

## Information on referrals for suspected mast cell activation syndrome (MCAS)

**Relates to all referrals that suggest EDS, POTS requires exclusion of MCAS**

Please note that we do not offer a service for non-clonal idiopathic mast cell activation syndrome.

The AAAAI Mast Cell Disorders Committee Work Group Report states that the following disorders have **no scientific basis** for being associated with mast cell activation: Ehlers-Danlos syndrome (EDS), **postural orthostatic tachycardia syndrome (POTS)**, typically with hypotension, irritable bowel syndrome, gastroesophageal reflux disease, idiopathic non-ischemic cardiomyopathy, metabolic syndrome, ADHD, depression, multiple chemical sensitivity syndrome, autoimmune disorders, endometriosis, PCOD, celiac disease and nonceliac gluten intolerance, migraine headaches, neurogenic pain syndrome or restless leg syndrome. (Ref: *J Allergy Clin Immunol* 2019;144(4):883-896).

As per existing consensus criteria, the term MCAS applies when

- i) typical clinical signs of severe recurrent acute systemic mast cell activation features are present (must fulfil criteria for 'idiopathic anaphylaxis' with involvement of at least 2 organ systems),
- ii) the involvement of mast cells should be demonstrated by biochemical analyses (preferably through an increase in tryptase to >20% of baseline plus 2 ng/ml, e.g., if baseline tryptase is 5 ng/ml, an increase >8 ng/ml suggests the possibility of mast cell activation) and
- iii) the symptoms respond to treatment with MC stabilizing agents or drugs targeted against MC mediator production, secretion or receptor binding.

All three criteria **must be met** to establish the diagnosis of MCAS.

(Ref: *J Allergy Clin Immunol Pract.* 2019 April; 7(4): 1125–1133.e1)

**POTS not considered** associated with mast cell activation but more of an autonomic dysfunction of blood vessels. Patients with hyperadrenergic form of postural orthostatic tachycardia syndrome (POTS) and hypermobility-type Ehlers-Danlos syndrome (EDS) have been discussed to suffer from symptoms resembling or mimicking mast cell activation or even mast cell activation syndrome, but **objective evidence to incriminate mast cell in their pathology has been lacking**.

Urticaria or dermatographism are not a form of mast cell activation syndrome. Please be aware that some patients might be very quickly **misdiagnosed** with MCAS when they might actually be experiencing other far more common conditions, such as chronic spontaneous urticaria or asthma. Misdiagnosis can be deleterious for patients.

We strongly recommend that patients **NOT** labelled with this condition, but referred to Clinics that may specialise in this area (example, multidisciplinary mastocytosis clinics).

Our clinic does not specialise in this area, and therefore unable to accept these referrals.