

Getting the Facts: Hodgkin Lymphoma

Hodgkin lymphoma (HL), also known as Hodgkin disease, represents about 10 percent of all lymphomas in the United States.

It is estimated that 8,540 new cases of HL will be diagnosed in the United States in 2022. HL can occur in both children and adults, but it is most common in young adults between the ages of 20 and 29 years, with an average age of 39 years at diagnosis.

HL is often characterized by the presence of very large cells called Reed-Sternberg (RS) cells and usually starts in the lymph nodes; however, it can spread to other lymph nodes and, rarely, to other organs.

Common signs and symptoms of HL include swelling of the lymph nodes (which is usually painless), fever, night sweats, unexplained weight loss, itching, and lack of energy. While most people who have these complaints do not have HL, anyone with persistent symptoms should see a physician to make sure that lymphoma is not present.

COMMON TYPES OF HL

HL is divided into two main classifications: classical HL (cHL), which accounts for the majority of cases, and nodular lymphocyte-predominant HL. The type of HL a patient has may affect their treatment choices.

CLASSICAL HL

Nodular Sclerosis cHL is the most common subtype of HL in developed countries, accounting for about 70% of all cHL cases. In nodular (knot-like) sclerosis cHL, the involved lymph nodes contain RS cells mixed with normal white blood cells. The lymph nodes often contain a lot of scar tissue, which is where the name nodular sclerosis (scarring) originates. The disease is more common in women than in men, and it usually affects adolescents and adults under the age of 50 years. The majority of patients are cured with current treatments.

Mixed Cellularity cHL accounts for about 40% of all HL cases. The disease is more common in men than in women, and primarily affects older adults and people with human immunodeficiency virus (HIV) infection. With this type of cHL, the lymph nodes contain many RS cells in addition to several other cell types. More advanced disease is usually present by the time this subtype is diagnosed.

Lymphocyte-Rich cHL accounts for less than 5% of HL cases. The disease is characterized by the presence of numerous normal-appearing lymphocytes and classic RS cells. This subtype of HL is usually diagnosed at an early stage in older adults and has a low risk of relapse (disease return after treatment).

Lymphocyte-Depleted cHL is rarely diagnosed (about 1% of all cHL cases) and occurs mainly in older people and those with HIV infection. Abundant RS cells and few normal lymphocytes are present in the lymph nodes of patients with this subtype, which is aggressive and usually not diagnosed until it is widespread throughout the body.

LYMPHOCYTE-PREDOMINANT HL

Nodular Lymphocyte-Predominant HL (NLPHL) accounts for 5% of all HL cases. It affects men more often than women and is usually diagnosed before the age of 35 years. This subtype is characterized by the appearance of large *lymphocytic* and *histiocytic* cells (sometimes called popcorn cells), as well as small B cells. This form of HL is not very aggressive (grows slowly) and can *relapse* (disease comes back) many years later.



TREATMENT OPTIONS

The majority of patients with newly diagnosed HL can be cured. While the treatment depends on the type of HL and the patient's overall health, most patients treated for HL receive some form of chemotherapy, sometimes followed by radiation therapy as their first treatment. Standard *frontline* (initial) chemotherapy for stage I or II HL is ABVD (adriamycin, bleomycin, vinblastine, and dacarbazine) with or without radiation therapy or other agents. The ABVE-PC (adriamycin, bleomycin, vinblastine, etoposide, prednisone and cyclophosphamide) chemotherapy regimen is generally standard for higher risk pediatric patients. Other chemotherapy regimens (like BEACOPP, which includes bleomycin, etoposide, doxorubicin, cyclophosphamide, vincristine, procarbazine, and prednisone) may be suggested for patients with more advanced-stage disease. The antibody-drug conjugate brentuximab vedotin (Adcetris) can also be used as frontline therapy for stage III or IV cHL in combination with doxorubicin, vinblastine, and dacarbazine (A+AVD).

Patients with early stage NLPHL may be treated with radiation therapy or surgery alone. If B symptoms (fever without infection, drenching night sweats and unexplained weight loss) are present or the disease is more advanced, treatment options include chemotherapy with or without rituximab (Rituxan) followed by radiation therapy. Some patients without B symptoms might be given rituximab (Rituxan) alone.

A vast number of single-agent and combination treatments are also available for patients with relapsed (disease returns after treatment) or refractory (disease does not respond to treatment) HL, including stem cell transplantation, brentuximab vedotin (Adcetris), nivolumab (Opdivo), pembrolizumab (Keytruda), and chemotherapy regimens.

For more information on relapsed and refractory HL, view the *Hodgkin Lymphoma: Relapsed/Refractory* fact sheet on the Lymphoma Research Foundation's (LRF's) website (lymphoma.org/publications).



TREATMENTS UNDER INVESTIGATION

The overall 5-year survival rate of HL is 88.3% and can reach up to 94% when detected in earlier stages. Although the cure rate in HL is already high, research continues in order to develop therapies that have less side effects and are even more effective. New drugs under study include immune checkpoint inhibitors like avelumab, camrelizumab, sintilimab and tislelizumab.

Investigators are also looking for ways to treat the minority of patients who are refractory to treatment and those who relapse. Studies for relapsed/refractory HL are looking at new combinations of targeted therapies or immunotherapies including umbralisib (Ukoniq), ibrutinib (Imbruvica), lenalidomide (Revlimid), and other drugs like antibody-drug conjugates (camidanlumab tesirine). Clinical trials are also ongoing for a modified interleukin-2 (THOR-707), regenerative medicine advanced therapy (AB-205) and chimeric antigen receptor (CAR) T cell therapy directed against CD-30.



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 (lymphoma.org/publications), 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 computed tomography [CT]/positron emission tomography [PET] scans) may be required at various times during remission to evaluate the need for additional treatment.

Since HL is a highly curable malignancy mostly affecting young people with long life expectancy, there is a growing number of HL survivors who may have special medical needs such as screening for secondary cancers or monitoring for long-term toxicities of therapy. Long-term follow up in a survivorship clinic is a way to quickly identify and address these issues. Additional resources for adolescents and young adults living with cancer are available on LRF's website (lymphoma.org/publications).

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 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 HL, and 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 HL, 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.

LYMPHOMA CARE PLAN

Keeping your information in one location can help you feel more organized and in control. This also makes it easier to find information pertaining to your care and saves valuable time. LRF offers a Lymphoma Care Plan as a resource for patients and their caregivers. LRF's Lymphoma Care Plan document organizes information on your health care team, treatment regimen, and follow-up care. You can also keep track of health screenings and any symptoms you experience to discuss with your health care provider during future appointments. The Lymphoma Care Plan document can be accessed by visiting lymphoma.org/publications.

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 HL. 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/HL or lymphoma.org, or contact the LRF Helpline at **(800) 500-9976** or helpline@lymphoma.org.

Para información en Español, por favor visite lymphoma.org/es. (For Information in Spanish please visit lymphoma.org/es.)

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