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About Non-Hodgkin Lymphoma in Children

Overview and Types

If your child has just been diagnosed with non-Hodgkin lymphoma or you are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- [What Is Non-Hodgkin Lymphoma in Children?](#)
- [What Are the Differences Between Cancers in Adults and Children?](#)
- [Types of Non-Hodgkin Lymphoma in Children](#)

Research and Statistics

See the latest estimates for new cases of childhood non-Hodgkin lymphoma in the US and what research is currently being done.

- [Key Statistics for Non-Hodgkin Lymphoma in Children](#)
 - [What's New in Research and Treatment of Non-Hodgkin Lymphoma in Children?](#)
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What Are the Differences Between Cancers in Adults and Children?

The types of cancers that develop in children are often different from the types that develop in adults. Childhood cancers are often the result of DNA changes in cells that take place very early in life, sometimes even before birth. Unlike many cancers in adults, childhood cancers are not strongly linked to lifestyle or environmental risk factors.

There are exceptions, but childhood cancers tend to respond better to treatments such as chemotherapy. Children's bodies also tend to tolerate chemotherapy better than adults' bodies do. But cancer treatments such as chemotherapy and radiation therapy can have long-term side effects, so children who survive cancer need careful attention for the rest of their lives.

Since the 1960s, most children and teens with cancer have been treated at [specialized centers](#)¹ designed for them. Being treated in these centers offers the advantage of a team of specialists who know the differences between adult and childhood cancers, as well as the unique needs of children with cancer and their families. This team usually includes pediatric oncologists, surgeons, radiation oncologists, pathologists, pediatric oncology nurses, and nurse practitioners.

These centers also have psychologists, social workers, child life specialists, nutritionists, rehabilitation and physical therapists, and educators who can support and educate the entire family.

Most children with cancer in the United States are treated at a center that is a member of the Children's Oncology Group (COG). All of these centers are associated with a university or children's hospital. As we have learned more about treating childhood cancer, it has become even more important that treatment be given by experts in this area.

Any time a child or teen is diagnosed with cancer, it affects every family member and nearly every aspect of the family's life. You can read more about coping with these changes in [When Your Child Has Cancer](#)².

Hyperlinks

1. www.cancer.org/treatment/finding-and-paying-for-treatment/choosing-your-treatment-team/pediatric-cancer-centers.html
2. www.cancer.org/treatment/children-and-cancer/when-your-child-has-cancer.html

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What Is Non-Hodgkin Lymphoma in Children?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see [What Is Cancer?](#)¹

Lymphoma is a type of cancer that starts in cells called *lymphocytes*, which are part of the body's immune system.

Types of lymphoma

The 2 main kinds of lymphomas are:

- **Hodgkin lymphoma** (also known as **Hodgkin disease**), which is named after Dr. Thomas Hodgkin, who first described it
- **Non-Hodgkin lymphoma (NHL)**

These types of lymphomas differ in how they behave, spread, and respond to treatment, so knowing which type your child has is important.

Both of these types are more common in adults, but they can also occur in children and teens: NHL tends to occur in younger children, while Hodgkin lymphoma is more likely to affect older children and teens.

Hodgkin lymphoma is very similar in adults and children, and treatment is the same for both. For more information on this disease, see [Hodgkin Lymphoma](#)².

The lymph (lymphatic) system

The lymph system is part of the body's immune system, which helps fight infections and some other diseases. It also helps fluids move around in the body.

Lymphocytes

The lymph system is made up mainly of cells called *lymphocytes*, a type of white blood cell. The main types of lymphocytes are:

