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# The Mastocytosis, MCAS & IA Patient Experience

We are sharing information about the rare disorders Mastocytosis (Masto), Mast Cell Activation Syndrome (MCAS) and Idopathic Anaphylaxis (IA), which we have derived from a number of sources in Canada and internationally. Our intention is to educate by sharing mastocytosis patient experiences reported over 10 years. These experiences are often beyond what has been researched and published in medical journals. Masto, MCAS, and IA can be severe and equally disabling. This is intended for general information purposes only and of course does not constitute medical advice. PLEASE SEE YOUR PHYSICIAN for medical advice and treatment.

It is unknown what causes or triggers Mastocytosis, MCAS or IA. Prolonged stress, emotional or physical trauma, illness/ infections, surgery of any kind, and even childbirth, have all been reported as contributing factors to the development or escalation of the disease. The basis of Mastocytosis & MCAS is a malfunctioning yet important immune system cell called a mast cell. Anything which triggers the immune system, including surgery, will trigger mast cell degranulation. The chemicals released through mast cell degranulation are the initiators of severe and prolonged symptoms of Mastocytosis, MCAS and contributes to IA.

Mast cells are created in the bone marrow and are found in every part and system of the human body, including the brain. Everyone has mast cells. In healthy people, mast cells function correctly. In Mastocytosis & MCAS patients, the mast cells may be malformed, increased in number, and/or behaving abnormally. Mastocytosis is classified as a neoplastic disorder (like cancer) and recently, also as a stem cell disorder. MCAS patients will have various markers of mastocytosis in their bone marrow but may not meet the entire WHO diagnostic criteria for Masto. It is theorized that MCAS may actually be the early stages of Systemic Mastocytosis but more research needs to be done in that area to know for sure. Due to the mast cell being part of the immune system, Masto and MCAS patients often experience autoimmune type responses but these responses are secondary to the mast cell disease.

Mastocytosis Society Canada strives to educate patients, caregivers, the general public and physicians fully about these rare disorders. This is crucial to target helpful treatments to lessen suffering and hopefully slow down progression of the illness. Current treatment focuses on managing the ongoing symptoms of the disease but do very little, if anything, to slow it's progression. There is presently no cure for Mastocytosis, MCAS or IA.

Most physicians do not recognize or understand Mastocytosis, MCAS, or IA as most doctors have not been trained to recognize these disorders. In fact, most mast cell disease patients go years before getting a proper diagnosis. Misdiagnosis is very common as a doctor cannot diagnose what he or she has never heard of or witnessed. Additionally, the existing diagnostic tests have proven to be

unreliable and inconsistent, evidenced by disparate results amongst the international Mastocytosis, MCAS and IA patient base.

Internationally, there appears to be a consensus amongst Mastocytosis patient groups, mast cell researchers, and physicians experienced in treating Mastocytosis, MCAS and IA, as to the following:-

1. Existing mastocytosis diagnostic tests need to be improved or completely new tests need to be developed. We estimate that approximately half the mastocytosis patient base worldwide does not meet the current diagnostic parameters, however they do experience benefits from being treated for mastocytosis. Many patients are being diagnosed clinically with MCAS and/or IA, based solely on their symptoms, in an effort to help them find some relief. This approach is working, resulting in saving patients lives, and physicians acknowledging that medicine just does not know nearly enough about mastocytosis yet.

2. Patient experiences of symptoms and secondary conditions (arthritis, osteoporosis, pancreatic tumours, chronic fatigue, ehlers danlos, connective tissue diseases, POTS, fibromyalgia, raynauds, iritis/uveitis, vertigo, interstitial cystitis, autonomic nervous dysfunction, thyroid malfunction or cancer, cerebral aneurysms), are often beyond what has been medically outlined as within the mastocytosis definition. However, many mastocytosis patients suffer at least one of these complications. Some have/had more than one.

3. Medical research has barely scratched the surface in fully defining, diagnosing and treating Mastocytosis, MCAS and IA. As a result, the stages or classifications of Mastocytosis, and the resulting effects or symptoms of each stage, are misleading or incomplete. MCAS and IA are often not addressed. Medicine does not know this disorder or mast cells nearly well enough to provide answers to all that patients endure and suffer. Physicians experienced in treating Mastocytosis, MCAS and IA patients are aware of this. Many Mastocytosis patients experience some or all of the symptoms in the same severity. (*Reference: The Mastocytosis Research Institute in France* http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2386235/). For example, Cutaneous (skin) forms and Indolent Systemic Mastocytosis presents the same symptoms as Aggressive Systemic Mastocytosis, both in type and severity. Thus, many patients with Skin involvement, Indolent Systemic Mastocytosis, MCAS and/or IA, suffer continuously and live extremely restricted, largely isolated, quiet lives in order to best control their immediate environments. These are not benign or minor diagnoses. All forms of Mastocytosis, including MCAS and IA, are in many cases life threatening and patients diagnosed with all forms typically experience continual escalation of current symptoms, and development of new symptoms over time.

