

Follicular Lymphoma

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Lymphoma occurs when cells of the immune system called lymphocytes, a type of white blood cell, grow and multiply uncontrollably. Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood, or other organs, and form a mass called a tumor. The body has two main types of lymphocytes that can develop into lymphomas: B lymphocytes (B cells) and T lymphocytes (T cells).

Follicular lymphoma (FL) is the most common indolent (slow-growing) form of NHL, accounting for approximately 20 percent of all B-cell NHLs. Common symptoms of FL include enlargement of the lymph nodes in the neck, underarms, abdomen, or groin, as well as fatigue, shortness of breath, night sweats, and weight loss. Often, patients with FL have no obvious symptoms of the disease at diagnosis.

Over time, some patients with FL may eventually develop a transformed lymphoma, which is often more aggressive and usually requires more intensive types of treatment. For more information on transformed lymphomas, view the *Transformed Lymphomas* fact sheet on the Lymphoma Research Foundation's (LRF) website at www.lymphoma.org.

Treatment Options

There are various treatment options for FL based on the severity of associated symptoms and the rate of cancer growth. If patients show no or very few symptoms, physicians may decide not to treat the disease right away, an approach referred to as "watch and wait" or "watchful waiting." Studies have shown that patients suitable for a "watch and wait" approach have outcomes similar to those being treated early in the course of their disease.

FL is generally very responsive to radiation and chemotherapy. Radiation can provide a cure in some patients with limited disease. In more advanced stages, physicians may use one or more chemotherapy drugs or the monoclonal antibody rituximab (Rituxan), alone or in combination with other agents. Monoclonal antibodies can act more directly than chemotherapy agents by targeting particular markers found on tumor cells and recruiting immune

cells to promote tumor destruction, which can increase a patient's response to treatment. Common combination regimens include:

- R-Bendamustine (rituximab and bendamustine)
- R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone)
- R-CVP (rituximab, cyclophosphamide, vincristine, and prednisone)

Rituximab can also be used as maintenance therapy in order to prolong remission for patients with no signs of lymphoma. Another treatment sometimes used for FL is ibritumomab tiuxetan (Zevalin), which is a radioactive particle connected to an antibody that targets cancer cells.

After treatment, many patients can go into a remission that lasts for years; however, the disease often returns. For these patients, additional therapies are often successful in providing another remission. For more information on relapsed (disease returns after treatment) and refractory (disease does not respond to treatment) FL, view the *Follicular Lymphoma: Relapsed/Refractory* fact sheet on LRF's website at www.lymphoma.org.

The U.S. Food and Drug Administration granted idelalisib (Zydelig) accelerated approval to treat patients with relapsed follicular B-cell NHL, as well as small lymphocytic lymphoma and chronic lymphocytic leukemia. Idelalisib is a phosphoinositide 3-kinase (PI3K) delta inhibitor, blocking cell signals known to be highly active in tumor cells and needed for growth and survival of those cells. For some patients with relapsed FL, high-dose chemotherapy and stem cell transplantation may be an option. For more information on transplantation, view the *Transplant in Lymphoma* fact sheet on LRF's website at www.lymphoma.org.

Treatments Under Investigation

Many treatments are currently being tested in clinical trials for patients who are newly diagnosed or have relapsed/refractory FL. For example, the combination of rituximab and the immunomodulatory drug lenalidomide (Revlimid) is being assessed in patients who have not previously received treatment for FL. The combination of bendamustine (Treanda) and the monoclonal antibody ofatumumab (Arzerra) is being investigated

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for patients with untreated FL with and without the proteasome inhibitor bortezomib (Velcade). Trials are also ongoing with the Bruton's tyrosine kinase inhibitor ibrutinib (Imbruvica) in combination with rituximab for patients with previously untreated FL. Combinations of treatment modalities, immunochemotherapy, ibritumomab tiuxetan, and stem cell transplantation are under investigation and may help patients achieve prolonged remission. It is critical to remember that today's scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with their physician or with LRF for any treatment updates that may have recently emerged.

Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Patients interested in participating in a clinical trial should talk to their physician or contact the LRF Helpline for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

Follow-up

Because FL is generally characterized by multiple disease relapses after responses to a variety of treatments, patients in remission should have regular visits with a physician who is familiar with their medical history and the treatments they have received. Medical tests (such as blood tests and computed axial tomography [CAT] scans) may be required at various times during remission to evaluate the need for additional treatment.

Some treatments can cause long-term effects or late effects, which can vary based on duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. A physician will check for these effects during follow-up care. Visits may become less frequent the longer the disease remains in remission.

Patients and their caregivers are encouraged to keep copies of all medical records and test results as well as information on the types, amounts, and duration of all treatments received. This documentation will be important for keeping track of any effects resulting from treatment or potential disease recurrences.

Support

A lymphoma diagnosis often triggers a range of feelings and raises concerns. In addition, cancer treatment can cause physical discomfort. Support groups and online message boards can help patients connect with other people who have lymphoma. One-to-one peer support programs, such as the LRF Lymphoma Support Network, match lymphoma patients (or caregivers) with volunteers who have gone through similar experiences.

Resources

LRF offers a wide range of resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as E-Updates that provide the latest disease-specific news and treatment options. For more information about any of these resources, visit our websites at www.FocusOnFL.org or www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.