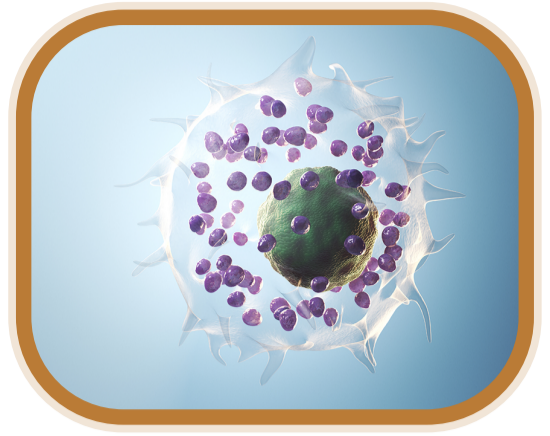


Learning About Systemic Mastocytosis (SM)

What is a mast cell?

Before you learn about systemic mastocytosis (SM), it's important to first know what a mast cell is.

A mast cell is a type of white blood cell that is a key part of the immune system. Mast cells are designed to help keep the body safe from diseases and infections.



What causes SM?

If mast cells are abnormal and uncontrolled, they often increase or spread throughout the body releasing proteins. These proteins are often presented as allergic reactions, inflammation, and other immune responses. SM is a rare disease that results from the spreading of these abnormal mast cells.

In about 95% of cases, SM forms because of a genomic change known as *KIT D816V*. This change (or mutation) leads to the production of too many abnormal mast cells.¹

95%

What can happen as a result of SM?

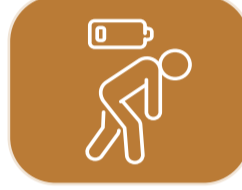
SM can lead to attacks that look and act like severe allergic reactions. As a result of these attacks, a few of the following symptoms may present impacting the daily life of people living with SM:



Anaphylaxis
(a serious allergic reaction)



Stomach or digestive issues



Fatigue



Bone pain



Itching



Flushing



Migraines

What are the types of SM?

Non-advanced SM

about ~90% of SM cases^{2,3}

Advanced SM

about ~10% of SM cases^{2,3}

Non-advanced SM

Non-advanced SM may cause many difficult symptoms. Subtypes of non-advanced SM:

Bone marrow mastocytosis (BMM)

In BMM, abnormal mast cells build up only in the bone marrow, not the skin.

Indolent SM (ISM)

ISM is the most common type of SM. Abnormal mast cells are mainly found in the bone marrow but also in other parts of the body, like the skin.

Smouldering SM (SSM)

SSM is rare and may progress slowly. Abnormal mast cells build up in the bone marrow and organs, and can cause more significant symptoms.

Advanced SM

Advanced SM occurs when abnormal mast cells continue to build and form throughout the body. This can lead to potentially life-threatening organ damage. Subtypes of advanced SM:

Aggressive SM (ASM)

A severe form of SM, in which abnormal mast cells can cause organ damage.

SM with an associated hematological neoplasm (SM-AHN)

SM-AHN occurs in combination with other blood or bone marrow disorders.

Mast cell leukemia (MCL)

MCL affects the bone marrow and white blood cells. It is extremely rare, but an aggressive form of SM.

How many people have SM?

SM is a rare disease that affects

1 in 10,000 people^{4*}

20–50 years old

People with SM may first start to notice signs or symptoms at 20–50 years of age²

~7 years

People with SM report averaging 7 years from first noticing symptoms to getting a diagnosis^{5†}

To gain a better understanding of SM, visit: www.navigatingSM.com

¹Based on a Swedish cohort of 195 adults with SM diagnosed in 2006–2020.
²Based on data from 149 patients with self-reported mastocytosis in Mast Cell Connect registry in Jennings 2018 study.

References: 1. Kristensen T, et al. *J Mol Diagn.* 2011;13(2):180–188. 2. Gülen T, et al. *J Intern Med.* 2016;279(3):211–228. 3. Pardanani A. *Am J Hematol.* 2021;96(4):508–525. 4. Ungerstedt J, et al. *Cancers (Basel).* 2022;14(16):3942. 5. Jennings SV, et al. *Immunol Allergy Clin North Am.* 2018;38(3):505–525.