

Hodgkin Lymphoma

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A HANDBOOK FOR FAMILIES

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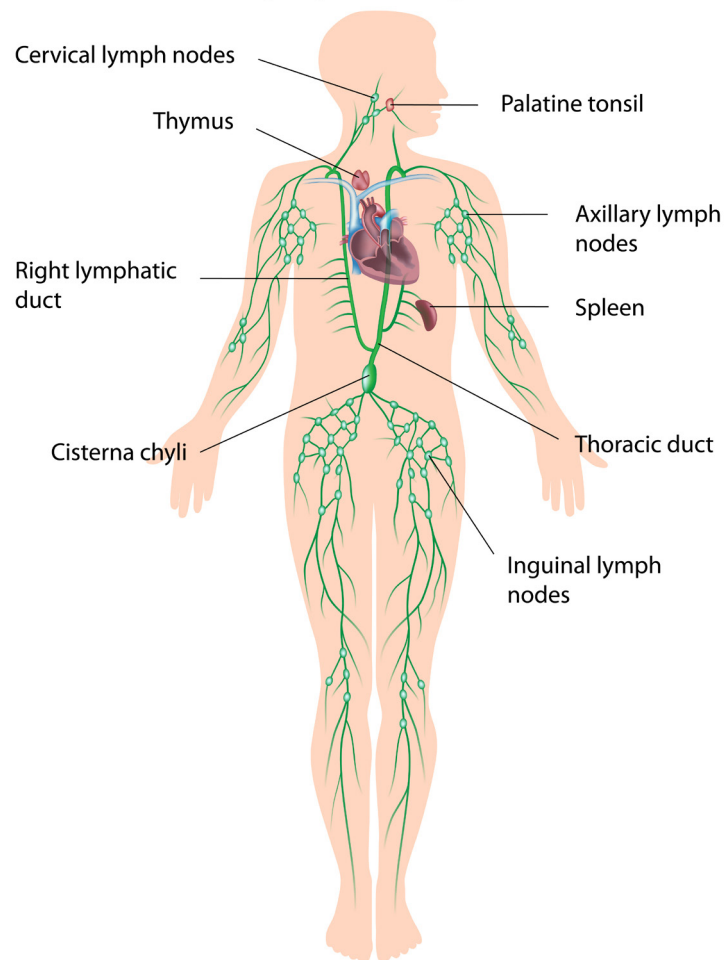
■ WHAT IS HODGKIN LYMPHOMA?

Hodgkin lymphoma, also known as Hodgkin's disease, is a cancer of the lymphatic system. The lymphatic system is part of the body's immune system, which protects the body from infection and disease. The lymphatic system is made up of a network of tissues and organs, including lymph nodes, tonsils, bone marrow, the spleen, and the thymus. Lymph tissue also is found in other parts of the body including the gut, skin, and brain. The lymphatic system circulates watery fluid called lymph throughout the body. This lymph fluid includes a type of white blood cells called lymphocytes that help the immune system filter viruses, bacteria, and other foreign substances out of the body.

Because lymph tissue is found throughout the body, Hodgkin lymphoma can develop almost anywhere and spread to almost any tissue or organ. Researchers have determined it is a cancer that develops from an abnormal lymphocyte (i.e., white blood cell) that divides too rapidly and grows without order or control.

The diagnosis of Hodgkin lymphoma depends on the presence of a particular kind of abnormal blood cells called Hodgkin Reed-Sternberg cells. These large, abnormal lymphocytes are seen when the involved tissue from a person with Hodgkin lymphoma is examined under a microscope.

The Lymphatic System



■ WHAT ARE SOME SYMPTOMS OF HODGKIN LYMPHOMA?

Most patients with Hodgkin lymphoma experience swollen lymph nodes that usually are painless, firm, and movable. These nodes most commonly are found in the neck and chest and less commonly under the arms or in the groin. Other symptoms of Hodgkin lymphoma include

- fever
- night sweats
- weight loss for no known reason
- fatigue
- difficulty breathing
- itchy skin.

■ WHAT CAUSES HODGKIN LYMPHOMA?

The causes of Hodgkin lymphoma are unknown. Individuals are at greater risk of developing Hodgkin lymphoma when they have the following:

- Epstein-Barr virus (EBV), which is the virus that causes infectious mononucleosis, often called “mono” (however, most people who have an EBV infection never develop Hodgkin lymphoma)
- human immunodeficiency virus (HIV)
- a family history of Hodgkin lymphoma (although hereditary cases are very rare).

