



THE MASTOCYTOSIS SOCIETY

Mast Cell Disorders

MEDICAL REFERENCE HIGHLIGHTS

Mastocytosis and Mast Cell Activation Syndromes

FILE DESCRIPTION: This file contains selected references, *listed by topic*, that might be of interest to mast cell disorder patients, their caregivers, physicians or others. Although efforts were made to compile a list of references containing information on a variety of topics related to mast cell disorders, this is NOT a complete list of all articles available on this subject. All references were obtained through searches of the PubMed database (<http://www.ncbi.nlm.nih.gov/pubmed>). Additional individually relevant references can be obtained by searching the PubMed database. Selected additional references may also be found at www.tmsforcure.org/research/research-resources.

DISCLAIMER: Listing of an article in this file does not imply TMS support of its authors or contents and an article that is not listed in this file does not imply a lack of support of its authors or contents. **Patients should consult with their physicians, or, if necessary, mast cell specialists, regarding any questions or concerns related to applicability, accuracy and individual usefulness of information presented in these articles.**

TOPICS INCLUDED IN THIS FILE:

- International Consensus Statements, Position Papers and WHO Classification/Criteria ¹⁻¹³
- Reviews and Expert Opinions ¹⁴⁻⁵²
- Laboratory Tests, Molecular Testing, Pathology, Immunohistology and Flow Cytometry ^{5, 16, 24, 46, 48, 50, 51, 53-59}
- Perioperative Care/Pre-Medication for Dental Work, Diagnostic Testing or Surgical Procedures ^{4, 45, 60-62}
- Therapy ^{15, 16, 24, 27, 28, 44, 45, 48, 63-68}
- Mastocytosis Quality of Life and Symptom Severity Questionnaires^{69, 70}
- The Mastocytosis Society Survey on Mast Cell Disorders ⁷¹

1. Arber DA, Orazi A, Hasserjian R, Thiele J, Borowitz MJ, Le Beau MM, et al. **The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia.** Blood. 2016 May 19;127(20):2391-405. <http://www.ncbi.nlm.nih.gov/pubmed/27069254>
2. Hartmann K, Escribano L, Grattan C, Brockow K, Carter MC, Alvarez-Twose I, et al. **Cutaneous manifestations in patients with mastocytosis: Consensus report of the European Competence Network on Mastocytosis; the American Academy of Allergy, Asthma & Immunology; and the European Academy of Allergology and Clinical Immunology.** J Allergy Clin Immunol. 2016 Jan;137(1):35-45. <http://www.ncbi.nlm.nih.gov/pubmed/26476479>
3. Ustun C, Gotlib J, Popat U, Artz A, Litzow M, Reiter A, et al. **Consensus Opinion on Allogeneic Hematopoietic Cell Transplantation in Advanced Systemic Mastocytosis.** Biol Blood Marrow Transplant. 2016 Aug;22(8):1348-56. <http://www.ncbi.nlm.nih.gov/pubmed/27131865>
4. Bonadonna P, Pagani M, Aberer W, Bilo MB, Brockow K, Oude Elberink H, et al. **Drug hypersensitivity in clonal mast cell disorders: ENDA/EAACI position paper.** Allergy. 2015 Jul;70(7):755-63. <http://www.ncbi.nlm.nih.gov/pubmed/25824492>
5. Arock M, Sotlar K, Akin C, Broesby-Olsen S, Hoermann G, Escribano L, et al. **KIT mutation analysis in mast cell neoplasms: recommendations of the European Competence Network on Mastocytosis.** Leukemia. 2015 Jun;29(6):1223-32. <http://www.ncbi.nlm.nih.gov/pubmed/25650093>
6. Valent P, Escribano L, Broesby-Olsen S, Hartmann K, Grattan C, Brockow K, et al. **Proposed diagnostic algorithm for patients with suspected mastocytosis: a proposal of the European Competence Network on Mastocytosis.** Allergy. 2014 Oct;69(10):1267-74. <http://www.ncbi.nlm.nih.gov/pubmed/24836395>
7. Valent P, Sotlar K, Sperr WR, Escribano L, Yavuz S, Reiter A, et al. **Refined diagnostic criteria and classification of mast cell leukemia (MCL) and myelomastocytic leukemia (MML): a consensus proposal.** Ann Oncol. 2014 Sep;25(9):1691-700. <http://www.ncbi.nlm.nih.gov/pubmed/24675021>
8. Gotlib J, Pardanani A, Akin C, Reiter A, George T, Hermine O, et al. **International Working Group-Myeloproliferative Neoplasms Research and Treatment (IWG-MRT) & European Competence Network on Mastocytosis (ECNM) consensus response criteria in advanced systemic mastocytosis.** Blood. 2013 Mar 28;121(13):2393-401. <http://www.ncbi.nlm.nih.gov/pubmed/23325841>
9. Valent P, Akin C, Arock M, Brockow K, Butterfield JH, Carter MC, 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):215-25. <http://www.ncbi.nlm.nih.gov/pubmed/22041891>
10. Horny HP, Akin C, Metcalfe DD, Escribano L, Bennett JM, Valent P, et al. **Mastocytosis (mast cell disease)** Swerdlow SH, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H, et al., editors. World

