

Understanding Lymphoma

CNS Lymphoma

Primary central nervous system (CNS) lymphoma is an aggressive form of non-Hodgkin lymphoma (NHL) in which *malignant* (cancer) cells are found exclusively in the CNS (the brain and spinal cord). Primary CNS lymphoma can start in the brain, spinal cord, meninges (sheets of tissue that protect the CNS) or the eye. When the lymphoma originated in other parts of the body and subsequently spread to the CNS, it is referred to as secondary CNS lymphoma. More than 95% of cases of primary CNS lymphoma are of B-cell origin.

The cause of primary CNS lymphoma is unknown, but having a compromised immune system (for example, people with acquired immunodeficiency syndrome [AIDS] or patients who have undergone organ transplant) may increase the risk of developing the disease. Having one or more of these risk factors does not mean a person will develop primary CNS lymphoma. Most people diagnosed with primary CNS lymphoma 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. Upon diagnosis, tests are done to find out which parts of the CNS are involved as well as to determine if there is cancer (lymphoma) elsewhere in the body. These can include computed tomography (CT) or positron emission tomography (PET) scans of the body, magnetic resonance imaging (MRI) of the brain and spinal cord, lumbar puncture, ophthalmologic evaluation, and bone marrow biopsy. Primary CNS lymphoma usually does not spread beyond the CNS or the eye, but it often relapses (disease returns after treatment).



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 regimens or consolidation therapies, to reduce the risk of recurrence. These consolidation therapies include high-dose chemotherapy alone such as cytarabine (Cytosar); cytarabine (Cytosar) 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).



TREATMENTS UNDER INVESTIGATION

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

- Abemaciclib (LY2835219)
- Ibrutinib (Imbruvica)
- Lenalidomide (Revlimid)
- Nivolumab (Opdivo)
- Obinutuzumab (Gazyva)
- Pembrolizumab (Keytruda)
- Copanlisib (Aliqopa)
- Venetoclax (Venclexta)
- Acalabrutinib (Calquence)
- Orelabrutinib (ICP-022)
- Zanubrutinib (Brukinza)

- Paxalisib (GDC-0084)
- Tirabrutinib (ONO-4059)
- Lisocabtagene maraleucel (Breyanzi)

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 on LRF's website (visit 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 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 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.

LRF'S HELPLINE AND LYMPHOMA SUPPORT NETWORK

A lymphoma diagnosis often triggers a range of feelings and concerns. In addition, cancer treatment can cause physical discomfort. The LRF Helpline staff members are available to answer your general questions about a lymphoma diagnosis and treatment information, as well as provide individual support and referrals to you and your loved ones. Callers may request the services of a language interpreter. A part of the Helpline is LRF's one-to-one peer support programs, Lymphoma Support Network. This program connects patients and caregivers with volunteers who have experience with primary 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.

MOBILE APP

Focus On Lymphoma is the first mobile application (app) that provides patients and caregivers comprehensive content based on their lymphoma subtype, including primary CNS lymphoma, and tools to help manage their lymphoma such as, keep track of medications and blood work, track symptoms, and document treatment side effects. The *Focus On Lymphoma* mobile app is available for download for iOS and Android devices in the Apple App Store and Google Play. For additional information on the mobile app, visit FocusOnLymphoma.org. To learn more about any of these resources, visit our website at lymphoma.org, or contact the LRF Helpline at **(800) 500-9976** or helpline@lymphoma.org.

Resources

LRF offers a wide range of free resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma and primary CNS lymphoma. LRF also provides many educational activities, including our in-person meetings, and webinars for people with lymphoma. For more information about any of these resources, visit our websites at lymphoma.org/aboutlymphoma/nhl/cns/ or lymphoma.org, or contact the LRF Helpline at **(800) 500-9976** or helpline@lymphoma.org.

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