■ WHO GETS HODGKIN LYMPHOMA?

Hodgkin lymphoma is a rare disease, accounting for about 5% of childhood cancers in the United States. It rarely occurs in patients younger than 5 years, but the risk of developing Hodgkin lymphoma increases in the second decade of life, making it one of the most common cancers in teenagers (usually between the ages of 15 and 19 years) and young adults.

■ IS HODGKIN LYMPHOMA INHERITED?

Though rare, some cases of Hodgkin lymphoma have been reported within families of certain ethnic groups. This leads to the premise that there may be some genetic predisposition that can increase the risk of developing Hodgkin lymphoma. There is a slightly greater chance that Hodgkin lymphoma will occur in siblings and cousins of those who have or have had the disease. Although the risk is about two to five times higher for siblings compared with nonsiblings, the likelihood that a sibling will get Hodgkin lymphoma is still very low.

■ WHAT IS METASTASIS?

Metastasis refers to the spread of a tumor from its original location (i.e., primary site) to other parts of the body. Hodgkin lymphoma often spreads from one lymph node to another and also can spread to organs outside the lymphatic system. If a biopsy reveals Hodgkin lymphoma, additional tests will be performed to find out if the cancer has spread to other parts of the body.

■ WHAT IS STAGING?

Staging is the process of determining the location and extent of a disease at the time of diagnosis. The information gathered from the staging process determines the extent of the involvement of the disease and helps guide treatment for that disease. There are four stages in Hodgkin lymphoma which are determined by the locations of the lymph node chains that have tumors. A higher stage number indicates that the disease has spread further throughout the body.

Hodgkin Lymphoma Staging

Within each stage, the letters "E" and "S" are used to further classify the disease. "E" indicates that cancer has extended into an organ or tissue that is not part of the lymphatic system but is next to an involved lymph node. "S" indicates that cancer is found in the spleen.

STAGE I

- Stage I: Cancer is found in one group of lymph nodes.
- Stage IE: Cancer is found in one group of lymph nodes and has extended to a nearby organ or tissue that is not part of the lymphatic system.

STAGE II

- Stage II: Cancer is found in two or more lymph node groups on the same side of the diaphragm (the breathing muscle that separates the chest and belly areas).
- Stage IIE: Cancer is found in two or more lymph-node groups on the same side of the diaphragm and has extended from one of those lymph nodes to a nearby organ or tissue that is not part of the lymphatic system.

STAGE III

- Stage III: Cancer is found on both sides of the diaphragm.
- Stage IIIE: Cancer is found on both sides of the diaphragm and has extended from one of these lymph nodes to a nearby organ or tissue that is not part of the lymphatic system.
- Stage IIIS: Cancer is found on both sides of the diaphragm and in the spleen.
- Stage IIIE+S: Cancer is found on both sides of the diaphragm and in the spleen and has extended from one of these lymph-node groups to a nearby organ or tissue that is not part of the lymphatic system.

STAGE IV

- Stage IV: Cancer is found throughout one or more organs that are not part of the lymphatic system (e.g., the lungs, bones, or bone marrow) and may be in lymph nodes that are near or far from those organs.

Each stage is further classified as either "A" or "B." "A" means the child has no specific symptoms. "B" indicates the child has experienced specific symptoms prior to diagnosis. These specific "B" symptoms include an unexplained fever that exceeds 100.4 °F/38.0 °C, drenching night sweats, or weight loss of at least 10% of the individual's body weight during a 6-month period.

Risk Groups

In addition to staging, Hodgkin lymphoma is divided into risk groups based on information obtained during staging, the size and location of the tumor, and whether the patient has “B” symptoms. Once the risk group is determined, the treatment team will use this information to decide which type of treatment is needed.

■ WHICH TESTS AND PROCEDURES WILL MY CHILD NEED?

Several tests are necessary to diagnose Hodgkin lymphoma and determine the extent of your child’s disease. The lymph nodes, chest, and abdomen will be examined. An enlarged spleen or liver may indicate Hodgkin lymphoma is present in those organs. The presence of small, soft lymph nodes in children may be misleading because only lymph nodes that have been increasing in size or that are significantly enlarged are a concern.

