



# Childhood Leukemia and Lymphoma

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*RN.com acknowledges the valuable contributions of...*

***Kim Maryniak, PhD, MSN, RNC-NIC, NEA\_BC;***

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## Purpose

The purpose of this course is to provide information about childhood leukemia and lymphoma. The course includes information on signs and symptoms of leukemia, how leukemia is diagnosed, childhood cancer statistics, current treatment patterns, long term side effects, and the emotional and psychosocial issues surrounding a childhood cancer diagnosis.

## Learning Objectives

After successful completion of this course, you will be able to:

1. Define childhood leukemia and lymphoma.
2. Identify the eight warning signs of childhood cancer.
3. Identify signs and symptoms of childhood leukemia and lymphoma.
4. Describe how leukemia and lymphoma is diagnosed and staged.
5. Identify the current treatment for childhood leukemia and lymphoma.
6. Describe side effects associated with treatment.
7. Describe statistics for childhood leukemia and lymphoma.
8. Describe treatment for pain associated with childhood leukemia and lymphoma.
9. Identify stressors, psychosocial issues and positive coping strategies.

## Introduction

Children who are diagnosed with leukemia or lymphoma develop symptoms and side effects which are different than those experienced by adults. A pediatric patient can become acutely ill quickly, presenting a medical challenge for the healthcare team. Diagnosis, treatment, and intervention by a specialized pediatric hematology oncology center can help ensure a positive outcome for ill children and adolescents with cancer.

Acute lymphocytic leukemia, also known as acute lymphoblastic leukemia (ALL) is the most common hematologic malignancy found in children. Fortunately ALL is treatable and can be cured. Survival rates have increased dramatically in recent decades due to advances in research and chemotherapy treatment (National Cancer Institute, 2018a).

This course will discuss childhood leukemia and lymphoma, childhood cancer statistics, life-saving treatment, side effects of treatment, and the stressors and psychosocial issues surrounding a diagnosis of childhood cancer.

### Did You Know?

Acute lymphocytic leukemia, also known as acute lymphoblastic leukemia (ALL) is the most common hematologic malignancy found in children.

## The Warning Signs of Childhood Cancer

The initial signs and symptoms of childhood cancer can be insidious. Young children may be unable to communicate pain or discomfort. Older children might not report symptoms of prolonged tiredness or fever (American Society of Clinical Oncology [ASCO], 2018).

### Early Warning Signs of Childhood Cancer

**C**ontinued, unexplained weight loss

**H**eadaches, often with vomiting in the morning

**I**ncreased swelling or persistent pain in bones, joints, back, or legs

**L**ump or mass, particularly in the abdomen, neck, underarms, chest, pelvis, or elsewhere

**D**evelopment of excessive bleeding or bruising

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Constant infections

A whitish color behind the pupil

Nausea that persists or vomiting without nausea

Constant tiredness or noticeable paleness

Eye or vision changes with sudden occurrence and persistence

Recurrent or persistent fevers without a known origin

(ASCO, 2018)

## Childhood Cancer Statistics

Although many forms of childhood cancer can be treated and cured, the loss of any child to cancer can be devastating. Statistics regarding cancer and children report that:

- Leukemia is the most common cancer in children and adolescents. It accounts for about one out of three cancers in children, or approximately 33%. Overall, however, childhood leukemia is a rare disease (American Cancer Society [ACS], 2018e).
- Acute lymphocytic leukemia (ALL) accounts for about three out of four leukemia cases among children and teens. Most of the remaining cases are acute myelogenous leukemia (AML). Chronic leukemias are rare in children (ACS, 2018e).
- ALL is most common in early childhood, peaking between two and four years of age. Cases of AML are more spread out across the childhood years, but it is slightly more common during the first 2 years of life and during the teenage years (ACS, 2018e).
- **ALL** is slightly **more common** among **Caucasian and Hispanic** children than among African-Americans and Asian American children and is more common in **boys** than in girls. AML (acute myelogenous leukemia) occurs equally among boys and girls of all races (ACS, 2018f).
- The **five year survival rate for ALL is now more than 85% overall** (ACS, 2018e). This is mainly due to advances in treatment.
- The five year survival rate for AML is approximately 60-70% (ACS, 2018e).

Children develop different types of cancer than adults. They also respond differently to treatment than adults.

## Test Your Knowledge

Childhood leukemia accounts for \_\_\_% of child cancers.

- A. 15%
- B. 22%
- C. 33%
- D. 41%

**The correct answer is: 33%.**

## What is Leukemia?

Leukemia is a cancer and disease of the bone marrow and blood cells. Bone marrow fills the cavities within the bones and makes blood cells. In the presence of leukemia, the bone marrow makes large amounts of abnormal white cells. As the leukemia cells increase and multiply, normal cells decrease and are pushed out. The abnormal white cells crowd out red blood cells and platelets resulting in anemia and thrombocytopenia. As the abnormal white cells multiply, the child will become ill and exhibit signs of leukemia (ACS, 2018e).