Health Organization (WHO) Classification of Tumours. Pathology and Genetics. Tumours of Haematopoietic and Lymphoid Tissues. Lyon: IARC Press; 2008.

11. Valent P, Akin C, Escribano L, Fodinger M, Hartmann K, Brockow K, et al. **Standards and standardization in mastocytosis: consensus statements on diagnostics, treatment recommendations and response criteria.** Eur J Clin Invest. 2007 Jun;37(6):435-53. <http://www.ncbi.nlm.nih.gov/pubmed/17537151>
12. Valent P, Horny H-P, Li CY, Longley JB, Metcalfe DD, Parwaresch RM, et al. **Mastocytosis.** Jaffe ES, Harris NL, Stein H, Vardiman JW, editors. World Health Organization (WHO) Classification of Tumours. Pathology and Genetics. Tumours of Haematopoietic and Lymphoid Tissues. Lyon: IARC Press; 2001.
13. Valent P, Horny HP, Escribano L, Longley BJ, Li CY, Schwartz LB, et al. **Diagnostic criteria and classification of mastocytosis: a consensus proposal.** Leuk Res. 2001 Jul;25(7):603-25. <http://www.ncbi.nlm.nih.gov/pubmed/11377686>
14. Gonzalez-de-Olano D, Alvarez-Twose I. **Insights in Anaphylaxis and Clonal Mast Cell Disorders.** Front Immunol. 2017;8:792. <http://www.ncbi.nlm.nih.gov/pubmed/28740494>
15. Akin C. **Mast cell activation syndromes.** J Allergy Clin Immunol. 2017 Aug;140(2):349-55. <http://www.ncbi.nlm.nih.gov/pubmed/28780942>
16. Valent P, Akin C, Hartmann K, Nilsson G, Reiter A, Hermine O, et al. **Advances in the Classification and Treatment of Mastocytosis: Current Status and Outlook toward the Future.** Cancer Res. 2017 Mar 15;77(6):1261-70. <http://www.ncbi.nlm.nih.gov/pubmed/28254862>
17. Zanoni G, Zanotti R, Schena D, Sabbadini C, Opri R, Bonadonna P. **Vaccination Management In Children and Adults With Mastocytosis.** Clin Exp Allergy. 2017 Jan 12. <http://www.ncbi.nlm.nih.gov/pubmed/28079293>
18. Schuch A, Brockow K. **Mastocytosis and Anaphylaxis.** Immunol Allergy Clin North Am. 2017 Feb;37(1):153-64. <http://www.ncbi.nlm.nih.gov/pubmed/27886904>
19. Metcalfe DD, Mekori YA. **Pathogenesis and Pathology of Mastocytosis.** Annu Rev Pathol. 2017 Jan 24;12:487-514. <http://www.ncbi.nlm.nih.gov/pubmed/28135563>
20. Lei D, Akin C, Kovalszki A. **Management of Mastocytosis in Pregnancy: A Review.** J Allergy Clin Immunol Pract. 2017 Jul 21. <http://www.ncbi.nlm.nih.gov/pubmed/28739366>
21. Pardanani A. **Systemic mastocytosis in adults: 2017 update on diagnosis, risk stratification and management.** Am J Hematol. 2016 Nov;91(11):1146-59. <http://onlinelibrary.wiley.com/doi/10.1002/ajh.24553/epdf>

