



## **Q+A: Mast Cell Disorders: More Than Just Hives**

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*All questions answered by Dr. Moshe Ben-Shoshan, except where indicated.*

### **Q: I have Cutaneous Mastocytosis, what are the chances of it becoming Systemic?**

A: In children, there is a higher than average probability they will outgrow the Cutaneous Mastocytosis, thus significantly reducing the chances of it going Systemic. If an adult has Cutaneous Mastocytosis, there is a good chance they already have Systemic Mastocytosis but it has gone undetected. Hence, in adults the investigation might include a bone marrow biopsy to rule out systemic forms.

### **Q: Can you have a Mast Cell Activation Disorder without Mastocytosis?**

A: Yes, but you will need to fulfill at least these three criteria:

1. Clinical symptomatology that is in keeping with the disorder.
2. A transient, measurable increase in either Serum Tryptase or other markers of mast cell mediators.
3. A response to agents that interfere with mast cell mediators (such as cetirizine).

### **Q: I have been diagnosed with MCAS and am managed well with Reactine, Montleukast and Ranitidine. How important is it to have further testing to rule out Mastocytosis?**

A: I would consider more testing (such as bone marrow biopsy) if you present with systemic symptoms consistent with anaphylaxis of an unclear trigger.

### **Q: Are there specific requirements or protocols that should be followed when processing a Tryptase sample (i.e. temperature must be kept cold)? Can any lab process Tryptase or is it typically sent away to be analyzed?**

A: Tryptase is very stable and does not require the same delicate care as other tests. Every province is different when it comes to where Tryptase can be tested.

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**Q: I was seen by a specialist who thinks I have Autoimmune Urticaria, however the test to confirm the absence of a particular protein in my blood is not available in Canada. Is this true? Is there another avenue I should explore to confirm Mast Cell Disease?**

A: Most cases of Chronic Urticaria are considered autoimmune (mainly to IL24). However, the tests are currently available only in research labs (mainly in Germany) and the test results will not practically change the treatment.

**Q: Is an Allergist the appropriate doctor to diagnose and treat Mast Cell Activation?**

A: Usually it requires an Allergist, Dermatologist and sometimes a Hematologist (when a bone marrow biopsy is needed).

**Q: My son has Mastocytoma and his Tryptase is normal, however he wasn't tested during or following a reaction. Should I ask for another test? Should I use corticosteroids cream on his spot?**

A: No, if it is Mastocytoma the diagnosis is clear (Tryptase is required mainly in cases that do not have clear diagnosis) and prognosis is excellent.

**Q: Is it common for people with Mast Cell Disorders to have angioedema in the gastrointestinal tract? Sometimes during a reaction, I have severe pain in my gall bladder area or on my left side under my ribs. Benadryl seems to ease the pain. Could angioedema be the cause of the pain?**

A: Theoretically, yes, although there are not many studies in the literature. It would be helpful if Tryptase were drawn during the pain episodes. If elevated, it would suggest mast cells played a role in the abdominal pain.

**Q: Which non-drowsy anti-histamines are least damaging to our organs?**

A: The main options would be Rupal or Blexten.

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**Q: How do I know when to use my epi-pen if I'm having Mast Cell reactions but am not in anaphylactic shock?**

A: Typically, if two or more systems are involved in the reaction, we recommend using your epi-pen. If symptoms are progressing and the patient is feeling concerned, we recommend using your epi-pen. It is important to visit the hospital after you've received epi to manage any rebound reactions.

**Q: Is it safe to use an epi-pen if significant hypertension (rather than hypotension) is one of the symptoms of a severe reaction that includes two or more organ systems?**

A: If it is anaphylaxis, it is better to use the epi-pen even if blood pressure is high.

**Q. Do second or rebound reactions happen more frequently in Mast Cell Disease anaphylactic episodes vs. allergy?**

A: There are no studies to support this question.

**Q: Is the Registry only for diagnosed cases of Mastocytosis or can diagnosed MCAS patients (meet the diagnostic criteria) also be included in the Registry?**

A: The Registry includes both Mastocytosis and MCAS.

**Q: How likely is it that children will outgrow Mastocytosis if symptoms persist into puberty? (increasing skin spots, GI symptoms, etc.)**

A: We need better studies, like the Registry, to provide a definitive answer to this. However, based on what we know today, if childhood symptoms continue into puberty it is less likely they will outgrow the Mastocytosis.



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**Q: Is elevated Chromogranin A in blood work an indicator for Mast Cell Disease?**

A: No, an elevated Chromogranin A is a biomarker for a neuroendocrine tumor like carcinoid.

**Q: Would you recommend patients follow a low histamine diet to control Mast Cell Disease symptoms?**

A: More research is required to determine if a low histamine diet is effective in the control of Mast Cell symptoms.

**Q: For those patients taking Nalcrom/Cromolyn 100mg capsules, what do you recommend these patients do during the current Canada wide drug shortage?**

A: Physicians and patients may look to other mast cell stabilizers such as Zaditen (Ketotifen), a higher dose of second generation anti-histamines, or perhaps Xolair (Omalizumab).