## Types of Childhood Leukemia

There are many types of childhood leukemia; however, the most common types are acute lymphoblastic leukemia (ALL) and acute myelogenous leukemia (AML).

Types of leukemia are identified by the type of cell affected. A pediatric patient's diagnosis of childhood leukemia will be classified by cell type and will aid in determining the prognosis for the child.

The prognosis is a determination of how the child will respond to treatment and how long the patient is expected to live. Each child can respond differently to leukemia treatment. For this reason many hematologists are reluctant to give a parent a definitive prognosis or expected timeline (ACS, 2011b & 2018g).

### **Test Your Knowledge**

Sarcoma is the most common cancer occurring in children.

- A. True
- B. False

**The correct answer is: False. ALL (Acute Lymphatic Leukemia) is the most common occurring cancer in children.**

## Leukemia Signs and Symptoms

Children who present with leukemia may appear to be asymptomatic. Signs and symptoms of leukemia which do occur are a result of inadequate blood cells. Common symptoms include:

- Petechiae, with easy bruising and bleeding
- Frequent or severe nosebleeds
- Bleeding gums
- Fatigue, weakness
- Temperature instability
- Headaches
- Vertigo
- Shortness of breath
- Pallor
- Infections

(ACS, 2018g)

## Leukemia Diagnosis

A diagnosis of childhood cancer can precipitate a crisis for the child, family, healthcare team, and the community. Since time affects prognosis, the healthcare team must act rapidly to ensure a positive outcome. Today, advances in treatment have improved the outlook of survival for children diagnosed with leukemia.

The most important factor in the management of leukemia is to determine the subtype of the leukemia. This is done by testing samples of the blood, bone marrow, and sometimes lymph nodes or CSF (ACS, 2018g).

## Classification of Leukemia

Classification of the leukemia plays a major role in determining both treatment options and a child's outlook (prognosis) (ACS, 2018g).

### Did You Know?

**Leukemia is not staged like most other cancers. It already involves the bone marrow and blood. But it is important to know whether the leukemia cells have started to collect in other organs such as the liver, spleen, or lymph nodes.**

## Why Do Children Get Leukemia?

The etiology of pediatric leukemia remains unknown. There are risk factors and signs of childhood cancer that parents and healthcare workers should be watchful for. Cancer and leukemia detected early may have a better chance of responding to treatment. The risk of leukemia cells spreading to the child's other organs is also decreased with early detection.

### Risk Factors for Childhood Leukemia

Certain risk factors can help to identify the chances of a child being diagnosed with leukemia. They include:

- Age
- Inherited genetic conditions including Down syndrome (trisomy 21), Li-Fraumeni syndrome, Fanconi anemia, Klinefelter syndrome, neurofibromatosis, and ataxia telangiectasia.
- Children receiving medication to suppress the immune system.
- Children who have rare immune disorders.
- Children with a sibling that has been diagnosed.
- Children previously diagnosed with cancer or leukemia.
- Children who have received chemotherapy or radiation.
- Viruses or infections early in life.

(ACS, 2018f)

## Diagnosing Leukemia in Children

An initial diagnosis assessment for childhood leukemia may include:

- A complete in-depth physical assessment.
- A health history from the caregiver and pediatrician. Has the child lost weight? Is there a lack of energy? Has there been bruising and prolonged bleeding? Was the child taking medication? Is there a family history of cancer or bleeding? Does the child have any additional health problems?
- Laboratory analysis, including a complete blood count (CBC) with differential, obtain slide for assessment, electrolytes, type and cross, liver function tests, blood glucose, chromosome analysis, and blood gases.
- Chest x-ray, a computed tomography (CT) scan, magnetic resonance imaging (MRI), and/or bone scan may be indicated.
- Bone marrow aspiration and bone marrow biopsy.
- Lumbar puncture to examine the cerebral spinal fluid (CSF).
- Chromosome studies.

(ACS, 2018g)

## Prognosis and Recovery

The following factors can affect a child's prognosis for recovery:

(ACS, 2018g)

### Age

Age at diagnosis of leukemia. Ages one year to nine years respond well.

### White Blood Count (WBC)

The higher the white blood count (WBC), the higher the risk. WBC counts greater than 100,000 are especially dangerous and increase the risk of relapse.

### **Gender**

Gender affects prognosis. Girls have a better chance of recovery than boys.

### **Race**

Race affects response. African American and Hispanic children have a lower cure rate than Caucasians.

### **Metastases**

Metastases to other parts of the body. When leukemia cells have spread to the spleen, liver, brain, testicles, and other organs, the prognosis for recovery is lower.

### **Immunophenotype**

Immunophenotype (the presence of B or T cell antigens) of the leukemia cell. T cells are higher risk.

### **Chromosomes**

The number, types, and translocation of chromosomes.

### **Response**

Response to therapy. A primary indicator of a patient's recovery is how well they respond to treatment. Many children with poor prognosis will surprise the medical team by responding well. Infrequently, a child with a good prognosis may not respond to treatment. While prognostic statistics are helpful, a patient's response and survival is individual.

