Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Mantle cell lymphoma (MCL) is a rare B-cell NHL, comprising about 6 percent of all NHL cases in the United States. Read below for more about the diagnosis, treatment, and prognosis of MCL. This "Patient Education" tear sheet was produced in collaboration with the Lymphoma Research Foundation (LRF) (www.lymphoma.org).

**What is Mantle Cell Lymphoma?**
Lymphoma occurs when cells of the immune system called lymphocytes, a type of white blood cell, grow and multiply uncontrollably. The body has two main types of lymphocytes that can develop into lymphomas: B lymphocytes (B cells) and T lymphocytes (T cells). MCL is a rare B-cell NHL. The disease is called “mantle cell lymphoma” because the tumor cells originally come from the “mantle zone” of the lymph node.

**How is Mantle Cell Lymphoma Diagnosed?**
MCL is usually diagnosed as a late-stage disease that has spread to the gastrointestinal tract and bone marrow.
A diagnosis of MCL requires taking a small sample of tumor tissue, called a biopsy, and looking at the cells under a microscope. A blood test may also be necessary to measure the white blood cell count and certain proteins, which help to diagnose MCL.
Other tests, such as a bone marrow biopsy, computed axial tomography (CAT) scan, or positron emission tomography/computed tomography (PET/CT) scan may be used to confirm a diagnosis and to determine what areas of the body are involved by the cancer.
Overproduction of a protein called cyclin D1 is found in more than 90 percent of patients with MCL. Each cyclin D1 from a biopsy is considered a very sensitive tool for diagnosing MCL.
One-quarter to one-half of patients with MCL also have higher-than-normal levels of certain proteins that circulate in the blood, such as lactate dehydrogenase (LDH) and beta-2 microglobulin. Measuring these and other proteins can help physicians determine how aggressive the disease may be and how to treat it.

**How is Mantle Cell Lymphoma Treated?**
The type of treatment selected for a patient with MCL depends on multiple factors, including the stage of disease, the age of the patient, and the patient’s overall health.

**Watchful Waiting**
For the subset of patients who do not yet have symptoms and who have a relatively small amount of slow-growing disease, monitoring the disease for progression may be an acceptable option.

**Chemotherapy**
Initial treatment approaches for aggressive MCL in younger patients include combination chemotherapy, typically with the monoclonal antibody rituximab as first-line treatment, followed by autologous hematopoietic cell transplantation (HCT).
HyperCVAD (cyclophosphamide, vincristine, doxorubicin, and dexamethasone alternating with methotrexate and cytarabine) plus rituximab are recommended as aggressive induction therapy and are associated with durable remissions in newly diagnosed patients.
For older patients, chemotherapy followed by a prolonged course of rituximab alone, known as maintenance therapy, is often recommended.
A common chemotherapy treatment approach used to treat MCL is called R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). Bendamustine in combination with rituximab is another common first-line treatment option. Several additional intensive chemotherapy combinations are also used in combination with rituximab, particularly in younger patients.

**Proteasome Inhibitors**
These drugs disrupt a molecular pathway that is critical for the elimination of proteins in both normal and cancer cells. Bortezomib is a proteasome inhibitor that has been approved by the U.S. Food and Drug Administration for the treatment of MCL patients who have received at least one prior therapy.

**Hematopoietic Cell Transplantation**
There are two types of HCTs: autologous (patients receive stem cells from another person) and allogeneic (patients receive their own cells).
Debate exists among researchers regarding which type of transplant is most efficacious and whether or when transplant should be used in the treatment of MCL. High-dose chemotherapy coupled with HCT can be used to treat MCL patients who have failed initial chemotherapy, but are responsive to a second chemotherapy regimen. Some researchers feel that autologous HCT is better for patients who have had a relapse and that autologous HCT should only be used to treat patients as part of initial therapy.

**Clinical Trials**
Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Because the optimal initial treatment of MCL is not clear and it is such a rare disease, clinical trials are very important and will identify the best treatment options in this disease. Patients interested in participating in a clinical trial should speak with their physician to discuss clinical trial opportunities.

**Follow-Up**
Patients with MCL in remission should have regular visits with a physician who is familiar with their medical history and the treatments they have received. Medical tests may be required at various times during remission to evaluate the need for additional treatment.

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**Lymphoma Research Foundation Patient Resources**
The Lymphoma Research Foundation (LRF) offers patients with lymphoma and chronic lymphocytic leukemia a wide range of resources that provide a comprehensive overview as well as address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma.
LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as an understanding Non-Hodgkin Lymphoma booklet e-Updates that provide tools to help manage the disease.
For more information about any of these resources, visit the Lymphoma Research Foundation’s websites at www.lymphoma.org or www.FocusOnMCL.org or contact the LRF Helpline at 800-500-9976 or helpline@lymphoma.org.