To determine the best treatment possible, it is very important to identify the exact type of Hodgkin lymphoma and where it is located in the body. Your child’s physician will decide which tests and procedures are needed.

Most Commonly Performed Tests and Procedures

TISSUE BIOPSY

The removal of all or part of a lymph node or other tissue is necessary to diagnose Hodgkin lymphoma. A lymph node tissue biopsy can be performed either in an operating room or in the hospital’s radiology department. Your child’s comfort and anxiety level always will be considered when planning procedures that involve needles.

Once the biopsy sample is obtained, a pathologist will examine the tissue under a microscope to look for cancer cells, specifically Hodgkin Reed-Sternberg cells, which are common abnormal lymphocytes in Hodgkin lymphoma.



X-RAY EXAMINATION

A chest X-ray, which takes a picture of the organs and bones inside the chest, may be necessary to determine whether Hodgkin lymphoma is present in the upper middle area of the chest (the mediastinum). This is the area just above the heart but below the neck, and it is a common site for Hodgkin lymphoma. A chest X-ray also can help determine if the lungs are affected.

CT SCAN

A computerized axial tomography (CT) scan is a computer-assisted X-ray that provides very detailed images of areas inside the body from different angles. The scan is painless and quick, but your child must be able to lie completely still during the scan. Younger children may require sedation to help them lie still. Your child may not be allowed to eat or drink for several hours before the exam. To make the CT images clearer, it also may be necessary for your child to have a small amount of dye injected into a vein or to drink a liquid containing a flavorless dye. Typically, side effects do not occur from either type of dye, although allergic reactions are possible. These scans are obtained at the time of diagnosis and at different times during treatment to determine how your child is responding to it.

PET SCAN

A positron emission tomography (PET) scan creates images of the body that show the size and shape of organs and tumors. PET scans are like CT and magnetic resonance imaging (MRI) scans, except they also can show chemical and functional changes within the body. A PET scan has three steps: injection of a radioactive material; a waiting period, usually 30–60 minutes; and scanning by the PET machine. During the waiting period and the scanning, your child must be able to lie completely still. The scanning process is painless; however, some children may require sedation to help them lie still. Like the CT scan, a PET scan should be obtained at diagnosis and is used to evaluate your child’s response as treatment progresses.

BONE MARROW BIOPSY AND ASPIRATION

Some children may need a bone marrow biopsy and aspiration, which is a procedure that involves collecting a sample of bone marrow. Bone marrow is found in the hollow part of the bone and is where white blood cells (infection fighters), red blood cells (oxygen carriers), and platelets (cells that help blood clot properly) are made. Once the bone marrow sample is collected, it is examined under a microscope by a pathologist to see whether Hodgkin lymphoma is present. Children usually require sedation or anesthesia during this procedure because it is performed with a biopsy needle.

Other Tests and Procedures that May Be Performed

MRI

An MRI is a scan that produces very exact pictures of organs and tumors inside the body. During an MRI scan, your child will lie on a table that is moved into a tube-like machine that surrounds him or her with a magnetic field. The scan is painless; however, the machine makes a loud banging noise that may frighten some children. Your child must be able to lie completely still during the scan. If your child is unable to remain still, sedation may be necessary.



BLOOD TESTS

Blood tests are performed to monitor your child’s blood cells, body salts, and certain proteins or compounds of interest in the blood. These tests primarily are used to monitor your child’s response to treatment and any possible side effects of therapy. A complete blood count helps detect decreases in the number of red blood cells, white blood cells, or platelets in the blood. Blood chemistry tests, such as tests that check the individual’s blood urea nitrogen and creatinine levels, are performed to detect changes in kidney function. Other tests, such as those monitoring the individual’s levels of alanine aminotransferase, aspartate aminotransferase, and bilirubin, may be necessary to assess liver function. Blood tests, such as those monitoring the individual’s rates of erythrocyte sedimentation and C-reactive protein—which sometimes are elevated in cases of Hodgkin lymphoma—may be used to measure the individual’s response to treatment. During your child’s first visit to the hospital or clinic, your treatment team may request blood tests to rule out other diseases, but these tests are not diagnostic for Hodgkin lymphoma.