## **Antigen Testing**

Acute lymphocytic leukemia are classified by their B-cell or T-cell status, which is determined by identifying the antigens on the cell surfaces. Tests for antigens can help determine whether the leukemia cells started in B cells or T cells, as well as how mature these cells are (ACS, 2018g).

## **Treatment for Childhood Leukemia**

Childhood leukemia is treated with a combination of chemotherapy, radiation, and bone marrow or stem cell transplant. The goal of treatment includes the following:

- Kill the leukemia/cancer cells.
- Control the symptoms of disease in the child.
- Achieve a remission of the disease and the symptoms.
- Cure the disease.

(ACS, 2018h)

## **Treatment Considerations**

Some children with leukemia are critically ill when they are first diagnosed with leukemia.

- A shortage of normal white blood cells may lead to very serious infections.
- Low blood platelet levels can cause severe bleeding.
- Not having enough red blood cells can lower the amount of oxygen getting to body tissues and put a tremendous strain on the heart muscle.

These problems must often be considered before treatment of the leukemia can begin. Antibiotics, blood growth factors, and transfusions of platelets and red blood cells may be given to treat or prevent some of these conditions (ACS, 2018h).

## Treatment Modalities

The initial treatment for leukemia depends on several different factors including:

- Leukemia cell type and stage.
- Eligibility for a clinical trial. Clinical trials are studies which evaluate the effectiveness of new treatments.
- The parents' understanding of the treatment, approval, and their written legal consent. Social, cultural, and economic factors need to be considered. Examples might include the populations without access to health insurance, individual beliefs in herbal remedies, or families of religious faith who believe in bloodless treatment. The healthcare team should make every effort to include the family's beliefs while providing effective treatment for the child.
- The hospital and hematologist caring for the child. The hematologist is ultimately responsible for writing the orders which determine treatment.
- The child's age and physical condition. Each child should be evaluated individually. Physical conditions which could affect treatment modalities include, heart, liver or organ failure, poor kidney function, decreased pulmonary function, among others. Treatments may need to be modified to decrease further risk of side effects.

**It is important that children with ALL have their treatment planned by a team of doctors with expertise in treating childhood leukemia (National Cancer Institute (NCI, 2018a).**

### ***Test Your Knowledge***

Which of the following factors will not affect the child's initial treatment?

- A. Eligibility for a clinical trial
- B. Age
- C. Gender
- D. The hematologist

***The correct answer is: Gender.***

## Treatment of Childhood ALL

The treatment of childhood ALL is done in three phases and always includes combination chemotherapy:  
(Source: NCI, 2018a)

### **Phase One**

Remission Induction Therapy

This is the first phase of treatment, with the purpose to kill the leukemia cells in the blood and bone marrow. This puts the leukemia into remission.

### **Phase Two**

Consolidation/ Intensification Therapy

This is the second phase of therapy that begins once the leukemia is in remission. The purpose of consolidation/ intensification therapy is to kill any remaining leukemia cells that may not be active but could begin to re-grow and cause a relapse.

## **Phase Three**

### Maintenance Therapy

This is the third phase of treatment, with the purpose to kill any remaining leukemia cells that can re-grow and cause a relapse. Usually the cancer treatments are given in lower doses than those used for induction and consolidation/intensification therapy.

## **Supportive Care**

Supportive care is an important part of treatment for childhood leukemia before, during, and after therapy. Supportive care includes:

- Antibiotics to prevent or treat infection caused by immune suppression.
- Hydration and electrolyte replacement. Vomiting and nausea, poor appetite, treatment, and disease predispose the child to a poor oral intake and dangerous electrolyte imbalances.
- Red blood cell transfusions to treat anemia, and platelet transfusions to treat thrombocytopenia (decreased platelet count) and control bleeding.
- Granulocyte stimulating agents to increase low white blood cell and decreased granulocyte counts.
- Red blood cell stimulating agents to increase the red blood cell count.
- Allopurinol medication to help decrease uric acid in the body and prevent tumor lysis syndrome. Tumor lysis occurs when killed tumor cells flood into the blood stream and organs causing life threatening conditions.
- Nutritional support for the child unable to maintain sufficient calorie intake.

(NCI, 2018a)

## **Surgery**

The child may need surgery to:

- Insert a central venous line to administer medication and hydration.
- Obtain a bone marrow biopsy under anesthesia.
- Treat the side effects caused by treatment. An example would be a large abscess needing incision and draining.

(ACS, 2018d)

### **Did You Know?**

**Surgery is rarely used to treat childhood leukemia as there is seldom a solid tumor to remove.**

## **Chemotherapy**

Chemotherapy is the backbone of treatment for children diagnosed with leukemia. Chemotherapy is used to kill or control leukemia or cancer cells. There are currently dozens of medications used to treat childhood leukemia. These medications may be administered by the following routes:

- Intravenously via central lines or peripherally venous access
- Orally, by mouth
- Intramuscular injection
- Subcutaneous injections
- Intrahepatic
- Intracranial
- Intrathecal (into the spinal fluid)
- Intra-organ (chemotherapy administered directly into organs)

(NCI, 2018a)

## Common Chemotherapeutic Drugs

Common medications used to treat childhood leukemia also include combinations of the following drugs:

- Vincristine sulfate
- Daunorubicin hydrochloride
- Doxorubicin hydrochloride
- Asparaginase Cytarabine Methotrexate
- Mercaptopurine Prednisone
- Cyclophosphamide

For a current list of approved chemotherapy medications, refer to the National Cancer Institute list at <http://www.cancer.gov/cancertopics/druginfo/leukemia#dal1>

## Radiation

Radiation is the administering of x-rays used to kill cancer cells.

