

Mast Cell Activation Syndrome (MCAS) is a **formally recognised Mast Cell Disorder**.

The American Centre for Disease Control (CDC) included a **diagnostic classification** of MCAS within their 2017 revised edition of the International Classification of Diseases (ICD-10).

The **ICD-10-CM codes** for Mast Cell Activation Syndrome and Mastocytosis can be found on The Mast Cell Disease Society website which is available at: <https://tmsforacure.org/icd-10-cm/>

This document provides the details of some key literature in the MCAS disease area. The document is divided into core topics, and outlines the subject of each publication. Hyperlinks to **full free texts** are provided where available, otherwise there are hyperlinks to **abstracts or posters**.

Medical consensus on MCAS:

1. **Recent international publication on diagnosing MCAS:** Afrin LB, Ackerley MB, Bluestein LS, et al. Diagnosis of mast cell activation syndrome: a global "consensus-2" [published online ahead of print, 2020 Apr 22]. *Diagnosis (Berl)*. Available at: <https://www.degruyter.com/view/journals/dx/ahead-of-print/article-10.1515-dx-2020-0005/article-10.1515-dx-2020-0005.xml?language=en>
2. **AAAAI Working Group Report:** Weiler CR, Austen KF, Akin C, et al. AAAAI Mast Cell Disorders Committee Work Group Report: Mast cell activation syndrome (MCAS) diagnosis and management. *J Allergy Clin Immunol*. 2019;144(4):883-896. Available at: [https://www.jacionline.org/article/S0091-6749\(19\)31116-9/pdf](https://www.jacionline.org/article/S0091-6749(19)31116-9/pdf)
3. **First proposed consensus of MCAS classification:** Valent P, Akin C, Arock M, et al. Definitions, criteria and global classification of mast cell disorders with special reference to mast cell activation syndromes: a consensus proposal. *Int Arch Allergy Immunol*. 2012;157(3):215-225. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3224511/>

Clinical manifestations of MCAS:

4. **Literature analysis of mast cell disorders:** Theoharides TC, Tsilioni I, Ren H. Recent advances in our understanding of mast cell activation - or should it be mast cell mediator disorders?. *Expert Rev Clin Immunol*. 2019;15(6):639-656. doi:10.1080/1744666X.2019.1596800. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7003574/>
5. **Patient Perceptions of MCAS:** Jennings S, Russell N, Jennings B, et al. The Mastocytosis Society survey on mast cell disorders: patient experiences and perceptions. *J Allergy Clin Immunol Pract*. 2014;2(1):70-76. Available at: [https://www.jacionline.org/article/S0091-6749\(18\)32704-0/fulltext](https://www.jacionline.org/article/S0091-6749(18)32704-0/fulltext). Poster available at: <https://tmsforacure.org/mcas-survey-poster/?fbclid=IwAR113YRom5M28B8eRuMXthDI5ZG1AS4Xv9wV4kr7bXcfbRiJ8hD-bFIP3TI>
6. **Anaphylaxis and MCAS:** Jimenez-Rodriguez TW, Garcia-Neuer M, Alenazy LA, Castells M. Anaphylaxis in the 21st century: phenotypes, endotypes, and biomarkers. *J Asthma Allergy*. 2018;11:121-142. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6016596/>

7. **Characterization of MCAS:** Afrin LB, Self S, Menk J, Lazarchick J. Characterization of Mast Cell Activation Syndrome. Am J Med Sci. 2017;353(3):207-215. doi:10.1016/j.amjms.2016.12.013. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5341697/>
8. **Dermatology and MCAS:** Cookson H, Grattan C. An update on mast cell disorders. Clin Med (Lond). 2016;16(6):580-583. doi:10.7861/clinmedicine.16-6-580. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6297339/?fbclid=IwAR3kL1IM9jcz8HzdUpo2hcJpgpEfK-M-RE9LAZKAHWWhaSQYCov3gB-J3JX5k>
9. **Mast cells, mastocytosis and related disorders:** Theoharides TC, Valent P, Akin C. Mast Cells, Mastocytosis, and Related Disorders. N Engl J Med. 2015;373(2):163-172. Available at: <http://www.mastcellmaster.com/documents/2018-07/Mast-cells-mastocytosis-and-supplemental-NEJM-July-9-2015.pdf>
10. **Clinical manifestations of MCAS:** Hamilton MJ, Hornick JL, Akin C, Castells MC, Greenberger NJ. Mast cell activation syndrome: a newly recognized disorder with systemic clinical manifestations. J Allergy Clin Immunol. 2011;128(1):147-152.e2. Available at: http://williams.medicine.wisc.edu/mast_cell_activation_2011.pdf

Diagnosing MCAS:

