MAST CELL ACTION

A Mast Cell Activation Syndrome (MCAS) diagnosis is supported by a set of criteria that were agreed by an international group of doctors in 2011.[1] MCAS was coded into ICD-10-CM in October 2016.[2]

Whilst the current understanding of MCAS is limited, it is a starting point, recognising that a growing patient population is presenting with symptoms that look like they are mast cell-driven, but are not due to mastocytosis or any other defined allergic or inflammatory condition. More work is needed to identify cohorts of patients, foster research and to develop treatments for people with MCAS.

Mast Cell Action is being contacted by an increasing number of parents who have spent years trying to get treatment for their children; often having to see private doctors. Almost all have a core set of similar allergy-like symptoms and a long list of diagnoses that do not explain their symptoms.

Treatment for MCAS can be cheap and effective, and can greatly improve the patient's quality of life. However, in the absence of a diagnosis, many are not receiving this simple and effective treatment for their disabling symptoms, and are undergoing costly investigations and consultations.

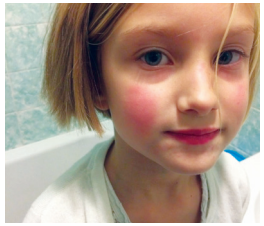
1. Valent P et al. Int Arch Allergy Immunol. 2012;157(3):215–225
2. International Classification of Diseases, Tenth Revision, Clinical Modification (ICD-10-CM). Available online at: www.cdc.gov/nchs/icd/icd10cm.htm

For further information contact: info@mastcellaction.org
Visit us at: mastcellaction.org

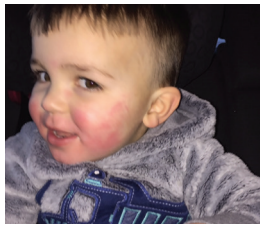
Visit our website resources for a bibliography of the latest papers on Mast Cell Activation Disorders.

Registered Charity Number 1164917

A unifying feature experienced by many children in our patient support group, including Lily, Jensen and Caspyn, is the perplexing range of reactions to foods and environmental triggers from birth. Most of these children struggle to find access to coordinated care and achieve a clear pathway towards a diagnosis. Their symptoms are too frequently been met with suspicion or doubt, causing significant impact on their mental well-being and that of their family.



Lily
For Lily daily life is an enormous struggle, she has to stick to a very restricted diet and reacts to a wide range of environmental triggers, including scents and vibration. She does not tolerate many medications, which makes her treatment very difficult.



Jensen
Jensen reacts to a great many foods and external triggers. He also suffers from syncope (loss of consciousness) and some days he cannot walk due to his genitourinary pain.

Caspyn
Caspyn can only tolerate ~20 foods and reacts to external sensory triggers. He has never slept more than a few hours at a time, often due to gastrointestinal discomfort. He tolerates some foods after gradual introduction, but many continue to worsen symptoms including spasms, hyperactivity and anxiety.

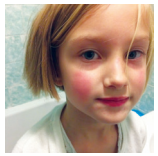
Although it took several years, Lily has now been diagnosed with MCAS and Jensen has been diagnosed with Hereditary alpha tryptasemia (HATs). Lily is under the care of a paediatric clinic with specialist knowledge of MCAS and her quality of life has improved enormously. In this leaflet we compare a historical picture of the three children's clinical symptoms.

Mast Cell Action is a new patient support and advocacy charity for MCAS Sufferers.

MAST CELL ACTION

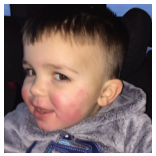
We are asking doctors to consider whether the role of the mast cell and mast cell mediators may be a cause of the multiple symptoms experienced by children like Lily, Jensen and Caspyn

Lily's parents have been trying to find compounded, additive-free medication, which are widely available in other developed countries



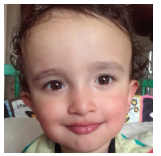
LILY'S SYMPTOMS

Jensen's parents are currently trying to find a urologist to help with his painful genitourinary symptoms



JENSEN'S SYMPTOMS

Caspyn's parents are hoping for a referral to someone with an understanding of mastocytosis and mast cell activation disorders



CASPYN'S SYMPTOMS

	LILY'S SYMPTOMS	JENSEN'S SYMPTOMS	CASPYN'S SYMPTOMS
DIGESTIVE:	Diarrhoea, vomiting, abdominal pain, nausea	Diarrhoea, reflux	Silent reflux, stomach pains, constipation
GENITOURINARY:	Vulvodynia, cystitis, urgency to urinate	Urinary incontinence, strong smelling urine, urinary frequency, pain - unable to walk	Urinary urgency when reacting, suspected urinary tract infections
CUTANEOUS:	Hives, itching, flushing, contact urticaria	Hives, flushing, sores, facial, tongue and lips swelling	Mastocytoma mast cell tumour, (which flares when reacting), rashes, hives and cradle cap (triggered by foods), flushing
CONSTITUTIONAL:	Fevers, fatigue, feeling unwell	Night sweats, fevers, feeling cold	Excessive sweating, pronounced hyperactivity when trialling new foods
NEUROLOGICAL:	Mood changes, anxiety, headaches, brain fog	Insomnia, anxiety	Body spasms, jaw tensing and head shaking, hyperactivity, anxiety linked to certain foods
RESPIRATORY:	Throat swelling	Wheezing, rattling chest, rhinitis, croup-type cough	Constant sneezing (in response to foods)
CARDIOVASCULAR:	Rapid change in heart rate at rest, dizziness	Turning blue with sudden sleepiness, syncope	Sudden episodes of drowsiness/sleepiness
MUSCULOSKELETAL:	Back, leg, shoulder pain	Leg pain	Foot and joint pain, some hypermobility, (family history of Ehlers-Danlos syndrome)
DIET:	Limited to 11 foods that she can tolerate	Reacts to a wide range of foods, that constantly change	Limited to 20 foods
MEDICATION:	Ketotifen, sodium cromoglicate, montelukast, desloratadine, hydrocortisone cream, adrenaline autoinjector, multiple drug intolerances*	Ketotifen, sodium cromoglicate, cetirizine, fexofenadine, ferrous gluconate, chlorphenamine maleate, adrenaline autoinjector	Ranitidine, cetirizine for flare ups, trialling sodium cromoglicate
ALLERGY TESTS:	Normal	Normal	Normal
MEDIATOR TESTS:	Normal tryptase, elevated histamine and prostaglandin levels	Elevated tryptase; confirmed as having gene triplication of the alpha tryptasemia gene	Normal tryptase
DIAGNOSES:	MCAS, previous diagnoses included chronic idiopathic urticaria with angioedema, fructose malabsorption, lactose intolerance, salicylate sensitivity, paraben mix allergies	HAT (Hereditary alpha tryptasemia), previous diagnosis were iron deficiency anemia, fructose malabsorption, histamine intolerance, chronic idiopathic urticaria and angioedema	Solitary mastocytoma, silent reflux

**Lily has recently trialled low-dose naltrexone, resulting in a dramatic improvement in her symptoms.*