- External radiation: uses a machine outside the body to send radiation toward the cancer.
- Internal radiation: uses radioactive substance sealed in needles, seeds, wires, or catheters that are placed directly near the cancer.

Radiation may be used for:

- Children who present with leukemia cells in their brain or spinal fluid.
- Children who suffer from a central nervous system (CNS) relapse.
- Patient with metastasis/spread to other organs.
- Children being prepared for a bone marrow or stem cell transplant.

(NCI, 2018a)

## Test Your Knowledge

Radiation may be used for treatment of leukemia cells in the brain or spinal fluid.

- A. True
- B. False

**The correct answer is: True.**

## Bone Marrow and Stem Cell Transplants

Bone marrow and stem cell transplants may also be used to treat childhood leukemia. In a transplant, a patient receives an intravenous transfusion of bone marrow or stem cells, replacing the patient's own marrow. The child will receive chemotherapy and/or radiation to kill all the bone marrow cells prior to infusing the new marrow and cells. The goal is for the new healthy marrow to be accepted by the patient to prevent the patient's body from making leukemia cells (NCI, 2018a).

## What is a Stem Cell Transplant?

Stem cells are immature blood cells that are removed from the blood or bone marrow of a donor. After the patient receives very high doses of chemotherapy and occasionally radiation therapy, the donor's stem cells are given back to the patient through an infusion. The re-infused stem cells grow into and restore the patient's blood cells. A stem cell transplant may use stem cells from a donor who is or is not related to the patient.

Stem cell transplant is rarely used as initial treatment for children and teenagers with ALL. It is used more often as part of treatment for ALL that relapses (NCI, 2018a).

## Indications for Stem Cell Transplants

Indications for a transplant include:

- Relapse. The leukemia returns, remission is not sustained.
- Children whose disease is unresponsive to chemotherapy or conventional treatment. The child has not achieved remission.
- Rare leukemia's which can be cured with a bone marrow transplant.
- Children with rare leukemia who have a perfect sibling bone marrow match.
- Stem cell aphaeresis donation for children receiving regimens that suppress the immune system and that will benefit from a bone marrow rescue or boost.

(NCI, 2018a)

## Sources of Bone Marrow & Stem Cell Transplants

Bone marrow transplants may come from several sources:

- **Autologous:** The patient's own marrow. This type of transplant is not used in childhood leukemia due to the risk that the leukemia will relapse.
- **Allogeneic:** Matched related or unrelated donor. Usually the patient's sibling.
- **Matched unrelated donor (MUD transplant):** Usually from a bone marrow donor bank.
- **Umbilical cord stem cell transplant:** Patient's own or matched donor.

### Did You Know?

Bone marrow transplants may come from the patient's own marrow.

## Determining Type of Transplant Needed

The type of transplant deemed necessary will depend on the following:

- Child's protocol (treatment plan).
- The disease status and disease type, the relapse diagnosis. Relapsed leukemia can change cell type.
- The available bone marrow or donor. Does the child have a sibling? What if the child is adopted? Can the patient donate his/her own marrow?
- Can the child physically withstand a bone marrow transplant and associated pre-treatment?

(NCI, 2018a)

## Targeted Therapy

Targeted therapy is treatment that uses drugs or other substances to identify and attack specific cancer cells without harming normal cells.

Tyrosine kinase inhibitors (TKIs): targeted therapy drugs that block the enzyme, tyrosine; which causes stem cells to generate more white blood cells or blasts than the body needs.

This treatment is often used in children with Philadelphia chromosome type ALL (NCI, 2018a).

## Understanding Lymphoma: The Lymph System

The lymphatic system is part of the immune system and helps the body fight against infection. Elements of the immune system include:

- Lymph nodes, including intestinal lymph nodes
- Lymphatic vessels
- Tonsils and adenoids
- Spleen and bone marrow
- Skin
- T lymphocytes and B lymphocytes
- Natural killer (NK) cells
- Plasma cells
- Lymphokines

The parts of the lymph system combine to fight disease and infection. When normal cells within the lymph system mutate to malignant cells, they can multiply into a dangerous lymphoma that can spread throughout the body (ACS, 2018a).

## What is Lymphoma?

Lymphoma is a type of cancer which develops in the lymphatic system and is more commonly diagnosed in adolescents and young adults. Pediatric patients and adolescents diagnosed with lymphoma usually respond well to treatment, particularly chemotherapy and/or radiation. Abnormal lymphocytes grow, multiply, and form tumor masses. Lymphomas generally start in the lymph nodes or in organs. Lymphomas can spread to the bone marrow and throughout the body (ACS, 2018a).

