

Understanding CNS Lymphoma

Non-Hodgkin Lymphoma (NHL) is broadly categorized as B-cell lymphomas or T-cell lymphomas. B-cell lymphomas develop from abnormal B cells and account for approximately 90 percent of all NHLs. T-cell lymphomas develop from abnormal T cells and account for about 5 to 10 percent of all NHLs. NHLs may also be classified as *indolent* (slow-growing) or *aggressive* (fast-growing).

Primary central nervous system (CNS) lymphoma is an aggressive form of NHL in which *malignant* (cancer) cells exclusively in the central nervous system including the brain, spinal cord, cerebrospinal fluid and/or eye. In more than 90 percent of cases, it is a diffuse large B-cell lymphoma. When lymphoma has originated in other parts of the body and subsequently has spread to the CNS, it is referred to as secondary CNS lymphoma.

The cause of CNS lymphoma is unknown, but there are some factors that may increase the risk of developing it such as having a compromised immune system (for example, people with acquired immunodeficiency syndrome [AIDS] or patients who have undergone organ transplant). Having one or more of these risk factors does not mean a person will develop NHL. Most people with risk factors never develop the disease and most people diagnosed have never been exposed to any clearly identifiable risk factors.

The common symptoms of CNS lymphoma are focal neurological deficits based on which part of the CNS is involved by the cancer (such as language problems, weakness, paralysis, loss of vision), but headaches, vomiting, confusion, seizures, personality changes, and blurred vision can also occur. Symptoms can occur suddenly or develop over time.

Early recognition of symptoms and diagnosis of CNS lymphoma are critical, followed by prompt initiation of treatment in order to reverse neurological deficits and improve outcomes in this disease.

TREATMENT OPTIONS

Until the mid-1990s, radiation was the standard therapy for patients with CNS lymphoma. Now, high-dose methotrexate-based therapy with rituximab (Rituxan) is recommended for most patients. Sometimes, this is given in combination with other agents such as cytarabine (Cytosar), vincristine (Oncovin), procarbazine (Matulane), ifosfamide (Ifex), thiotepa (Tepadina) or temozolomide (Temodar). Furthermore, this initial treatment, or induction, is often followed by other therapy regimens, or consolidation, to reduce the risk of recurrence. These consolidation therapies include high-dose chemotherapy alone such as cytarabine; cytarabine and etoposide (Etopophos, Toposar); high-dose chemotherapy (including thiotepa) with *autologous stem cell transplant* (patient's own cells are infused after high-dose chemotherapy); or whole brain radiation therapy (WBRT). WBRT is seldom used in patients over the age of 65 because of permanent and progressive neurological complications (for example, loss of memory, problems with balance or muscle coordination).

If lymphoma cells are found in the spinal fluid, chemotherapy may be delivered directly into the spinal fluid, in addition to the above chemoimmunotherapy regimens.

TREATMENTS UNDER INVESTIGATION

Various agents are being investigated in clinical trials for patients with newly diagnosed CNS lymphoma and those who are *relapsed* (disease returns after treatment) or with refractory (disease does not respond to initial treatment) lymphoma. Some of the agents being investigated for the treatment of patients with CNSL include:

- Ibrutinib (Imbruvica)
- Lenalidomide (Revlimid)
- Nivolumab (Opdivo)
- Obinutuzumab (Gazyva)
- Pembrolizumab (Keytruda)
- Copanlisib (Aliqopa)
- Venetoclax (Venclexta)
- PQR309/Bimralisib
- Tisagenlecleucel (Kymriah)
- Axicabtagene ciloleucel (Yescarta)
- Acalabrutinib (Calaquence)
- Paxalisib
- Tiraburitinib

Treatment options are changing as new therapeutics become available and current treatments are improved. Because today's scientific research is continuously evolving, it is important that patients check with their physician or with the Lymphoma Research Foundation (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. Because CNS lymphoma is a rare disease, clinical trial enrollment is critical for establishing more effective, less toxic treatments. The rarity of the disease also means that the most novel treatments are often available only through clinical trials.

Patients interested in participating in a clinical trial should view the *Understanding Clinical Trials* fact sheet LRF's website at lymphoma.org/publications, and the *Clinical Trials Search Request Form* at 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 CNS 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, MRI [magnetic resonance imaging] scans of the brain and/or spine) and eye (ophthalmological) examination may be required at various times during *remission* (disappearance of signs and symptoms) to evaluate the need for additional treatment.

Some treatments can cause long-term side effects or late side effects, such as memory problems, which can vary based on the 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 side 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 side effects resulting from treatment or potential disease recurrences. LRF's award-winning *Focus On Lymphoma* mobile app (lymphoma.org/mobileapp) and the *Lymphoma Care Plan* (lymphoma.org/publications) can help patients manage this documentation.

PATIENT AND CAREGIVER SUPPORT SERVICES

A lymphoma diagnosis often triggers a range of feelings and concerns. In addition, cancer treatment can cause physical discomfort. One-to-one peer support programs, such as LRF's *Lymphoma Support Network*, connect patients and caregivers with volunteers who have experience with CNS lymphoma, similar treatments, or challenges, for mutual emotional support and encouragement. Patients and loved ones may find this useful whether the patient is newly diagnosed, in treatment, or in remission.

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 webinars for people with lymphoma and CNS lymphoma, as well as patient guides and e-Updates that provide the latest disease-specific news and treatment options. To learn more about any of these resources, visit our websites at lymphoma.org/CNS or lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.

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Supported through grants from:



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The *Understanding Lymphoma* series is published by the Lymphoma Research Foundation (LRF) for the purpose of informing and educating readers. Facts and statistics were obtained using published information, including data from the Surveillance, Epidemiology, and End Results (SEER) Program. Because each person's body and response to treatment is different, no individual should self-diagnose or embark upon any course of medical treatment without first consulting with his or her physician. The medical reviewer, the medical reviewer's institution, and LRF are not responsible for the medical care or treatment of any individual.

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Last updated 2021

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