Diagnostic guide — Child

For the attention of the physician · Mast cell activation in children · PMCHS / SHMP

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Clinical context: The parents of this child suspect programmed mast cell hyperreactivity (PMCHS / SHMP). This document outlines the recommended diagnostic and therapeutic approach when biological proof is difficult to obtain.

1. Episodes to recognise — attenuated multi-systemic presentation

A formal MCAS episode requires symptoms in ≥ 2 organ systems and a transient mediator rise. In children, these episodes are often attenuated and difficult to capture.

Skin

Peri-oral redness, dermographism, localised itching within 5–30 min of a trigger.

Digestive

Abdominal pain (knees to belly), alternating diarrhoea/constipation, reflux, food refusal.

Neuro-behavioural

"Tipsy", vacant look, paroxysmal irritability after a trigger food.

Autonomic nervous system

Sudden pallor, cold sweats, abrupt drowsiness after a meal.

2. Background terrain (between flares)

Skin

Easy dermographism, transient redness, intermittent itching without frank eczema.

Digestive

Recurrent abdominal pain, bloating, reflux, alternating bowel — impact on sleep/mood.

Neuro-behaviour

Fluctuating irritability, sensory intolerance (sounds/light/smells), brief hypo-reactive episodes.

■■ **Pitfalls:** Normal baseline tryptase ≠ absence of MCAS (mediators are fluctuating). Negative IgE tests do not rule out non-IgE histamine release.

3. Pragmatic diagnostic approach

Without biological "proof" — maximise safety and clinical relevance.

a) Structured clinical documentation (1–2 months)

· Observation form: link trigger → ≥ 2 systems → resolution.
· Dated photographs of skin lesions.

b) Ruling out common differential diagnoses

· IgE allergy (patch tests, prick tests if suspected)
· Pathological reflux, parasitosis
· Coeliac disease / enteropathy, IBD if red flags present

c) Opportunistic, non-traumatic blood / urine tests

· Baseline serum tryptase (outside a flare)
· 24h urine after a notable flare: N-methylhistamine, LTE4, PGD2 metabolite
· Negativity does not exclude the mast cell hypothesis in children
· Do not provoke a flare to capture a mediator (unethical)

4. Step-by-step therapeutic trial

Treat mast cell symptoms and observe functional impact.

1 Non-pharmacological (2–4 weeks)

· Avoid identified major triggers (tomato, citrus, chocolate, heat, smells).

· Light low-histamine diet: limit fermented foods, aged cheeses, old leftovers.

· Sleep hygiene and soothing sensory routines.

2 Non-sedating H1 antihistamine (if approved)

· Age-appropriate paediatric dosage, continuously or before predictable triggers.

· ± H2 if significant digestive component.

3 Oral sodium cromoglycate (if GI predominant)

· Mast cell stabiliser, mainly digestive benefit.

· Known paediatric safety profile in mastocytosis / MCAD.

4 Leukotriene antagonists / specialist referral

· If strong suspicion, previous steps failed, functional impact.

· Allergology / paediatric gastroenterology opinion.

Response assessment (8–12 weeks)

· Fewer documented multi-systemic episodes
· Reduced intensity / duration
· Improved sleep / comfort / school participation

Clinical response to anti-mediator treatment supports the mast cell hypothesis when biological workup is non-contributory. This is a recognised supporting criterion.