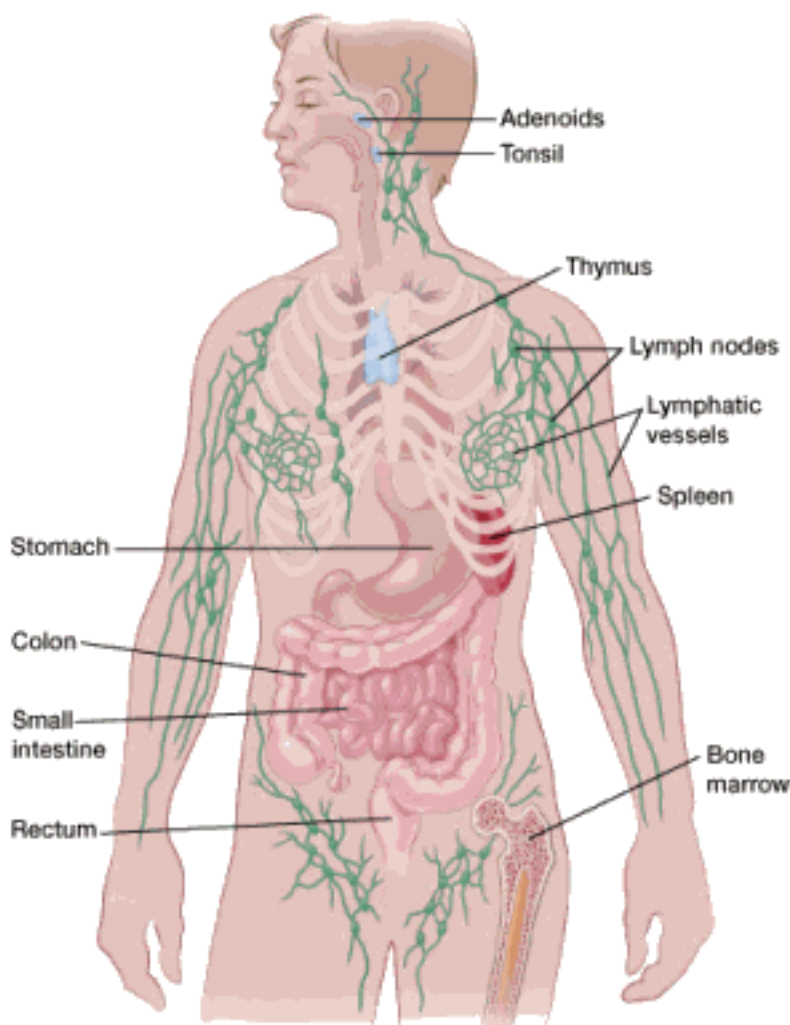
- **B lymphocytes (B cells):** B cells normally help protect the body against germs (bacteria or viruses) by making proteins called *antibodies*. The antibodies attach to the germs, marking them for destruction by other parts of the immune system.
- **T lymphocytes (T cells):** There are several types of T cells, each with a special job. Some T cells destroy germs or abnormal cells in the body. Other T cells help boost or slow the activity of other immune system cells.

Both types of lymphocytes can develop into lymphoma cells.

[Different types of NHL](#) can develop in children. Treatment depends on which type of NHL it is, so determining the exact type a child has is important.

Parts of the lymph system

The lymph system is in many parts of the body, so lymphomas can start almost anywhere. (This can affect what [symptoms](#)³ a child has.)



Lymph tissue is found in:

Lymph nodes: Lymph nodes are bean-sized collections of lymphocytes and other immune cells throughout the body. They can sometimes be felt under the skin in the neck, under the arms, and in the groin. Lymph nodes are connected to each other by a system of lymphatic vessels.

Lymph nodes get bigger when they fight infection. Lymph nodes that grow because of infection are called *reactive nodes* or *hyperplastic nodes* and are often painful when they are touched. An enlarged lymph node in a child is not usually a sign of a serious problem. Lymph nodes in the neck are often enlarged in children with sore throats or colds. But a large lymph node is also the most common sign of lymphoma. This is discussed more in [Signs and Symptoms of Non-Hodgkin Lymphoma in Children⁴](#).

Spleen: The spleen is an organ under the lower ribs on the left side of the body. The spleen makes lymphocytes and other immune system cells. It also stores healthy blood cells and filters out damaged blood cells, bacteria, and cell waste.

Bone marrow: The bone marrow is the spongy tissue inside certain bones, which is where new blood cells (including some lymphocytes) are made.

Thymus: The thymus is a small organ behind the upper part of the breast bone and in front of the heart. It's important in the development of T lymphocytes.

Adenoids and tonsils: These are collections of lymph tissue in the back of the throat. They help make antibodies against germs that are breathed in or swallowed. They are easy to see when they become enlarged during an infection, which occurs often in children, or if a lymphoma develops.

Digestive tract: Lymph tissue is also in the stomach and intestines, as well as many other organs.

Hyperlinks

1. www.cancer.org/cancer/cancer-basics/what-is-cancer.html
2. www.cancer.org/cancer/hodgkin-lymphoma.html
3. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/detection-diagnosis-staging/signs-symptoms.html
4. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/detection-diagnosis-staging/signs-symptoms.html

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Types of Non-Hodgkin Lymphoma in Children

Non-Hodgkin lymphomas (NHLs) are most often classified by how the cancer cells look under the microscope. Key features include the size and shape of the cells and how they are arranged (their pattern of growth).