22. Monnier J, GeorGIN-Lavialle S, Canioni D, Lhermitte L, Soussan M, Arock M, et al. **Mast cell sarcoma: new cases and literature review.** *Oncotarget*. 2016 Oct 04;7(40):66299-309. <http://www.ncbi.nlm.nih.gov/pubmed/27602777>
23. Azana JM, Torrelo A, Matito A. **Update on Mastocytosis (Part 1): Pathophysiology, Clinical Features, and Diagnosis.** *Actas Dermosifiliogr*. 2016 Jan-Feb;107(1):5-14. <http://www.ncbi.nlm.nih.gov/pubmed/26546030>
24. Azana JM, Torrelo A, Matito A. **Update on Mastocytosis (Part 2): Categories, Prognosis, and Treatment.** *Actas Dermosifiliogr*. 2016 Jan-Feb;107(1):15-22. <http://www.ncbi.nlm.nih.gov/pubmed/26525106>
25. Bonadonna P, Bonifacio M, Lombardo C, Zanotti R. **Hymenoptera Allergy and Mast Cell Activation Syndromes.** *Curr Allergy Asthma Rep*. 2016 Jan;16(1):5. <http://www.ncbi.nlm.nih.gov/pubmed/26714690>
26. Pieri L, Bonadonna P, Elena C, Papayannidis C, Grifoni FI, Rondoni M, et al. **Clinical presentation and management practice of systemic mastocytosis. A survey on 460 Italian patients.** *Am J Hematol*. 2016 Jul;91(7):692-9. <http://www.ncbi.nlm.nih.gov/pubmed/27060898>
27. Valent P, Akin C, Metcalfe DD. **Mastocytosis 2016: Updated WHO Classification and Novel Emerging Treatment Concepts.** *Blood*. 2016 Dec 28. <http://www.ncbi.nlm.nih.gov/pubmed/28031180>
28. Gonzalez de Olano D, Matito A, Orfao A, Escribano L. **Advances in the understanding and clinical management of mastocytosis and clonal mast cell activation syndromes.** *F1000Res*. 2016 Nov 14;5:2666. <https://www.ncbi.nlm.nih.gov/pubmed/27909577>
29. Alvarez-Twose I, Jara-Acevedo M, Morgado JM, Garcia-Montero A, Sanchez-Munoz L, Teodosio C, et al. **Clinical, immunophenotypic, and molecular characteristics of well-differentiated systemic mastocytosis.** *J Allergy Clin Immunol*. 2016 Jan;137(1):168-78 e1. <http://www.ncbi.nlm.nih.gov/pubmed/26100086>
30. Carter MC, Clayton ST, Komarow HD, Brittain EH, Scott LM, Cantave D, et al. **Assessment of clinical findings, tryptase levels, and bone marrow histopathology in the management of pediatric mastocytosis.** *J Allergy Clin Immunol*. 2015 Dec;136(6):1673-9 e3. <http://www.ncbi.nlm.nih.gov/pubmed/26044856>
31. Valent P. **Diagnosis and management of mastocytosis: an emerging challenge in applied hematology.** *Hematology Am Soc Hematol Educ Program*. 2015 Dec 5;2015(1):98-105. <http://www.ncbi.nlm.nih.gov/pubmed/26637707>
32. Bonadonna P, Bonifacio M, Lombardo C, Zanotti R. **Hymenoptera Anaphylaxis and C-kit Mutations: An Unexpected Association.** *Curr Allergy Asthma Rep*. 2015 Aug;15(8):49. <http://www.ncbi.nlm.nih.gov/pubmed/26149588>

