

## Hodgkin 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).

HL, also known as Hodgkin disease, is not as common as NHL. Approximately 9,000 new cases of HL are projected each year. Although HL can occur in both children and adults, it is most commonly diagnosed in young adults between the ages of 20 and 34 years.

HL is characterized by the presence of very large cells called Reed-Sternberg (RS) cells, although other abnormal cell types may be present. HL usually starts in the lymph nodes; however, it often spreads from one lymph node to another and can also spread to other organs.

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

### Common Types of HL

HL has been divided into two main classifications: classical HL (CHL), which accounts for 90 to 95 percent 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, accounting for 60 to 80 percent of all HL 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. The majority of patients are cured with current treatments.

**Mixed Cellularity CHL** accounts for about 15 to 30 percent of all HL cases. The disease is found more commonly in men than in women, and it primarily affects older adults. 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-Depleted CHL** is rarely diagnosed. 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-Rich CHL** accounts for less than five percent of HL cases. The disease may be *diffuse* (spread out) or nodular in form and 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 adults and has a low *relapse* (disease returns after treatment) rate.

### Lymphocyte Predominant HL

**Nodular Lymphocyte Predominant HL** accounts for five to 10 percent of all HL cases. It affects men more often than women and is usually diagnosed before the age of 35. In nodular lymphocyte predominant HL, most of the lymphocytes found in the lymph nodes are normal (not cancerous). Typical RS cells are usually not found in this subtype, but large, abnormal B cells (sometimes referred to as popcorn cells) can be seen as well as small B cells, which may be distributed in a nodular pattern within the tissues. This subtype is usually diagnosed at an early stage and is not very aggressive. In many ways, this form of HL resembles *indolent* (slow-growing) B-cell NHL with late recurrences.

### Treatment Options

Over 80 percent of patients with HL survive for five years, and many are cured. Most patients treated for HL will receive some form of chemotherapy, and sometimes radiation therapy, as their first treatment. The recommended *front-line* (initial) therapy for HL is ABVD (adriamycin, bleomycin, vinblastine, and dacarbazine) with or without radiation therapy or other agents, depending on the patient's type and stage of HL as well as their overall health status. Other chemotherapy regimens may also be recommended by the physician, but not always as initial therapies. ABVD tends to be recommended for stage I or II favorable disease, but it may also be used to treat more advanced HL. The Stanford V (spoken as Stanford Five) regimen, which consists of doxorubicin, vinblastine, mechlorethamine, etoposide, vincristine, bleomycin, and prednisone, is typically used for patients with stage I or II *bulky disease* (patient has tumor greater than 6–10 centimeters) or more advanced disease. BEACOPP, which includes bleomycin, etoposide, doxorubicin, cyclophosphamide, vincristine, procarbazine, and prednisone, may also be suggested for patients with more advanced stage disease.

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Stem cell transplantation is typically used in the *relapsed* (disease returns after treatment) or *refractory* (disease does not respond to treatment) setting. Brentuximab vedotin (Adcetris) was approved in 2011 by the U.S. Food and Drug Administration (FDA) for the treatment of relapsed/refractory HL after failure of stem cell transplantation or after failure of two previous chemotherapy regimens in patients who are not eligible for stem cell transplantation. In 2015 brentuximab vedotin was FDA-approved as a consolidation treatment after autologous stem cell transplantation in patients with HL who are at high risk of disease relapse or progression. In May 2016, nivolumab (Opdivo) was approved by the FDA for the treatment of patients with CHL that has relapsed or progressed after autologous stem cell transplantation and post-transplantation brentuximab vedotin. 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 at [www.lymphoma.org](http://www.lymphoma.org).

## Treatments Under Investigation

Although the cure rate in HL is already high, research continues to look for ways to treat the minority of patients who are *refractory* to treatment and those who relapse. Many promising therapies are currently under investigation in clinical trials for HL including:

- Bendamustine (Treanda)
- Ifosfamide (Ifex)
- Panobinostat (Farydak)
- Gemcitabine (Gemzar)
- Mocetinostat (MGCD0103)
- Pembrolizumab (Keytruda)

## 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](http://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](mailto: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 positron emission tomography [PET]/computed tomography [CT] scans) may be required at various times during remission to evaluate the need for additional treatment.

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 concerns. One-to-one peer support programs, such as LRF's Lymphoma Support Network, connects patients and caregivers with volunteers who have experience with HL, similar treatments, or challenges, for mutual emotional support and encouragement. You may find this useful whether you or a loved one 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, *Focus On Lymphoma*. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as disease-specific websites, videos, and e-Updates for current lymphoma information and treatment options. To learn more about any of these resources, visit our websites at [www.FocusOnHL.org](http://www.FocusOnHL.org) or [www.lymphoma.org](http://www.lymphoma.org), or contact the LRF Helpline at (800) 500-9976 or [helpline@lymphoma.org](mailto:helpline@lymphoma.org).