VENOUS ACCESS DEVICE (VAD)

Chemotherapy usually is needed to treat Hodgkin lymphoma, and there are several line options to help with your child's treatment. Because treatment for Hodgkin lymphoma often is brief, a temporary catheter called a peripherally inserted central catheter may be used. Sometimes, a surgeon will insert a small plastic tube called a venous access line or device (i.e., a port) into a large blood vessel, usually under your child's collar bone. These VADs can be used for blood tests and to give medications, chemotherapy, blood products, and nutritional support when needed. You and your child's treatment team will decide whether a VAD is necessary. A VAD usually is left in place for the duration of treatment and then removed after the completion of therapy.

■ HOW IS HODGKIN LYMPHOMA TREATED?

Hodgkin lymphoma is one of the most treatable cancers, with a long-term survival rate of more than 90% after treatment. The goal of treatment for Hodgkin lymphoma is to cure the disease while minimizing any treatment-related side effects or adverse reactions. Chemotherapy and radiation therapy are the two most common therapies used to treat Hodgkin lymphoma.



Chemotherapy

Chemotherapy is medicine that helps kill cancer cells and prevents the cancer from spreading. Because no single chemotherapy medicine can control the disease by itself, a variety of medications are given in combination to kill the cancer cells. The exact combination of chemotherapy agents your child receives will depend on the stage of the disease and other treatment considerations. Many chemotherapy medications are administered through a vein or VAD, although some are given by mouth. Most chemotherapy treatments are administered in an outpatient clinic; however, some treatments may require hospitalization. Close monitoring of your child will be a priority when they receive chemotherapy. Your child's treatment team will explain the possible side effects of the specific chemotherapy medicines your child receives.

Radiation Therapy

Radiation therapy uses a special type of X-ray treatment that kills or damages cancer cells and may be given before or after chemotherapy. When radiation is used to treat Hodgkin lymphoma, it typically is given after the completion of chemotherapy. Because radiation can result in harm to normal tissues and organs, even long after completion of the therapy, the focus has been on avoiding radiation as much as possible in the treatment of Hodgkin lymphoma. The use of PET scans has enabled treatment providers to identify children who respond well to the first two cycles (1 month each cycle) of chemotherapy treatment. Current research has shown that this group of patients may be cured with only chemotherapy (i.e., they do not receive any radiation) or with small doses of radiation in addition to chemotherapy. However, this is still under investigation and may not be the standard practice where your child is receiving treatment. Children who do not respond well to 2 months of chemotherapy may go on to receive radiation. If radiation therapy is necessary for your child, the radiation therapy doctor will explain to you exactly how the radiation will be given and how long the treatments will last. Most children experience very few side effects while they are receiving radiation therapy. However, some children may feel more tired, have a decreased appetite, or experience redness or darkness of the treated skin. The side effects most likely to occur typically depend on the area of the body being treated. In general, most side effects stop after treatment; however, side effects also can occur after treatment stops. Your child's treatment team will explain any possible late effects in detail.

■ HOW LONG WILL MY CHILD'S THERAPY LAST?

The length of your child's therapy will depend on the stage of the disease and its risk group. Usually, treatment is given for 2–6 months, but it can take longer depending on the specific treatment plan. The duration of treatment may be modified based on your child's response to treatment after 4–8 weeks.

■ WHAT IF THE CANCER RECURS?

It is possible for Hodgkin lymphoma to recur, which is called a recurrence or relapse. If your child has a relapse, his or her treatment may involve additional chemotherapy, radiation therapy, newer agents like immunotherapy or targeted therapy, or stem cell transplant.

Immunotherapy

The immune system is the body's natural defense against infection and disease. Immunomodulators are drugs that modify different parts of that system to boost its function, stop cancer cells from increasing in number, and stop the growth of new blood vessels that provide nutrients to the cancer. This new form of treatment may be used to treat classical Hodgkin lymphoma that was not cured by first-time treatment, and your treatment team may consider using it if additional chemotherapy fails. Because this type of treatment still is new, it usually is administered as part of a clinical trial. We will discuss what this means in the next section on page 8.

Targeted Therapy

Like immunotherapy, targeted therapy also is a new form of treatment. This class of drugs stops the action of molecules that help cancer cells grow by targeting these molecules on the surface of cancer cells. Targeted therapy is not yet widely used to treat Hodgkin lymphoma, but it may be a treatment option if the cancer recurs. The most common targeted therapy used in Hodgkin lymphoma is a drug called brentuximab. This drug binds to a specific protein on the Hodgkin-Reed Sternberg cell to help destroy it.

