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.

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/>

**Consensus papers on MCAS:**

1. **Recent international publication on diagnosing MCAS:** Afrin L, Ackerley M, Bluestein L, Brewer J, Brook J, Buchanan A et al. Diagnosis of mast cell activation syndrome: a global “consensus-2”. Diagnosis [Internet]. 2020 [cited 25 March 2022];8(2):137-152.
Available at: <https://www.degruyter.com/document/doi/10.1515/dx-2020-0005/html>
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%2819%2931116-9/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:**

1. **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/>
2. **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/>
3. **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/](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5341697/%20)
4. **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=IwAR3kL1lM9jcz8HzdUpo2hcJpg pEfkM-RE9LAZKAHWhaSQYCov3gB-J3JX5k](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6297339/?fbclid=IwAR3kL1lM9jcz8HzdUpo2hcJpg%20pEfkM-RE9LAZKAHWhaSQYCov3gB-J3JX5k%20)
5. **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>
6. **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](http://williams.medicine.wisc.edu/mast_cell_activation_2011.pdf%20)

**Patient Perceptions of Mast Cell Disorders:**

1. **Gut Symptoms in MCAS:** Slater R, Bowes H. P345 Dear doctor, could your patient’s unexplained gastrointestinal symptoms be caused by mast cell activation syndrome? Gut [Internet]. 2021 [cited 25 March 2022].
Available at: <https://gut.bmj.com/content/gutjnl/70/Suppl_1/A220.1.full.pdf>
2. **MCAS and Patient Perception**: Jennings S, Slee V, Hempstead J, Slee A, Castells M, Akin C et al. The Mastocytosis Society Mast Cell Activation Syndrome Patient Perceptions Survey. Journal of Allergy and Clinical Immunology [Internet]. 2019 [cited 25 March 2022];143(2): AB427. Available at: [https://www.jacionline.org/article/S0091-6749(18)32704-0/fulltext#relatedArticles.](https://www.jacionline.org/article/S0091-6749%2818%2932704-0/fulltext#relatedArticles.) Poster available at: [https://tmsforacure.org/mcas-survey-poster/?fbclid=IwAR113YRom5M28B8eRuMXthDl5ZG1AS4Xv9wV4kr7bXcfbRiJ8hD-bFIP3TI](https://tmsforacure.org/mcas-survey-poster/?fbclid=IwAR113YRom5M28B8eRuMXthDl5ZG1AS4Xv9wV4kr7bXcfbRiJ8hD-bFIP3TI%20)
3. **MCAS and Patient Perception:** Jennings S, Russell N, Jennings B, Slee V, Sterling L, Castells M et al. The Mastocytosis Society Survey On Mast Cell Disorders: Patient Experiences and Perceptions. Journal of Allergy and Clinical Immunology [Internet]. 2013 [cited 25 March 2022];131(2):AB116. Available at: [https://www.jacionline.org/article/S0091-6749(12)03088-6/pdf](https://www.jacionline.org/article/S0091-6749%2812%2903088-6/pdf)

**Diagnosing MCAS:**

1. **Critical Review of MCAS:** Gülen T, Akin C, Bonadonna P, Siebenhaar F, Broesby-Olsen S, Brockow K et al. Selecting the Right Criteria and Proper Classification to Diagnose Mast Cell Activation Syndromes: A Critical Review. The Journal of Allergy and Clinical Immunology: In Practice [Internet]. 2021;9(11):3918-3928.
Available at: [https://www.jaci-inpractice.org/article/S2213-2198(21)00676-0/fulltext](https://www.jaci-inpractice.org/article/S2213-2198%2821%2900676-0/fulltext)
2. **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
3. **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>
4. **Diagnosis tools:** 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>
5. **Proposed diagnostic algorithm:** 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=IwAR 1jlNFhJeuns0bG-WuzYCUqJjD\_16ZgM\_OnJ3QC88mXh3vY-TEeCopTsh8](https://www.sciencedirect.com/science/article/pii/S221321981930056X?via%3Dihub&fbclid=IwAR%201jlNFhJeuns0bG-WuzYCUqJjD_16ZgM_OnJ3QC88mXh3vY-TEeCopTsh8)
6. **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-Various-Mast-Cell-Mediators-in](https://ashpublications.org/blood/article/126/23/5174/94641/Utilities-of-Various-Mast-Cell-Mediators-in%20)
7. **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.wjgnet.com/2218-6204/full/v3/i1/1.htm>
8. **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/](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3069946/%20)

**Managing MCAS:**

1. **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](https://doi.org/10.1016/j.jaip.2019.02.002%20%20)
2. **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>
3. **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](https://link.springer.com/article/10.1007/s00210-016-1247-1)

**MCAS Genetics and HAT:**

1. **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/>
2. **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](https://www.research.manchester.ac.uk/portal/files/50850071/alpha_tryptasemia_manuscript_Nat%20_Genet.pdf%20)
3. **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\](https://www.sciencedirect.com/science/article/abs/pii/S1040842814001498/%20)
4. **Familiar 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>

**MCAS and Long Covid:**

1. **MCAS symptoms and Long-Covid:** Weinstock L, Brook J, Walters A, Goris A, Afrin L, Molderings G. Mast cell activation symptoms are prevalent in Long-COVID. International Journal of Infectious Diseases [Internet]. 2021; 112:217-226.
Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8459548/>
2. **MCAS symptoms and Long-Covid:** Wechsler J, Butuci M, Wong A, Kamboj A, Youngblood B. Mast cell activation is associated with post‐acute COVID‐19 syndrome. Allergy [Internet]. 2021; Available at: <https://onlinelibrary.wiley.com/doi/10.1111/all.15188>
3. **MCAS symptoms and Hyperinflammation and Long-Covid:** Afrin L, Weinstock L, Molderings G. Covid-19 hyperinflammation and post-Covid-19 illness may be rooted in mast cell activation syndrome. International Journal of Infectious Diseases [Internet]. 2020; 100:327-332.
Available at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7529115/>
4. **MCAS, Multisystem Inflammatory Syndrome and Long-Covid:** Theoharides TC, Conti P. COVID-19 and Multisystem Inflammatory Syndrome, or is it Mast Cell Activation Syndrome? J Biol Regul Homeost Agents. 2020 09-10;34(5):1633-1636. Available at: <https://www.biolifesas.org/biolife/wp-content/uploads/2020/10/T.C.-Theoharides-open-access.pdf>