- **Size** is described as large or small.
- **Shape** is described as cleaved (showing folds or indentations) or non-cleaved.
- The **growth pattern** may be either diffuse (cancer cells are scattered) or follicular (cells are arranged in clusters).

Not every lymphoma is described using all 3 features. Special lab tests are often needed to accurately classify lymphomas. These are discussed in [Tests for Non-Hodgkin Lymphoma in Children](#)¹.

The most common types of NHL in children are different from those in adults. Nearly all NHLs in children are 1 of 3 main types:

- Lymphoblastic lymphoma
- Burkitt lymphoma (small non-cleaved cell lymphoma)
- Large cell lymphoma

All 3 types are high grade (meaning they grow quickly) and diffuse, but it's important to find out which type a child has because they are treated differently.

There are many other types of NHL. These are much more common in adults and are rare in children, so they are not discussed further heret.

Lymphoblastic lymphoma

Lymphoblastic lymphoma (LBL) accounts for about 25% to 30% of NHL in children in the United States. Boys are about twice as likely to get LBL as girls.

The cancer cells of LBL are very young lymphocytes called *lymphoblasts*. They are the same cells as those seen in [acute lymphoblastic leukemia \(ALL\)](#)² in children. In fact, if more than 25% of the bone marrow is made up of lymphoblasts, the disease is classified and treated as ALL instead of lymphoma.

Most cases of LBL develop from **T cells** and are called **precursor T-lymphoblastic lymphomas**. These lymphomas often start in the thymus, forming a mass in the area behind the breast bone and in front of the trachea (windpipe). This can cause problems breathing, which may be the first symptom of LBL.

Less often, LBL develops in the tonsils, lymph nodes of the neck, or other lymph nodes. It can spread very quickly to the bone marrow, other lymph nodes, the surface of the brain, and/or the membranes that surround the lungs and heart.

A small fraction of LBLs develop from B cells, and are called **precursor B-lymphoblastic lymphomas**. These lymphomas more often begin in lymph nodes outside the chest, particularly in the neck. They can also involve the skin and bones.

LBL can grow very quickly and can often cause trouble breathing, so it needs to be diagnosed and treated quickly.

Burkitt lymphoma

Burkitt lymphoma, also known as *small non-cleaved cell lymphoma*, accounts for about 40% of childhood NHL in the United States. It is most often seen in boys, usually when they are around 5 to 10 years old.

A subtype of Burkitt lymphoma, sometimes called *Burkitt-like lymphoma* or *non-Burkitt lymphoma*, shares some features with diffuse large B-cell lymphoma (described below) when seen under the microscope, but it is still treated like Burkitt lymphoma.

Burkitt lymphoma is named after the doctor who first described it in African children. In certain parts of Africa, Burkitt lymphoma accounts for nearly all childhood NHL and over half of all childhood cancers. In African children this lymphoma usually develops in the jaw or other facial bones.

Burkitt lymphomas in other parts of the world, including the United States, most often start in the abdomen (belly). Typically, a child will develop a large tumor in his or her abdomen that can sometimes block the bowels (intestines). This can cause belly pain, nausea, and vomiting. Burkitt lymphoma can also sometimes start in the neck or tonsils, or rarely in other parts of the body.

This lymphoma develops from **B lymphocytes (B cells)**. It can spread to other organs, including the surface of the brain or inside the brain. It is one of the fastest growing cancers known, so it needs to be diagnosed and treated quickly.

Large cell lymphomas

These lymphomas start in more mature forms of T cells or B cells and can grow almost anywhere in the body. They are not as likely to spread to the bone marrow or brain, nor do they grow as quickly as other childhood lymphomas. These lymphomas tend to occur more often in older children and teens.

The 2 main subtypes of large cell lymphoma are:

Anaplastic large cell lymphoma (ALCL): This lymphoma makes up about 10% of all NHL in children. It usually develops from mature T cells. It may start in lymph nodes in the neck or other areas, and may be found in the skin, lungs, bone, digestive tract, or other organs.

Diffuse large B-cell lymphoma (DLBCL): This lymphoma accounts for about 15% of childhood lymphomas. It starts in B cells, as the name implies. These lymphomas sometimes grow as large masses in the mediastinum (the space between the lungs), in which case they are referred to as **primary mediastinal B-cell lymphomas**. But they are also sometimes found in other areas such as lymph tissue in the neck or abdomen, or in the bones.