Stem Cell Transplant

Blood stem cells are cells that eventually develop into mature blood cells. The body makes stem cells in the bone marrow, which is the richest source of these cells. Stem cells also are present in the blood stream, although in small quantities, and their amount can be increased with certain drugs. In the treatment of cancer, stem cells are used to restore blood and immune cell formation after intense chemotherapy, radiation therapy, or both have been administered.

The goal of stem cell transplant in the treatment of Hodgkin lymphoma is to replace the healthy bone marrow stem cells that are destroyed during a very intense form of chemotherapy called high-dose chemotherapy. Unlike the chemotherapy used for first-time treatment of cancer, this high-dose chemotherapy kills off cancer cells and wipes out the stem cells in the bone marrow, meaning they no longer are able to produce blood cells. During a stem cell transplant, stem cells are collected from a patient with Hodgkin lymphoma before high-dose chemotherapy is administered. After the high-dose chemotherapy, the patient's collected stem cells are replaced through infusion so that they are able to function again.

■ WHAT NEW METHODS OF TREATMENT ARE AVAILABLE?

Hodgkin lymphoma has been studied more than any other type of lymphoma. With the many rapid advances in diagnosis and treatment, high survival rates have been reported in children with Hodgkin lymphoma using different treatment strategies. Children with stage I or II disease have a 95% survival rate. Children with stage III and IV disease typically have a survival rate near 90%.

Most advances in the treatment of childhood cancer have been made through a process known as clinical trials. During clinical trials, the best-known (or standard) treatment for a cancer is compared with a new (or experimental) treatment that is believed to be at least as good as, and possibly better than, the standard treatment. Clinical trials enable doctors to determine whether new treatments are safe and effective. Clinical trials for Hodgkin lymphoma have focused on decreasing long-term side effects by reducing or eliminating radiation therapy and tailoring chemotherapy treatments.

Participation in clinical trials is voluntary, and potential candidates must meet specific qualifications. Because clinical trials involve new treatment plans that are experimental, all of the possible risks of the treatment cannot be known ahead of time and unknown side effects may occur. However, children who participate in clinical trials can be among the first to benefit from new treatment approaches. Before making a decision about your child's participation in a clinical trial, you should discuss all of the potential risks and benefits with your child's treatment team.

Additional information about clinical trials is available in the free booklet *Taking Part in Clinical Trials: What Cancer Patients Need to Know* by the National Cancer Institute (Publication No. 98-4250). To obtain the booklet and other useful information about childhood cancer, call 800.4CANCER (800.422.6237). The booklet also can be downloaded from the National Cancer Institute website at www.cancer.gov/clinicaltrials/learning.

■ WHAT ARE THE POTENTIAL LATE EFFECTS OF TREATMENT?

Like all patients treated with chemotherapy and radiation, survivors of Hodgkin lymphoma may be at risk for developing side effects months or even years after treatment ends. Children who received radiation to the chest area as part of their cancer treatment are at risk for breast and cardiac complications. Other late effects may include delayed puberty in males, infertility, thyroid or lung problems, an increased risk of developing another cancer, delayed growth and development, and bone-health issues.

Cancer survivors should seek appropriate cancer-related follow-up care to help them monitor their overall health after treatment ends. Many cancer centers throughout the country include survivor clinics that offer access to experts who can address a wide range of follow-up concerns.

For more information about the specific long-term side effects of cancer treatment, visit curesearch.org.

■ HOW CAN I WORK WITH MY CHILD'S TREATMENT TEAM?

Your child's care and treatment requires a team approach. As a parent or guardian, you are an integral part of the team and your input is important. Because you know your child better than anyone else, the treatment team will need your help with managing your child's disease. It is important to communicate openly with your child's treatment team. Be sure to question your child's oncology provider or nurse whenever there is anything you are not sure about. It often helps to write down your questions when you think of them. Examples of some questions to ask include the following:



- Has the cancer spread beyond the primary site?
- What treatment options are available?
- What treatment do you recommend and why?
- What are the risks or side effects of the recommended treatment?
- What should we do to prepare for treatment?
- What is my child's outlook for survival?
- What are the chances of a recurrence?
- If my child's cancer recurs, are other treatment options available?

