

MANTLE CELL LYMPHOMA:

The Facts

What Is Mantle Cell Lymphoma?

Mantle cell lymphoma (MCL) is a sub-type of lymphoma—a cancer of white blood cells called lymphocytes, which normally help the body fight infection. Lymphomas, which usually begin in the lymph nodes, are divided in two categories: Hodgkin's lymphoma and non-Hodgkin's lymphoma (NHL), which is further divided into several sub-types, including MCL.¹

MCL accounts for less than 10% of NHLs.²

Symptoms of MCL and Other Forms of NHL³

- Swollen, painless lymph nodes in the neck, armpits or groin
- Unexplained weight loss
- Fever
- Soaking night sweats
- Coughing, trouble breathing or chest pain
- Weakness and tiredness that don't go away
- Pain, swelling or a feeling of fullness in the abdomen

Complications of MCL¹

Complications that arise from disease progression may include:

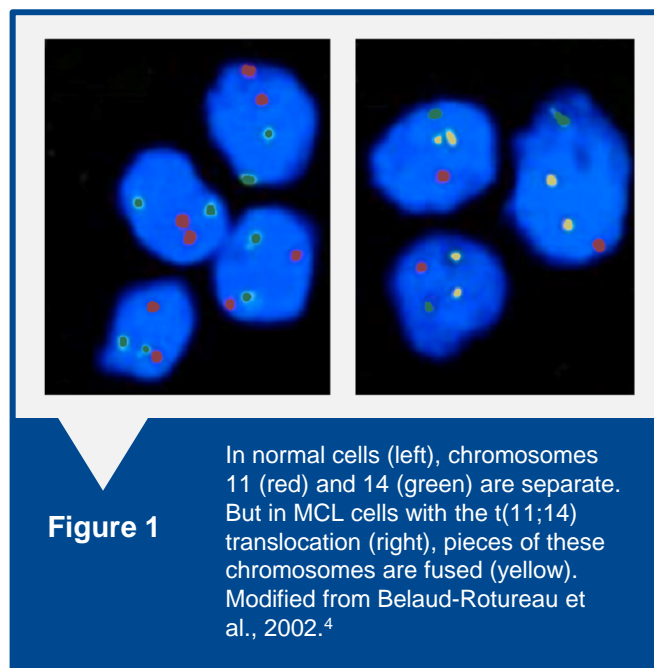
- Low blood counts (known as cytopenias), when cancer cells reach the bone marrow:
 - Neutropenia (low **white blood cell** counts)
 - Anemia (**low red blood cell** counts)
 - Thrombocytopenia (low numbers of **platelets**)
- Complications of the digestive track, lungs or brain, when the tumor cells reach these organs
- High white blood cell counts (known as leukocytosis), if the disease progresses to leukemia

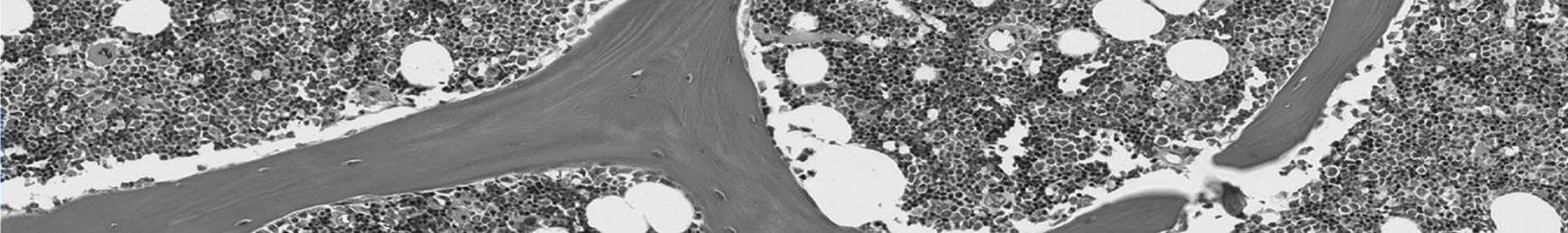
What Causes MCL?

MCL results when a white blood cell type known as a B lymphocyte grows in an uncontrolled manner in the outer edge of a lymph node follicle (called the mantle zone). The result is swelling of the lymph nodes. The MCL cells can enter the lymphatic system and the blood and spread to other lymph nodes or to tissues such as the liver, bone marrow and gastrointestinal tract.¹

In about 85 percent of MCL cases, patients have a genetic change in developing B cells that results in DNA on chromosomes 11 and 14 exchanging places (Figure 1). This swap, known as the t(11;14) translocation, causes the cells to produce too much of a protein called cyclin D1, which instructs the cells to divide uncontrollably.¹

Patients without this translocation usually have other genetic changes that cause cyclin levels to rise.⁴





Who Gets MCL?

- About 4,000 new cases of MCL are diagnosed in the United States each year¹
- Roughly half of people with MCL are over the age of 68 at the time of diagnosis⁵
- This type of lymphoma affects men more often than women⁴
- White males and females are at a higher risk than black males and females¹

What Is the Prognosis for Patients with MCL?

Because it is both incurable and rapidly growing, MCL has one of the worst prognoses of all lymphomas.²

With currently approved therapies, estimated survival is only 4–5 years after initial treatment.⁶⁻⁸ Following relapse, only 1 or 2 additional years of survival are expected with current therapeutic options.

How Is MCL Treated?

There is no generally accepted standard treatment for MCL. Chemotherapy in combination with rituximab followed by a stem cell transplant is one common first-line treatment for young, fit patients.⁹ Unfortunately, the majority of MCL patients are not young and fit, so these types of treatments are not always applicable.²

If the MCL does not respond to initial treatment or if the disease returns, patients may be treated with a different combination of drugs.²

Because patient outcomes are still less than optimal, researchers continue to develop and test new approaches to treatment.

¹ Leukemia and Lymphoma Society. Mantle Cell Lymphoma Facts. July 2012.

² Li ZM, Zucca E, Ghielmini M. Open questions in the management of mantle cell lymphoma. *Cancer Treat Rev.* 2013. doi: 10.1016/j.ctrv.2012.12.013.

³ NCI. What You Need to Know About™ Non-Hodgkin Lymphoma. 2008.

⁴ Belaud-Rotureau M-A, et al. A Comparative Analysis of FISH, RT-PCR, PCR, and Immunohistochemistry for the Diagnosis of Mantle Cell Lymphomas. *Mod Pathol.* 2002; 15:517–525.

⁵ Shah BD, Martin P, Sotomayor EM. Mantle cell lymphoma: a clinically heterogeneous disease in need of tailored approaches. *Cancer Control.* 2012; 19:227-235.

⁶ Goy A, Kahl B. Mantle cell lymphoma: the promise of new treatment options. *Crit Rev Oncol Hematol.* 2011; 80:69-86.

⁷ McKay P, Leach M, Jackson R, et al. Guidelines for the investigation and management of mantle cell lymphoma. *Br J Haematol.* 2012; 159:405-426.

⁸ Vose JM: Mantle cell lymphoma: 2012 update on diagnosis, risk-stratification, and clinical management. *Am J Hematol.* 2012; 87:604-609.

⁹ National Comprehensive Cancer Network. Clinical Practice Guidelines in Oncology™. Non-Hodgkin's Lymphomas (V1.2010). Accessed May 31, 2013. <http://www.24hmb.com/UpLoad/Editor/2010/1/4/2010010471345169.pdf>