11. **Laboratory tools for measuring mast cell mediators:** Butterfield J, Weiler CR. The Utility of Measuring Urinary Metabolites of Mast Cell Mediators in Systemic Mastocytosis and Mast Cell Activation Syndrome. J Allergy Clin Immunol Pract. 2020;8(8):2533-2541. Abstract available at: <https://doi.org/10.1016/j.jaip.2020.02.021>
12. **Laboratory tools for diagnosing MCAS:** Leru, PM, et al. Mast cell activation syndromes-evaluation of current diagnostic criteria and laboratory tools in clinical practice. Experimental and Therapeutic Medicine.2020;3:2348-51. Available at: <https://www.spandidos-publications.com/10.3892/etm.2020.8947>
13. **Differential diagnosis of MCAS:** Weiler CR. Mast Cell Activation Syndrome: Tools for Diagnosis and Differential Diagnosis. J Allergy Clin Immunol Pract. 2020;8(2):498-506. Abstract available at: <https://doi.org/10.1016/j.jaip.2019.08.022>
14. **Proposed diagnostic algorithm for MCAS:** Valent P, Akin C, Bonadonna P, et al. Proposed Diagnostic Algorithm for Patients with Suspected Mast Cell Activation Syndrome. J Allergy Clin Immunol Pract. 2019;7(4):1125-1133.e1. Available at: https://www.sciencedirect.com/science/article/pii/S221321981930056X?via%3Dihub&fbclid=IwAR1jIjNFhJeuns0bG-WuzYCUqJjD_16ZgM_OnJ3QC88mXh3vY-TEeCopTsh8
15. **MCAS testing challenges:** Zenker N, Afrin LB. Utilities of Various Mast Cell Mediators in Diagnosing Mast Cell Activation Syndrome. Blood. 2015;126(23): 5174. Abstract available at: <https://ashpublications.org/blood/article/126/23/5174/94641/Utilities-of-Variou-Mast-Cell-Mediators-in>
16. **MCAS diagnostic assessment guide:** Afrin LB, Molderings GJ. A concise, practical guide to diagnostic assessment for mast cell activation disease. World Journal of Hematology. 2014;3(1):1-17. Available at: <https://www.wjnet.com/2218-6204/full/v3/i1/1.htm>
17. **Diagnostic and treatment options guide for MCAS:** Molderings GJ, Brettner S, Homann J, Afrin LB. Mast cell activation disease: a concise practical guide for diagnostic workup and therapeutic options. J Hematol Oncol. 2011;4:10. Published 2011 Mar 22. Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3069946/>

Managing MCAS:

18. **Latest management guidelines:** Castells M, Butterfield J. Mast Cell Activation Syndrome and Mastocytosis: Initial Treatment Options and Long-Term Management. *J Allergy Clin Immunol Pract.* 2019;7(4):1097-1106. **Abstract available at:** <https://doi.org/10.1016/j.jaip.2019.02.002>
19. **MCAS diagnosis and therapeutic options:** Afrin LB, Butterfield JH, Raithel M, Molderings GJ. Often seen, rarely recognized: mast cell activation disease--a guide to diagnosis and therapeutic options. *Ann Med.* 2016;48(3):190-201. **Abstract available at:** <https://www.tandfonline.com/doi/abs/10.3109/07853890.2016.1161231?journalCode=iann20>
20. **Different treatment options for MCAS:** Molderings GJ, Haenisch B, Brettner S, et al. Pharmacological treatment options for mast cell activation disease. *Naunyn Schmiedebergs Arch Pharmacol.* 2016;389(7):671-694. **Available at:** <https://link.springer.com/article/10.1007%2Fs00210-016-1247-1>

MCAS Genetics and HAT:

21. **The genetics of mast cell activation:** Molderings GJ. The genetic basis of mast cell activation disease - looking through a glass darkly. *Crit Rev Oncol Hematol.* 2015;93(2):75-89. **Abstract available at:** <https://www.sciencedirect.com/science/article/abs/pii/S1040842814001498>
22. **Familial genetics for mast cell disorders:** Molderings GJ, Haenisch B, Bogdanow M, Fimmers R, Nöthen MM. Familial occurrence of systemic mast cell activation disease. *PLoS One.* 2013;8(9):e76241. Published 2013 Sep 30. **Available at:** <https://journals.plos.org/plosone/article?id=10.1371/journal.pone.0076241>
23. **Hereditary alpha-tryptasemia:** Lyons JJ. Hereditary Alpha Tryptasemia: Genotyping and Associated Clinical Features. *Immunol Allergy Clin North Am.* 2018;38(3):483-495. **Available at:** <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6411063/>
24. **Elevated tryptase and alpha-tryptasemia:** Lyons, J. J. et al. Elevated basal serum tryptase identifies a multisystem disorder associated with increased TPSAB1 copy number. *Nature Genetics*, 2016, vol. 48, no. 12, pp. 1564-1569 [Online]. **Available at:** https://www.research.manchester.ac.uk/portal/files/50850071/alpha_ tryptasemia_manuscript_Nat_Genet.pdf