

Hodgkin Lymphoma: Relapsed/Refractory

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. 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 has been studied more than any other type of lymphoma, and because of the treatment advances over the past few decades, five-year survival rates have improved.

The presence of Reed-Sternberg cells is necessary in the diagnosis of HL, although other inflammatory cell types are present. HL usually starts in the lymph nodes; however, it often spreads from one lymph node to another and can also involve 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, 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

The two main classifications of Hodgkin lymphoma are classical HL (cHL), which accounts for almost all (95 percent) cases of HL, and nodular lymphocyte predominant HL.

The four subtypes of classical HL are:

- Nodular sclerosis
- Mixed cellularity cHL
- Lymphocyte-depleted cHL
- Lymphocyte-rich cHL

The type of Hodgkin lymphoma patients have may affect their treatment choices.

Relapsed or Refractory

For patients who *relapse* (disease returns after treatment) or become *refractory* (disease does not respond to treatment), secondary therapies are often successful in providing another *remission* (disappearance of signs and symptoms) and may even cure the disease. For cHL, most relapses typically occur within the first three years following diagnosis, although some relapses occur much later.

Treatment Options

A number of treatment options are available for patients with relapsed or refractory HL. Exactly what type of treatment is prescribed for individual patients depends on several factors, including the timing of the relapse, age and overall health of the patient, the scope of disease, and the previous therapies received.

The standard secondary treatment for the majority of patients consists of *systemic* (throughout the body) therapy, usually followed by *autologous stem cell transplantation* (in which a patient's own stem cells are infused after high-dose chemotherapy). Involved site radiation therapy (ISRT) may also be used. For more information on transplantation, view the *Understanding the Stem Cell Transplantation Process* publication on the Lymphoma Research Foundation's (LRF's) website at lymphoma.org/publications.

There are a variety of single-agent and combination therapy regimens that may be used for relapsed/refractory HL, including:

- Brentuximab vedotin (Adcetris)
- Bendamustine (Treanda)
- Nivolumab (Opdivo)
- Pembrolizumab (Keytruda)
- DHAP (dexamethasone, cisplatin, and cytarabine)
- ESHAP (etoposide, methylprednisolone, cisplatin, and cytarabine)
- GVD (gemcitabine, vinorelbine, and liposomal doxorubicin)
- ICE (ifosfamide, carboplatin, and etoposide)
- IGEV (ifosfamide, gemcitabine, and vinorelbine)

Treatments Under Investigation

In addition to conventional chemotherapies, there are several new agents currently being tested in clinical trials:

- Anti-CD30-CAR T cells
- Atezolizumab (Tecentriq)
- Bortezomib (Velcade)
- Carfilzomib (Kyprolis)
- Everolimus (Afinitor)
- Ibrutinib (Imbruvica)
- Lenalidomide (Revlimid)
- Mocetinostat (MGCD103)
- Ruxolitinib (Jakafi)
- Umbralisib

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. Because the optimal initial treatment of HL is not clear, clinical trials are very important and will identify the best treatment options in this disease. Patients interested in participating in a clinical trial should view the *Understanding Clinical Trials* fact sheet on LRF's website at 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 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. LRF's award-winning mobile app (lymphoma.org/mobileapp) and *Lymphoma Care Plan* (lymphoma.org/publications) can help patients manage this information.

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 Hodgkin lymphoma, similar treatments, or challenges, for mutual emotional support and encouragement. Patients and loved ones may find this information 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 Hodgkin lymphoma, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts for people with lymphoma, as well as an *Understanding Hodgkin Lymphoma* patient guide and HL 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/HL or lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.

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