## Test Your Knowledge

Lymphoma is more commonly diagnosed in adolescents and young adults.

- A. True
- B. False

**The correct answer is: True.**

## Types of Lymphoma

There are several different types of lymphoma. The two main types of lymphoma are:

- Hodgkin lymphoma (also called Hodgkin disease)
- Non-Hodgkin lymphoma

(ACS, 2018a)

### Hodgkin Lymphoma

Hodgkin lymphoma is more common in adolescents and young adults. Hodgkin lymphoma is characterized by the presence of a type of malignant cells identified as Reed-Steinberg cells. It is more common in two age groups: early adulthood (age 15 to 40, usually people in their 20's) and late adulthood (after age 55). Hodgkin lymphoma is rare in children younger than five years of age. This is one type of cancer that is very similar in children and adults, including which types of treatment work best. The cause of Hodgkin lymphoma is unknown and the incidence of Hodgkin lymphoma increases with age. Hodgkin disease usually responds well to treatment and a cure is possible (ACS, 2018b).

## Non-Hodgkin Lymphoma (NHL)

Non-Hodgkin lymphoma (NHL) includes all lymphomas except Hodgkin lymphoma. These are a large diverse group of malignant cell clusters originating from the immune system cells. These cells in the lymph system are less differentiated. Non-Hodgkin lymphoma also makes up about 4% of childhood cancers. It is more likely to occur in younger children than Hodgkin lymphoma is, but it is still rare in children younger than age three. The most common types of NHL in children are different from those in adults. These cancers often grow quickly and require intensive treatment, but they also tend to respond better to treatment than most non-Hodgkin lymphomas in adults. The chance of recovery is good if the cancer is not wide spread. NHL is most often found in adults and less common in children and adolescents. NHL is classified as slowly progressing or aggressive rapidly progressing (ACS, 2018c).

## Risk Factors for Lymphoma

The majority of lymphomas occur without a known cause; the etiology is unknown. The incidence of lymphoma has risen over the past three decades and the principle cause for this is unknown. Known risk factors for lymphoma include:

- Children/adolescents with a disease of the immune system.
- Children/adolescents on medication that suppresses the immune system.
- Patients who have received an organ transplant and are receiving immune suppressant medication.
- Lymphoma rates increase with age, and is uncommon under the age of five.
- Gender is a known risk, as the disease is more prevalent in males.
- Prolonged chemical or environmental exposure to cancer causing agents.
- Certain virus including human immunodeficiency virus (HIV), Epstein Barr virus (EBV), and human T cell lymphocytotropic virus (HTLV). Helicobacter pylori bacterium is associated with stomach ulcers and lymphoma of the stomach wall

(ACS, 2018b)

## Test Your Knowledge

Lymphoma is more prevalent in which gender?

- A. Males
- B. Females
- C. Equally prevalent in both genders

**The correct answer is: Males.**

## Signs and Symptoms

The most common early sign of lymphoma is painless swelling or lump in the neck, upper chest, groin, armpits, and abdomen. Other symptoms include:

- Fatigue, the child exhibits unexplained tiredness
- Fever and/or night sweats, sweating occurs mainly at night
- Weight loss, particularly rapid and unexplained
- Loss of appetite, the teen or child may lose his appetite
- Itching, or a feeling of skin itching or skin crawling

(ACS, 2018b)

## Diagnosis

Diagnosing lymphoma may include the following:

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- In-depth physical exam as the child/teen may exhibit different symptoms than an adult. Feel for lymph node abnormality and any swelling.
- Medical and family history.
- Complete blood analysis (blood count, and blood chemistry).
- Chest x-ray.
- Computerized tomography (CT) scan.
- Positron emission tomography (PET) scan.
- Magnetic resonance imaging (MRI).
- Imaging of the chest, pelvis, and abdomen to look for spread of the disease.
- Gallium scan to check the lymphatic system for disease.
- Bone marrow aspiration and biopsy to see if lymphoma has spread to the bone marrow.
- Spinal tap to see if disease has spread into central nervous system.
- Exploratory staging surgery to look for extent of disease.
- Lymph angiogram to check the lymph system for disease.

(NCI, 2018b)

## Staging for Lymphoma

The stage of lymphoma determines how extensive the disease is and how it is likely to be treated. Most importantly, it is an indicator of how the disease will fare after treatment. Staging for lymphoma is based on the following factors:

- The **number** of abnormal lymph nodes present.
- The **location** of the lymph nodes.
- If the cancer **spread** outside the lymph nodes.
- If the abnormal cells are slow or fast-growing/**aggressive**.
- The cell **type**.
- If the abnormal cells are **orderly** or poorly differentiated.
- The cell **grade**.

(ACS, 2018c)

## Prognosis and Recovery

The following factors can affect a child's prognosis for recovery of NHL:

(NCI, 2018b)

### Age at Diagnosis

Infants, although rarely diagnosed, have poorer outcomes. Adolescents have been reported to have inferior outcomes compared with younger children.

