# 1 Recognising & Managing MCAS Overview

Key Information | Think About | Action | Caution

Mast Cell Activation Syndrome (MCAS) is part of a spectrum of mast cell disorders characterised by abnormal mast cell activation and mediator release, without evidence of clonal mast cell proliferation.

MCAS affects both children and adults and may present at any age. Symptoms are often long-standing but under-recognised, there may be mild or intermittent symptoms from childhood which escalate in adulthood, or symptoms may be severe from birth. Sometimes a clear precipitating event precedes escalation of symptoms, including infection, surgery, stress, hormonal changes.
**The condition is frequently characterised by unpredictable, relapsing–remitting symptoms involving multiple organ systems.**

**Patient presents with a multisystem constellation of allergy-like and inflammatory symptoms that remains unexplained by conventional investigation and are refractory to standard treatment.**

**Obtain a detailed history of symptoms and triggers (Checklist on P18 may help)**

Use clinical judgement to rule out differential diagnoses.

**Consider - Does the patient have any of the following?**

- Multi-speciality referral history
- Other historical symptoms that are potentially mast-cell mediated?
- A relapsing–remitting pattern of illness, often without an obvious unifying diagnosis
- Family history of similar symptoms
- Food, medication, or environmental intolerances

**Common co-morbid conditions?**
- Hypermobility spectrum disorders, EDS. Hypermobility
- PoTS and autonomic nervous system dysfunction
- Long COVID, (ME/CFS)
- Autism, ADHD

**Symptoms affecting multiple body systems, such as:**
- Skin (flushing, rashes, hives, itching, dermographism)
- Gastrointestinal (abdominal pain, diarrhoea, constipation, nausea, bloating)
- Respiratory (wheeze, throat tightness, breathlessness)
- Cardiovascular (palpitations, dizziness, fainting, chest pain)
- Neurological (headaches, brain fog, fatigue, mood changes)
- General (fatigue, muscle or joint pain, chemical/medication/food sensitivities

**Referrals**
Many patients with MCAS may be adequately managed in Primary Care. However, referral may be required for
- Ongoing symptoms that are not adequately controlled by first-line treatment
- Patients with recurrent anaphylaxis
- Those needing psychological support to cope with severe symptoms
- History of anaphylaxis
- Need for advanced diagnostic testing
- Suspected clonal mast cell disorders

**Affirmative answers to the above are often indicative of MCAS, but absence does not rule out MCAS.**

**Does Patient meet diagnostic criteria?**
- Inflammatory symptoms, 2 or more body systems
- Response to treatment - improvement with mast cell–targeted therapies
- Mediator testing where available - Elevated mast cell mediators N-Methyl Histamine /Prostaglandins (blood or urine)
- Exclusion of alternative diagnoses - Differential diagnosis is essential, as symptom overlap is common.

**Prescribe medication for mast cell mediated symptoms**

Encourage diary to log symptoms and triggers

Educate on Patient Management Strategies

Book a follow up appointment

**Medical Management**
- H1 and H2 Blockers
- Antileukotrienes
- Mast Cell Stabilisers
- Corticosteriods
- Epi-Pens where necessary

Patient, systemic trial may be necessary.

**Patient Management**
- Symptom Diary
- Trigger identification and avoidance
- Dietary modification to lessen/ avoid high histamine or other trigger foods
- Stress management

**Stabilisation**
Review medication response
Adjust accordingly
Trial different categories of medication, different medications within the same category, different brands and different combinations until symptoms are well controlled

If symptom control cannot be achieved, consider referral.

**Ongoing Management**
Review medication regularly for continued effectiveness, and adjust as appropriate

Viral infections, surgeries, stressful life events, unexpected trigger exposure etc may cause temporary or permanent illness progression which may necessitate adjustment of medication.

**Consider Possible Triggers**
- Foods (particularly high-histamine foods)
- Medications
- Fragrances, perfumes, and chemicals
- Household detergents and laundry products
- Pollen, dust, pet dander, and moulds
- Temperature changes, heat, sunlight
- Physical exertion or exercise
- Stress
- Hormonal fluctuations (e.g. puberty, menopause)
- Viral/bacterial/fungal/infections

**Anaphylaxis in MCAS**
Anaphylaxis is a mast cell mediated reaction.
People with MCAS are potentially at increased risk of anaphylaxis, affecting around 7% of MCAS patients.
May be atypical presentation
Emergency management follows standard UK guidance
- Establish whether there is a history of anaphylaxis
- Explain the signs, triggers and symptoms of anaphylaxis to the patient
- Prescribe 2 Epi-Pens
- Provide Personalised Management Plan

**Useful Resources**
Mast Cell Action's 'Brief Guide to MCAS'
Mast Cell Action's 'Self-management Toolkit'
CAMRare passports or medical bracelet can be helpful
Mast cell action have services and resources to help individuals navigate life with MCAS

**Caution**
- Medication sensitivities are common.
- Introduce drugs cautiously, titrate slowly.
- Avoid problematic medications eg Opioids, NSAIDs, contrast agents, alcohol, certain local and general anaesthetics, excipients like colourings, flavourings, fillers, binders etc.
- Patients may only tolerate specific brands.
- Medications may need to be compounded from specialist pharmacies.
- Surgeries and investigative procedures can trigger flares, consider premedication protocol.

View this Flowchart Online