* Nose: itching, running, sneezing
* Mouth and throat: itching, swelling of the tongue, lips, and/or throat
* Lungs: trouble breathing, wheezing
* Heart and blood vessels: low blood pressure, rapid heart rate
* Stomach and intestines: cramping, diarrhea, nausea, abdominal pain
* Nervous system: headache, dizziness, confusion, extreme tiredness

The patient typically exhibits recurring symptoms involving 2 or more organ systems in parallel and are found not to be caused by any other condition or disorder other than mast cell activation.

**HOW IS MCAS EVALUATED?**

Labs may be drawn to provide evidence that mast cells are directly involved in the symptoms. Some of these include:

* Serum tryptase level
* 24-hour urinary tests to evaluate for an increase in mast cell mediator levels

Additionally, patients may be treated and evaluated for a response to medications that inhibit the action of histamine, inhibitor other mediators produced by mast cells, or block mast cell mediator release.

**HOW IS MCAS TREATED?**

Treating MCAS consists primarily of managing the symptoms.

For mild to moderate symptoms, antihistamines (H1 and H2 blockers), mast cell stabilizers, and/or anti-leukotrienes may be used to block the effects of mast cell mediators.

For severe symptoms, such as anaphylaxis, a prescription for an epinephrine auto-injector may be prudent, especially if the patient’s history indicates such concerns.