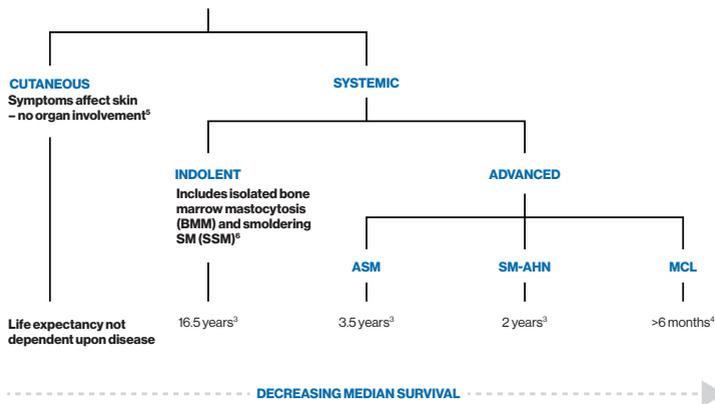


Advanced Systemic Mastocytosis (AdvSM)

Advanced Systemic Mastocytosis is a group of rare mast cell disorders where the disease has spread to more than one part of your body, and is stopping your body from working¹

Types of Mastocytosis²



If you have been told you have Advanced Systemic Mastocytosis, it is likely you will have one of these three types:²

1. Aggressive systemic Mastocytosis (ASM)
2. Systemic Mastocytosis with Associated Hematologic Neoplasm (SM-AHN)
3. Mantle Cell Leukaemia (MCL)

There is no cure for Advanced Systemic Mastocytosis, but there are treatments that can help.⁷

Some symptoms of AdvSM may include:



Losing weight without trying²



Fever or chills⁸



Bone pain⁸



Swollen lymph nodes, liver or spleen²



Tiny red spots on your skin⁸

Other symptoms may include:

Poor appetite, pain when you go to the toilet, anaemia, stomach problems such as feeling sick, being sick or the runs, anaphylactic shock, low blood pressure, low platelet count, low levels of albumin and too many bone marrow blasts.^{2,9,10}

Tests for AdvSM may include a combination of:⁷



Full blood count and blood tryptase levels



Bone marrow test (Biopsy)



Ultrasound scan



DEXA scan (Bone scan)

Other tests may include:¹¹

Samples taken from areas such as your spleen or lymph nodes to check for the number or shape of mast cells in other parts of the body

Questions to ask your doctor:



- What type of mastocytosis do I have?
- Is my mastocytosis a type of cancer?
- Should I carry an EpiPen?
- How will advanced SM affect me?
- What things can I no longer do?
- What are my treatment options?
- Are there any risks to the treatment?
- What are the side effects of the treatment?
- Will my symptoms go away after I start treatment?
- How will I know if my treatment is working?
- If treatment is not helping, what are my options?
- Should I take part in a clinical trial?
- How long do people with this disease live?
- Are there support groups I can join?

Treatments for AdvSM:

There is no cure for AdvSM but there are treatments available.⁹

There are two types of treatment approaches for AdvSM:

- Treatments to treat the symptoms, also known as supportive treatments. These reduce the symptoms that can cause quality of life issues.
- Treatments to stop the mast cells from growing and spreading. When mast cells grow too fast, they can stop some of the body's organs from working properly. Although treatment can't reduce the number of mast cells in your body, the treatments can try to stop them spreading.

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1. Valent et al. Blood. Mastocytosis 2016: updated WHO classification and novel emerging treatment concepts. 2017 Mar 16; 129(11): 1420-1427. 2. Pardanani A. Am J Hematol. 2015;90:251-262. 3. Lim KH, Te eri A, Lasho T et al. Blood. 2009;113:5727-5736. 4. Georjin-Lavaille S, Lhermitte L, Dubreuil P et al. Blood. 2013;121:1285-1295. 5. National Institutes of Health. Cutaneous mastocytosis. <https://rarediseases.info.nih.gov/diseases/7342/cutaneous-mastocytosis>. Accessed September 20, 2019. 6. National Institutes of Health. Current concepts on diagnosis and treatment of mastocytosis. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4000464/>. Accessed September 20, 2019. 7. National Health Service. Overview- Mastocytosis. <https://www.nhs.uk/conditions/mastocytosis/>. Accessed September 20, 2019. 8. OncoLive. Subtypes and Characteristics of Mastocytosis. <https://www.oncolive.com/insights-archival/aml-asm-advancements/subtypes-and-characteristics-of-asm>. Accessed September 23, 2019. 9. Cancer.net. Mastocytosis: Treatment Options. <https://www.cancer.net/cancer-types/mastocytosis/treatment-options>. Accessed September 23, 2019. 10. The Mastocytosis Society. Overview and diagnosis. <https://traffordcare.org/overview/>. Accessed September 23, 2019. 11. Horry H. Am J Clin Pathol. Mastocytosis: An Unusual Clonal Disorder of Bone Marrow-Derived Hematopoietic Progenitor Cells. 2009;132:438-447.