### Site of Disease

In general, patients with low-stage disease (i.e., single extra-abdominal/extra-thoracic tumor or totally resected intra-abdominal tumor) have an excellent prognosis (a five-year survival rate of approximately 90%), regardless of histology. Involvement of mediastinum, skin, or viscera is associated with poor outcomes.

### Response to Therapy

A primary indicator of a patient's recovery is how well they respond to treatment. Many children with a poor prognosis will surprise the medical team by responding well. Infrequently, a child with a good prognosis may not respond to treatment. While prognostic statistics are helpful, a patient's response and survival is individual.

## Treatment for Lymphoma

- ✓ Surgery
- ✓ Chemotherapy (common medications include daunorubicin, vincristine, doxorubicin, L-asparaginase, cyclophosphamide, cytarabine, methotrexate, 6-mercaptopurine, 6-thioguanine)
- ✓ Radiation
- ✓ Immunotherapy- including prednisone and intrathecal methotrexate
- ✓ Bone marrow transplant
- ✓ Stem cell transplant

For a current list of approved treatment recommendations based on staging and type of lymphoma, refer to the National Cancer Institute list at <http://www.cancer.gov/cancertopics/pdq/treatment/child-non-hodgkins/HealthProfessional/page4>

## Side Effects of Leukemia and Lymphoma Cancer Treatments

Healthcare workers need to train families and patients to recognize and report side effects such as (The Leukemia and Lymphoma Society, 2015):

Nausea, vomiting, decreased appetite, weight loss	Pain secondary to disease and treatment
Fatigue, lethargy	Decreased white blood count, anemia
Skin rash, mucositis, and mouth sores	Fevers, infection secondary to bone marrow immunosuppression
Cardiac toxicity, pulmonary toxicity	Infertility
Bleeding	Hair loss (alopecia)
Diarrhea	Extravasations of veins and subcutaneous skin burns from vesicant chemotherapy
Depression, anxiety, sleep disturbances	Neuropathy

### **Test Your Knowledge**

Side effects of treatment include all but one of the following:

- A. Skin rash
- B. Anemia
- C. Excessive sweating
- D. Nausea and vomiting

**The correct answer is: Excessive sweating.**

## Managing Side Effects of Leukemia and Lymphoma Treatment

Children receiving cancer treatment can suffer unpleasant side effects. The goal of treatment therapy is to control and kill the leukemia and lymphoma cells. While suppressing the malignant cells, treatment may also suppress the child's immune system leading to life threatening complications. Side effects can be treated and controlled leading to increased cure rates and decreased mortality rates.

These side effects can be mild or serious requiring hospitalization. Each child is an individual and will respond to the treatment uniquely. The healthcare worker needs to stay aware of each patient's response. A baby may experience diarrhea and weight loss. A five year old may suffer from anemia and fevers. A teenager could be prone to depression. The majority of patients receiving chemotherapy and other cancer treatments will experience more than one side effect.

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These side effects can be treated effectively and the child should be supported during therapy. Prompt, proper support can increase the child's chances to recover from leukemia or lymphoma (The Leukemia and Lymphoma Society, 2015).

## Pain Management for Children Diagnosed with Leukemia and Lymphoma

Children and adolescents experience pain from abnormal cells and side effects from treatment. Children relate pain differently from adults. A young toddler may not be able to voice pain. Crying and grimacing often indicates pain in babies. A school age child may withdraw and exhibit a lack of appetite. A teen may exhibit acting out and anger when in pain. Face charts and number charts are helpful in assessing pain in adolescents and children.

(Leukemia & Lymphoma Society, 2013a)