Hyperlinks

1. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/detection-diagnosis-staging/how-diagnosed.html
2. www.cancer.org/cancer/leukemia-in-children.html

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Allen CE, Kamdar KY, Bollard CM, Gross TG. Malignant non-Hodgkin lymphomas in children. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia Pa: Lippincott Williams & Wilkins; 2016:587–603.

Kamdar KY, Sandlund JT, Bollard CM. Malignant lymphomas in childhood. In: Hoffman R, Benz EJ, Silberstein LE, Heslop HE, Weitz JI, Anastasi J, eds. *Hematology: Basic Principles and Practice*. 6th ed. Philadelphia, Pa: Elsevier; 2013:1255-1266.

Rabin KR, Margolin JF, Kamdar KY, Poplack DG. Leukemias and lymphomas of childhood. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology*. 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015:1500–1510.

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Key Statistics for Non-Hodgkin Lymphoma in Children

Non-Hodgkin lymphoma (NHL) accounts for about 5% of all childhood cancers. [Hodgkin lymphoma](#)¹ accounts for about another 3%.

In children up to age 14, most lymphomas are non-Hodgkin lymphomas, with about 500 of these cancers being diagnosed in the United States each year. If all children and teens up to age 19 are included, the numbers of Hodgkin and non-Hodgkin lymphomas are about equal, and there are about 800 cases of NHL diagnosed each year.

NHL is about 2 to 3 times more common in boys than in girls, and it is more common in white children than black children.

About 2% of all NHLs occur in children and teens.

Overall, the risk of NHL in children increases with age. It can occur at any age but is uncommon in children younger than 3.

Statistics on survival can be found in [Survival Rates for Childhood Non-Hodgkin Lymphoma](#)².

Hyperlinks

1. www.cancer.org/cancer/hodgkin-lymphoma.html
2. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/detection-diagnosis-staging/survival-rates.html

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Allen CE, Kamdar KY, Bollard CM, Gross TG. Malignant non-Hodgkin lymphomas in children. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia Pa: Lippincott Williams & Wilkins; 2016:587–603.

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What's New in Research and Treatment of Non-Hodgkin Lymphoma in Children?

Research on the causes, diagnosis, and treatment of childhood non-Hodgkin lymphoma (NHL) is being done at many medical centers, university hospitals, and other institutions around the world.

Genetics

As noted in [What Causes Non-Hodgkin Lymphoma in Children?](#)¹ scientists are making great progress in understanding how changes in the DNA inside normal lymphocytes can cause them to develop into lymphoma cells.

Understanding the gene changes that often occur in lymphoma cells can help explain why these cells grow too quickly, live too long, and don't develop into normal, mature cells. This information is being used to develop new treatments for lymphoma.

This progress has also led to very sensitive lab tests for detecting and monitoring this disease during treatment. Tests such as the polymerase chain reaction (PCR) can identify lymphoma cells based on some of these gene changes. This test is useful in determining how completely the lymphoma has been destroyed by treatment, and whether a relapse is likely if no further treatment is given.

Clinical trials of new treatments

Most children with NHL are treated at major medical centers, where treatment is often given as part of a [clinical trial](#)² to get the most up-to-date care. Several important questions are now being studied in clinical trials, such as:

- Can early-[stage](#)³ (stages I and II) lymphomas be treated with less intense [chemotherapy](#)⁴ regimens?
- What is the best length of treatment for each type of NHL?
- Can less intense treatments provide an outcome as good as highly intense treatments, while possibly helping children avoid some [long-term side effects](#)⁵?
- Can new chemotherapy drugs and new combinations of drugs improve cure rates?
- Can the safety and effectiveness of [stem cell transplants](#)⁶ be improved on?
- Can newer, targeted drugs such as monoclonal antibodies be helpful in treating NHL, either alone or added to current treatments to make them better? (See [Other Drugs for Non-Hodgkin Lymphoma in Children](#)⁷.)
- Can newer forms of [immunotherapy](#)⁸ (treatments that boost the immune system), such as CAR T-cell therapy, be helpful in treating childhood NHL, especially if other treatments are no longer working?

Hyperlinks

1. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/causes-risks-prevention/what-causes.html
2. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
3. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/detection-diagnosis-staging/staging.html
4. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/treating/chemotherapy.html
5. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/after-treatment/long-term-effects.html
6. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/treating/bone-marrow-

- [stem-cell-transplant.html](#)
7. www.cancer.org/cancer/childhood-non-hodgkin-lymphoma/treating/monoclonal-antibodies.html
 8. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/immunotherapy.html

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