4. Current treatments do not remove or prevent all the symptoms. Patients with Mastocytosis, MCAS and IA, suffer a wide range of life threatening and/or disabling and permanently damaging symptoms. This prevents many patients from living a normal life and basically existing in a state of catering to their disease to try to lessen their suffering. The most basic daily life activities induce suffering and internal damage over time for mastocytosis patients. For example, symptoms are triggered by simple activities such as daily bathing, inhaling airborne scents/chemical toxins, walking or any other exercise, eating, cognitive processing (concentration, learning and memory impairments), being in extreme temperatures (hot or cold in weather or water), talking for extended periods of time, engaging in social activities, stress or fatigue, and even for some, getting dressed every day. Friction and sensitivity to clothing, pressure, and coming in contact with man made materials, are enough to trigger symptoms for some patients. Foods and drinks often play an enormous role in triggering the symptoms, with the main troubles caused by foods & drinks which are naturally high in Histamine, Tyramine, Gluten and/or Salicylates.

Mastocytosis, MCAS and IA, negatively impact working and personal lives. Many of these patients have a restricted diet of very few foods they can safely eat, constant fatigue, episodes of vertigo and anaphylaxis, as well as inability to exercise on a regular basis (includes walking) due to fatigue and weakness and all the other symptoms. On a daily basis, many patients experience difficulty with tasks requiring concentration, learning, retention or anything relying on memory or information processing skills. Many people suffering with these rare disorders cannot commit to anything in terms of a schedule or regular routine because their symptoms vary in type and severity from day to day and often hour to hour. They are continuously symptomatic with disabling effects such as fatigue, sudden unexplained episodes of drops in blood pressure (very common), anaphylaxis and anaphylactoid reactions, flushing, shaking & tremors, and poor concentration.

Most Mastocytosis, MCAS and IA patients find that medications do not eradicate or prevent their symptoms. This disorder requires avoidance or minimization of exposure to triggers in addition to taking medications. Triggers can be anything which causes mast cells to degranulate. This includes hormonal activity, emotions (positive or negative), physical activities, positive/negative stress, foods and drinks, temperature, and exercise/exertion of any kind. It is only by controlling their exposure to triggers in addition to taking medications, that they can achieve some relief from the symptoms, but it is not total relief and this does not return them to functioning normally on a consistent and reliable basis. Their good days are very few and far between, rarely even lasting a full day. Most of these patients share this same experience.

Typically, Mastocytosis, MCAS and IA patients experience increasing limitations and greater suffering over their lifetime. If they push against their symptoms, believing that "trying harder" or pushing to complete tasks (laundry, grocery shopping, visiting a friend, cleaning), the symptoms become more severe and prolonged and continue to ricochet setting off other symptoms for days, weeks or months on end. There is no predictability to the array of symptoms. There is no way to make the symptoms behave consistently or to be able to garner better control over them. The medical research community has not yet fully figured out mast cell disorders. In the meantime, patients are driving the medical research via their physicians reporting the escalating symptoms and secondary and tertiary complications suffered by their patients. Everyone wants to have the answers, patients most of all.

Mastocytosis, MCAS and IA patients often look normal or healthy to others, especially when they are enjoying one of their better days/moments when their symptoms are subdued or well controlled, as during these times they may be able to be more active. However, these stable periods do not endure and it is the fluctuating nature of the illness that causes much difficulty for the person with mastocytosis. Many of these patients are not seen regularly by people outside their immediate family or caregiver except for those rare occasions when they are feeling well. As a result, people fail to understand Mastocytosis, Mast Cell Activation Syndrome and Idiopathic Anaphylaxis, and the extent to which they prevent individuals from living normal and active lives.

Mastocytosis, Mast Cell Activation Syndrome, and Idiopathic Anaphylaxis, are irreversible and life threatening rare disorders and in most cases, the symptoms are extraordinarily difficult to manage.

By Mastocytosis Society Canada