

Diffuse Large B-Cell Lymphoma

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma 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).

B-cell lymphomas are much more common than T-cell lymphomas and account for approximately 85 percent of all NHLs. Diffuse large B-cell lymphoma (DLBCL) is the most common form of NHL, accounting for about 30 percent of newly diagnosed cases of NHL in the United States. DLBCL occurs in both men and women, although it is slightly more common in men. Although DLBCL can occur in childhood, its incidence generally increases with age and most patients are over the age of 60.

DLBCL is an aggressive (fast-growing) lymphoma that can arise in lymph nodes or outside of the lymphatic system, in the gastrointestinal tract, testes, thyroid, skin, breast, bone, or brain. There are several subtypes of DLBCL that may affect a patient's prognosis (how well a patient will do with standard treatment) and treatment options. For example, DLBCL that only affects the brain is called "primary central nervous system lymphoma" and is treated differently than DLBCL that affects areas outside of the brain. Another example is "primary mediastinal B-cell lymphoma," which often occurs in younger patients and grows rapidly in the chest (mediastinum). In addition, there are genetic and biologic markers in some DLBCLs that may be important for both prognosis and treatment choices.

Often, the first sign of DLBCL is a painless, rapid swelling in the neck, underarms, or groin that is caused by enlarged lymph nodes. For some patients, the swelling may be painful. Other symptoms may include night sweats, fever, and unexplained weight loss. Patients may notice fatigue, loss of appetite, shortness of breath (trouble breathing), or pain.

Diagnosis and Staging

A tissue biopsy is needed for a definitive diagnosis of DLBCL. A biopsy is usually a small surgical procedure to remove part or all of an affected lymph node or other abnormal area in order to look at it under the microscope. This can be done under local or general anesthesia.

Once the diagnosis of DLBCL is confirmed, the next step (called staging) is to understand how much lymphoma is present and where it is located in the body. Because DLBCL is a type of blood cancer, it is important to look at the entire body to determine all the locations of the lymphoma. This is usually done with a whole-body computed axial tomography (CAT) scan or positron emission tomography (PET)/CAT scan. Staging may also include a bone marrow biopsy to look for lymphoma cells in the bone and sometimes a spinal tap (lumbar puncture) to determine if there are lymphoma cells in the brain and spinal cord. The physician will use the results of these tests to assess the stage of the lymphoma. Limited stage disease represents lymphoma affecting only one area of the body. Advanced stage disease indicates that lymphoma has spread to several organs. Staging is needed to choose an appropriate course of treatment.

Prior to treatment, there are several blood tests that are important in determining if organs are healthy and functioning sufficiently to withstand the effects of treatment. These tests can include a complete blood count, liver and kidney function tests, and a lactate dehydrogenase test, which is a general indicator of tissue damage.

Treatment Options

Because DLBCL can advance quickly, it usually requires immediate treatment. A combination of chemotherapy and the monoclonal antibody rituximab (Rituxan), with or without radiation therapy, can lead to disease remission in a large number of patients with this form of lymphoma. The most widely used treatment for DLBCL is R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) that is usually given in 21-day cycles, although the number of cycles given can vary. Sometimes another chemotherapy drug, etoposide (Vepesid, Toposar, Etopophos), is added to the R-CHOP regimen, resulting in a drug combination called R-EPOCH. For many patients, the initial treatment is effective and DLBCL does not return after treatment; however, the disease does return for some patients. For patients where the disease becomes refractory (disease does not respond to treatment) or relapses (disease returns after treatment), secondary therapies may be successful.

High-dose chemotherapy coupled with stem cell transplantation can be used to treat patients with DLBCL whose disease is refractory or relapsed following initial chemotherapy. The majority of patients undergoing a stem cell transplant will receive their own stem cells (autologous stem cell transplant). Occasionally, a patient will receive stem cells from a donor (allogeneic stem cell transplant). For more information on stem cell transplants, view the

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Transplant in Lymphoma fact sheet on the Lymphoma Research Foundation's (LRF's) website at www.lymphoma.org.

Relapsed/refractory patients who are not candidates for stem cell transplant, or who choose not to have a stem cell transplant, do have various combination chemotherapy regimens that can be used for treatment. Bendamustine (Treanda) plus rituximab, single-agent rituximab, lenalidomide (Revlimid) plus rituximab, and gemcitabine-based combinations are secondary therapies that may also be used in these patients.

Treatments Under Investigation

Many promising therapies are currently being tested in clinical trials for the treatment of patients with newly diagnosed or relapsed/refractory DLBCL, including:

- Bortezomib (Velcade)
- Chimeric antigen receptor therapy (CAR-T)
- Everolimus (Afinitor)
- Ibrutinib (Imbruvica)
- Lenalidomide (Revlimid)
- Nivolumab (Opdivo)
- Panobinostat (Farydak)

Clinical trials are investigating the use of these agents at various treatment stages (first-line, maintenance, etc.) and for specific patient populations, including newly diagnosed patients, patients with relapsed/refractory disease, and the elderly. The clinical trials investigating these drugs are in various phases of development. 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 view the *Understanding Clinical Trials* fact sheet on LRF's website at www.lymphoma.org, 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

Patients with lymphoma 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 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.

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 disease-specific websites, videos, and eNewsletters for current lymphoma information and treatment options. To learn more about any of these resources, visit our website at www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.