Use this space to write down some additional questions.

■ ARE MY FEELINGS NORMAL?

Learning that your child has cancer can be shocking and overwhelming. At first, you may not believe it or may hope the diagnosis is wrong. However, the changes you see in your child and the experience of being in the hospital and beginning treatment no doubt will confirm the reality of your child's situation. Many family members feel they are somehow responsible for the child's disease or feel guilty that they were not able to detect it

sooner. Remember that this disease was not caused or triggered by anything anyone did to the child, anything the child ate, or anything that happened during pregnancy. In addition to shock and guilt, you and your family may feel anger and sadness. Even the youngest members of your family are likely to be affected. These feelings are normal, and each family member will express them in different ways and at different times. It can be very difficult to feel so many strong emotions all at once. Talking honestly with each other about your feelings, reactions, and questions will help everyone in the family. It may seem difficult to talk to friends, family, or even medical staff, but expressing your feelings will help you cope with this situation. Your child also will benefit from family members continuing to show their care through support and communication.

■ HOW CAN I HELP MY CHILD?

You will notice frequent changes in your child during his or her treatment for Hodgkin lymphoma. These changes or symptoms may make you feel even more helpless. It is important to remember that despite changes on the outside, your child is still the same person on the inside. Hair loss and other changes in physical appearance are temporary and often bother adults more than the child or his or her siblings and friends. All of your feelings about what your child is going through during treatment for cancer must be balanced by remembering that treatment provides an opportunity to cure the disease so your child can go on to live a full and meaningful life. It is important to reinforce to your

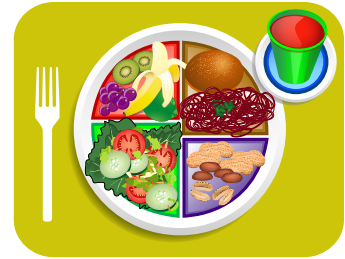


child that nothing he or she did or said caused this disease. Telling your child that your angry or sad feelings are directed at the cancer and not at him or her will help preserve honesty and closeness in your relationship. Like you, your child will need someone with whom to share feelings. Do not hesitate to ask your child to express his or her feelings and do not be afraid to explain what is happening and why.

In spite of the disease, your child is still developing and learning. All children, both sick and well, need love, attention, discipline, limits, and the opportunity to learn new skills and try new activities. As you learn about your child's special needs, it is important to remember that he or she also has all of the needs and rights of any other growing, developing person. Many patients with Hodgkin lymphoma are adolescents, which presents special considerations. Do not avoid using direct terms and explanations with your child. Children will tolerate treatment better if they understand it and are allowed to be active decision makers whenever possible. The same is true for parents and family members.

■ IS MY CHILD'S DIET DURING TREATMENT IMPORTANT?

Yes. We know from research that well-nourished children tolerate therapy better and have fewer treatment delays from illness. It may be difficult for your child to resume normal eating habits while receiving therapy, so you will need to be flexible and creative. Your child's food preferences and tastes may change throughout therapy. Often, numerous small meals are better tolerated than three large ones. Children usually are more interested in eating foods they helped prepare. It is important to include your child in the social activity of family meals even if he or she does not eat full meals. Remember, nobody wins food fights—it is best not to force your child to eat. Make sure foods that are high in protein and carbohydrates are readily available. Your treatment team should approve multivitamins, herbs, supplements, and all other medicines before you give them to your child because they may cause an interaction with your child's chemotherapy. A dietitian trained in the calorie and energy needs of children with cancer can offer you guidance. Your child's treatment team also can intervene if there is a nutritional problem.



■ CAN MY CHILD ATTEND SCHOOL DURING TREATMENT?

Most children who are being treated for Hodgkin lymphoma can attend school. However, some children may not tolerate chemotherapy and radiation as well as others and may need to miss school on occasion. You should discuss school attendance with your child's oncology team. School is important because it helps children and adolescents maintain social contact with their peers. It is important for your child to return to school as soon as he or she is medically able to do so.



Suggested Resources

www.curesearch.org

National Childhood Cancer Foundation and the Children's Oncology Group

www.cancer.gov

National Cancer Institute

www.cancer.gov/clinicaltrials/learning

Webpage from which the "Taking Part in Clinical Trials: What Cancer Patients Need to Know" booklet can be downloaded.



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