33. Theoharides TC, Valent P, Akin C. **Mast Cells, Mastocytosis, and Related Disorders.** N Engl J Med. 2015 Jul 9;373(2):163-72. <http://www.ncbi.nlm.nih.gov/pubmed/26154789>
34. Matito A, Carter M. **Cutaneous and systemic mastocytosis in children: a risk factor for anaphylaxis?** Curr Allergy Asthma Rep. 2015 May;15(5):22. <http://www.ncbi.nlm.nih.gov/pubmed/26139333>
35. Meni C, Bruneau J, Georgin-Lavialle S, Le Sache de Peuffeilhoux L, Damaj G, Hadj-Rabia S, et al. **Paediatric mastocytosis: a systematic review of 1747 cases.** Br J Dermatol. 2015 Mar;172(3):642-51. <http://www.ncbi.nlm.nih.gov/pubmed/25662299>
36. Valent P, Sotlar K, Sperr WR, Reiter A, Arock M, Horny HP. **Chronic mast cell leukemia: a novel leukemia-variant with distinct morphological and clinical features.** Leuk Res. 2015 Jan;39(1):1-5. <http://www.ncbi.nlm.nih.gov/pubmed/25443885>
37. Akin C. **Mast cell activation disorders.** J Allergy Clin Immunol Pract. 2014 May-Jun;2(3):252-7 e1; quiz 8. <http://www.ncbi.nlm.nih.gov/pubmed/24811013>
38. Akin C, Valent P. **Diagnostic criteria and classification of mastocytosis in 2014.** Immunol Allergy Clin North Am. 2014 May;34(2):207-18. <http://www.ncbi.nlm.nih.gov/pubmed/24745670>
39. Niedoszytko M, Bonadonna P, Oude Elberink JN, Golden DB. **Epidemiology, diagnosis, and treatment of Hymenoptera venom allergy in mastocytosis patients.** Immunol Allergy Clin North Am. 2014 May;34(2):365-81. <http://www.ncbi.nlm.nih.gov/pubmed/24745680>
40. Weiler CR, Butterfield J. **Mast cell sarcoma: clinical management.** Immunol Allergy Clin North Am. 2014 May;34(2):423-32. <http://www.ncbi.nlm.nih.gov/pubmed/24745684>
41. Rossini M, Zanotti R, Viapiana O, Tripi G, Orsolini G, Idolazzi L, et al. **Bone involvement and osteoporosis in mastocytosis.** Immunol Allergy Clin North Am. 2014 May;34(2):383-96. <http://www.ncbi.nlm.nih.gov/pubmed/24745681>
42. Brockow K. **Epidemiology, prognosis, and risk factors in mastocytosis.** Immunol Allergy Clin North Am. 2014 May;34(2):283-95. <http://www.ncbi.nlm.nih.gov/pubmed/24745674>
43. Moura DS, Georgin-Lavialle S, Gaillard R, Hermine O. **Neuropsychological features of adult mastocytosis.** Immunol Allergy Clin North Am. 2014 May;34(2):407-22. <http://www.ncbi.nlm.nih.gov/pubmed/24745683>
44. Carter MC, Metcalfe DD, Komarow HD. **Mastocytosis.** Immunol Allergy Clin North Am. 2014 Feb;34(1):181-96. <http://www.ncbi.nlm.nih.gov/pubmed/24262698>
45. Fried AJ, Akin C. **Primary mast cell disorders in children.** Curr Allergy Asthma Rep. 2013 Dec;13(6):693-701. <http://www.ncbi.nlm.nih.gov/pubmed/24150753>