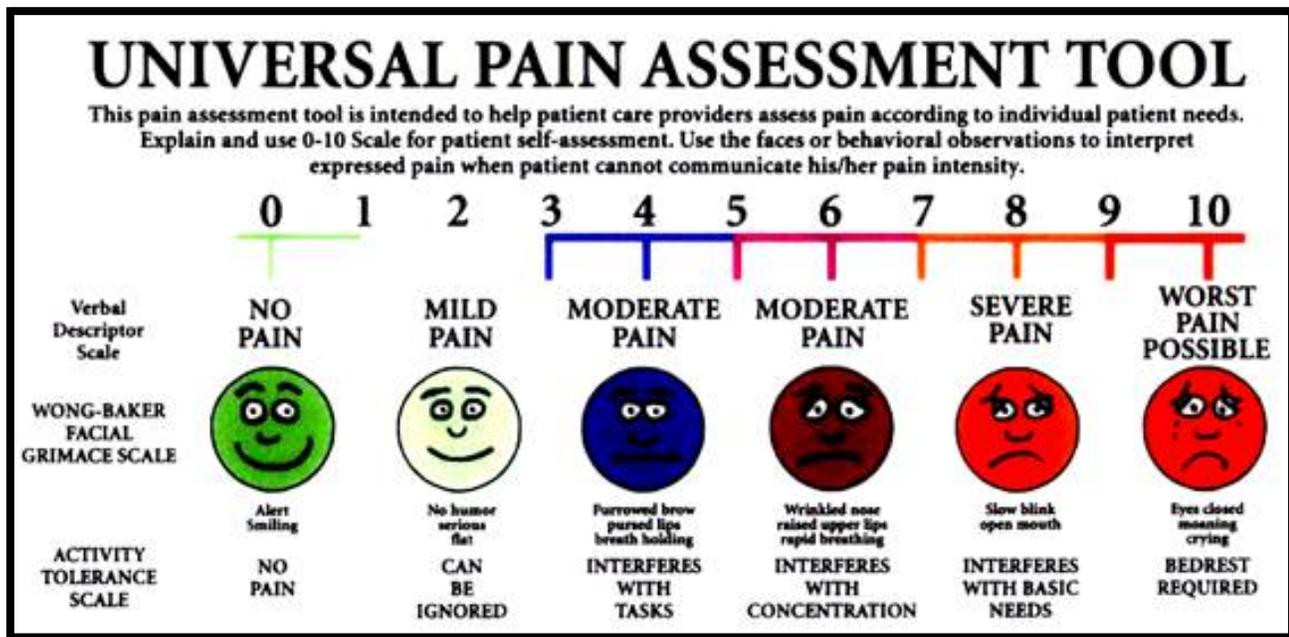


Image Source: [www.pacificu.edu](http://www.pacificu.edu)

## Pain Management Techniques

The following are five pain management techniques to utilize with your patients:

(Leukemia & Lymphoma Society, 2013a)

### Distraction

Young children react well to distracting games and toys, videos, and TV when in pain or during a painful procedure.

### Music

Soothing music and book tapes help children and teens cope with pain.

### Play Therapy

Board games and play activities allow young patients an outlet from pain.

### Massage

Massage therapy can be comforting and help relieve cramps, aching joints, tired back, and pain secondary to treatment.

### Visualization

Teaching techniques which allow a child or adolescent to visualize positive and calming scenes like a sunny beach can help relieve pain.

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## **Test Your Knowledge**

Young children react well to distractions when they are in pain or during a painful procedure.

- A. True
- B. False

*The correct answer is: True.*

## **Pain Medication: Analgesia**

When pain increases and the child exhibits signs and symptoms of discomfort, medication can effectively control pain. Commonly used analgesics for children and adolescents diagnosed with mild to moderate pain related to leukemia and lymphoma include local anesthetic topical via patches, creams, gels and the following:

- Oral analgesics
- Non-opioid analgesics
- Acetaminophen
- Ibuprofen
- Ketorolac
- Naprosyn
- Rofecoxib

(Leukemia & Lymphoma Society, 2013a)

## **Pain Medication: Opioids**

Children receiving opioids need to be frequently observed for pain response and respiratory depression.

Commonly used opioid pain medications include:

- Codeine
- Meperidine oxycodone
- Methadone
- Morphine
- Hydromorphone
- Fentanyl

(Leukemia & Lymphoma Society, 2013a)

## **Administration of Opioids**

Opioids may be administered orally, intravenously, via topical skin patches, implanted pumps, and reservoirs. The child needs to be frequently observed for pain response and respiratory depression while taking opioids. Pain control allows the young child to get on with the work of play and discovery and use positive resources to fight and recover from their disease.

The adolescent with effective pain control will have the ability to enjoy listening to music, watching television, and interact with peers. He will be better prepared to cope with the rigors of chemotherapy or radiation if his pain is controlled (Leukemia & Lymphoma Society, 2013a).

## **Long Term Effects**

Children are surviving leukemia and lymphoma in great numbers, but many suffer long term side effects. These side effects can be effectively managed with appropriate intervention.

Assessment in a pediatric long-term follow up clinic can help detect late effects, and referral to the appropriate professionals can minimize or control these side effects.

<b>Long-term effects include:</b>
Cognitive and learning deficits
MRDD (mental retardation developmental disabilities)
Vision changes, blindness, & hearing loss
Secondary leukemia & cancers
Infertility
Depression & anxiety disorders
Social disorders
Organ damage
Cardiology complications & pulmonary dysfunction
Gastroenterology & liver disease
Chronic pain

## Psychosocial and Emotional Aspects of Childhood Cancer

When a child is undergoing treatment for leukemia or lymphoma, the family faces many stressors. The diagnosis and treatment can precipitate a crisis for the pediatric patient, parents, siblings, and extended family. All these individuals need support and nurturing. Family centered care is a favorable goal that will help the child and family to cope effectively with the diagnosis.

Each family and patient will experience individual stressors and coping mechanisms. Individual response should be respected. Children and families cope differently with a diagnosis of leukemia or lymphoma.