46. Valent P. **Mast cell activation syndromes: definition and classification.** Allergy. 2013 Apr;68(4):417-24. <http://www.ncbi.nlm.nih.gov/pubmed/23409940>
47. Georjin-Lavialle S, Lhermitte L, Dubreuil P, Chandesris MO, Hermine O, Damaj G. **Mast cell leukemia.** Blood. 2013 Feb 21;121(8):1285-95. <http://www.ncbi.nlm.nih.gov/pubmed/23243287>
48. Cardet JC, Castells MC, Hamilton MJ. **Immunology and clinical manifestations of non-clonal mast cell activation syndrome.** Curr Allergy Asthma Rep. 2013 Feb;13(1):10-8. <http://www.ncbi.nlm.nih.gov/pubmed/23212667>
49. Afrin LB. Presentation, diagnosis and management of mast cell activation syndrome. In: Murray DB, editor. Mast cells: phenotypic features, biological functions and role in immunity. Hauppauge: Nova Science Publishers, Inc.; 2013. p. 155-232.
50. Alvarez-Twose I, Morgado JM, Sanchez-Munoz L, Garcia-Montero A, Mollejo M, Orfao A, et al. **Current state of biology and diagnosis of clonal mast cell diseases in adults.** Int J Lab Hematol. 2012 Oct;34(5):445-60. <http://www.ncbi.nlm.nih.gov/pubmed/22551157>
51. Torrelo A, Alvarez-Twose I, Escribano L. **Childhood mastocytosis.** Curr Opin Pediatr. 2012 Aug;24(4):480-6. <http://www.ncbi.nlm.nih.gov/pubmed/22790101>
52. Akin C, Valent P, Metcalfe DD. **Mast cell activation syndrome: proposed diagnostic criteria.** J Allergy Clin Immunol. 2010 Dec;126(6):1099-104 e4. <http://www.ncbi.nlm.nih.gov/pubmed/21035176>
53. Teodosio C, Mayado A, Sanchez-Munoz L, Morgado JM, Jara-Acevedo M, Alvarez-Twose I, et al. **The immunophenotype of mast cells and its utility in the diagnostic work-up of systemic mastocytosis.** J Leukoc Biol. 2015 Jan;97(1):49-59. <http://www.ncbi.nlm.nih.gov/pubmed/25381388>
54. Horny HP, Sotlar K, Valent P. **Mastocytosis: immunophenotypical features of the transformed mast cells are unique among hematopoietic cells.** Immunol Allergy Clin North Am. 2014 May;34(2):315-21. <http://www.ncbi.nlm.nih.gov/pubmed/24745676>
55. Doyle LA, Hornick JL. **Pathology of extramedullary mastocytosis.** Immunol Allergy Clin North Am. 2014 May;34(2):323-39. <http://www.ncbi.nlm.nih.gov/pubmed/24745677>
56. Horny HP, Sotlar K, Valent P. **Evaluation of mast cell activation syndromes: impact of pathology and immunohistology.** Int Arch Allergy Immunol. 2012;159(1):1-5. <http://www.ncbi.nlm.nih.gov/pubmed/22555026>
57. Horny HP, Sotlar K, Valent P. **Mastocytosis: state of the art.** Pathobiology. 2007;74(2):121-32. <http://www.ncbi.nlm.nih.gov/pubmed/17587883>

58. Horny HP, Valent P. **Diagnosis of mastocytosis: general histopathological aspects, morphological criteria, and immunohistochemical findings.** Leuk Res. 2001 Jul;25(7):543-51. <http://www.ncbi.nlm.nih.gov/pubmed/11377679>
59. Jawhar M, Schwaab J, Hausmann D, Clemens J, Naumann N, Henzler T, et al. **Splenomegaly, elevated alkaline phosphatase and mutations in the SRSF2/ASXL1/RUNX1 gene panel are strong adverse prognostic markers in patients with systemic mastocytosis.** Leukemia. 2016 Dec;30(12):2342-50. <http://www.ncbi.nlm.nih.gov/pubmed/27416984>
60. Matito A, Morgado JM, Sanchez-Lopez P, Alvarez-Twose I, Sanchez-Munoz L, Orfao A, et al. **Management of Anesthesia in Adult and Pediatric Mastocytosis: A Study of the Spanish Network on Mastocytosis (REMA) Based on 726 Anesthetic Procedures.** Int Arch Allergy Immunol. 2015;167(1):47-56. <http://www.ncbi.nlm.nih.gov/pubmed/26160029>
61. Dewachter P, Castells MC, Hepner DL, Mouton-Faivre C. **Perioperative Management of Patients with Mastocytosis.** Anesthesiology. 2013 Oct 16. <http://www.ncbi.nlm.nih.gov/pubmed/24135579>
62. Carter MC, Uzzaman A, Scott LM, Metcalfe DD, Quezado Z. **Pediatric mastocytosis: routine anesthetic management for a complex disease.** Anesth Analg. 2008 Aug;107(2):422-7. <http://www.ncbi.nlm.nih.gov/pubmed/18633019>
63. Gotlib J. **Tyrosine Kinase Inhibitors in the Treatment of Eosinophilic Neoplasms and Systemic Mastocytosis.** Hematol Oncol Clin North Am. 2017 Aug;31(4):643-61. <http://www.ncbi.nlm.nih.gov/pubmed/28673393>
64. DeAngelo DJ, George TI, Linder A, Langford C, Perkins C, Ma J, et al. **Efficacy and safety of midostaurin in patients with advanced systemic mastocytosis: 10-year median follow-up of a phase II trial.** Leukemia. 2017 Jul 24. <http://www.ncbi.nlm.nih.gov/pubmed/28744009>
65. Gotlib J, Kluin-Nelemans HC, George TI, Akin C, Sotlar K, Hermine O, et al. **Efficacy and Safety of Midostaurin in Advanced Systemic Mastocytosis.** N Engl J Med. 2016 Jun 30;374(26):2530-41. <http://www.ncbi.nlm.nih.gov/pubmed/27355533>
66. Rossini M, Zanotti R, Orsolini G, Tripi G, Viapiana O, Idolazzi L, et al. **Prevalence, pathogenesis, and treatment options for mastocytosis-related osteoporosis.** Osteoporos Int. 2016 Aug;27(8):2411-21. <http://www.ncbi.nlm.nih.gov/pubmed/26892042>
67. Ustun C, Arock M, Kluin-Nelemans HC, Reiter A, Sperr WR, George T, et al. **Advanced systemic mastocytosis: from molecular and genetic progress to clinical practice.** Haematologica. 2016 Oct;101(10):1133-43. <http://www.ncbi.nlm.nih.gov/pubmed/27694501>
68. Arock M, Akin C, Hermine O, Valent P. **Current treatment options in patients with mastocytosis: status in 2015 and future perspectives.** Eur J Haematol. 2015 Jun;94(6):474-90. <http://www.ncbi.nlm.nih.gov/pubmed/25753531>

69. Siebenhaar F, von Tschirnhaus E, Hartmann K, Rabenhorst A, Staubach P, Peveling-Oberhag A, et al. **Development and validation of the mastocytosis quality of life questionnaire: MC-QoL.** Allergy. 2016 Jun;71(6):869-77. <http://www.ncbi.nlm.nih.gov/pubmed/26797792>
70. van Anrooij B, Kluin-Nelemans JC, Safy M, Flokstra-de Blok BM, Oude Elberink JN. **Patient-reported disease-specific quality-of-life and symptom severity in systemic mastocytosis.** Allergy. 2016 Nov;71(11):1585-93. <http://www.ncbi.nlm.nih.gov/pubmed/27089859>
71. 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.** J Allergy Clin Immunol Pract. 2014 Jan-Feb;2(1):70-6. <http://www.ncbi.nlm.nih.gov/pubmed/24565772>