Feelings and stressors associated with a diagnosis of childhood leukemia or lymphoma on initial diagnosis include: (Leukemia & Lymphoma Society, 2013b)

Shock associated with diagnosis	Confusion
Fear of cancer, diagnostic tests, hospital environment, treatment, chemotherapy, and death	Denial
Helplessness related to loss of control	Depression
Grief related to an ill child, loss of a healthy child	Financial crisis
Anger- “why me” syndrome, feels diagnosis is “unfair”, anger over being ill	Strain on marriage and family relationships
Guilt	Role changes
	Developmental delay

## Survival and Life after Cancer Treatment

Patients and families find life after cancer therapy hopeful yet frightening. Staff can help this transition by providing clear guidelines for follow up care and education. Individuals cope differently in their reaction to survival as health improves and life returns to normal (The Leukemia and Lymphoma Society, 2013b).

## Test Your Knowledge

Consistent and clear information about follow up helps the family transition after a diagnosis of leukemia or lymphoma.

- A. True
- B. False

**The correct answer is: True.**

## Case One Study

A two year old Caucasian female, Lily, is rushed by her parents to the local children's hospital emergency room. The toddler appears acutely ill; she is chalky, pale, listless, bruised, and having difficulty breathing. The ER team responds immediately performing a comprehensive physical assessment. Lily's parents are fearful and anxious, the ER staff solemn and watchful. A preliminary diagnosis of childhood cancer is difficult for the family and healthcare workers to consider. Will the little girl survive? The emergency room staffs works quickly with the pediatric hematologist oncologist to treat and diagnose the ill child. Her preliminary blood work is consistent with Acute Lymphoblastic Leukemia, ALL. Lily will be admitted to the pediatric hematology oncology unit for observation and further assessment.

Lily's parents brought her to the emergency room quickly; within a week of symptoms and deteriorating health. Early detection of childhood leukemia may help her chances to survive. Lily exhibited signs and symptoms of anemia, lethargy, and fever. While fevers and childhood illness can mimic childhood leukemia, Lily's overall appearance of profound illness alerted her parents to a serious disorder.

## Test Your Knowledge

Which blood cells in Lily's bone marrow may have been affected?

- A. White blood cells
- B. Red blood cells
- C. Platelets
- D. All of the above

**The correct answer is: All of the above.**

## Case Study One: Risk Factors and Diagnosis

Two year old Lily's risk factor is her age. The incidence of childhood leukemia increases at age two through age eight years. Her age also increases her chances for survival. However her cell type and chromosome analysis will also be important to gauge how she will respond.

Lily was admitted to the Hematology Oncology Unit of a large teaching children's hospital affiliated with a university. Their multidisciplinary team will begin the process of finding out what kind of leukemia Lily has and which treatment offers the best hope for survival. Lily's physicians and nurses perform a complete physical assessment including medical and family history. Lily's parents are overwhelmed with many questions about their daughter's condition. Lily's laboratory results revealed an abnormally high WBC (white blood cell count) of 60,000/mcL, a low hemoglobin of 5.0 g/dL, and decreased platelet count of 5,000/mcL (thrombocytopenia). A bone marrow biopsy was performed with a spinal tap. The preliminary tests are consistent with ALL.

## Case Study One: Potential Side Effects

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Lily's treatment for ALL will be determined by the Hematology Team. Treatment will begin as soon as possible, after all her staging tests have been completed and after her parents provide their consent. The hematologist explains Lily's disease and the available treatment options. Lily's mother cries as she hears the possible side effects from treatment. Lily could experience hair loss, infection, infertility, and secondary tumors. Her parents both agree that this is Lily's best chance at life and the father signs the consent form for treatment. They agree with the oncologist's recommendation for a clinical trial for ALL. This trial is showing much promise for a cure for Lily's disease.

Lily's nurse uses distraction techniques and takes time to build trust before beginning interventions. Initially, she reads to Lily to calm her down and initiate a relationship, later she takes the little girl's vital signs without a struggle. Soon after, she is able to insert peripheral intravenous lines to initiate maintenance hydration by distracting Lily with puppets. Red blood cells were ordered for anemia and a platelet transfusion was given to increase her platelet count and prevent bleeding. Lily is monitored closely for any deterioration in vital signs.

**Staff can assist Lily in coping with the many invasive procedures she will experience by employing age appropriate distraction techniques such as scheduling play therapy with a child life specialist, providing age appropriate toys and books, and encouraging parental participation in care.**

### **Case Study One: Treatment**

Lily receives her first dose of intravenous chemotherapy that afternoon. She is irritable and nauseated, crying and clinging to her mother. Multiple visitors arrive; grandparents, aunts and uncles, cousins, and neighbors bringing balloons and toys. The many visitors excite Lily and tire her parents. The phone rings non-stop. Lily's father comments to the nurse, *"I wish I could tell her visitors to stay home. Could they bring in germs that could hurt her? She hasn't been able to take a nap. I am not sure what to do."* He looks overwhelmed and exhausted.

### **Case Study One: Controlling the Environment**

How can the nurse and team help the family gain control over the visitors and phone calls? Lily's parents talk with the nurse about options for controlling their hospital environment. Together they come up with a workable plan, appointing Lily's paternal grandparents to be the contacts for family and friends. Her grandparents let the relatives and friends know Lily's status and advise them on visitor restrictions. Adults without contagious illnesses may visit at the parent's request. Children are requested to stay home and the phone ringer is turned off in the room. Lily's nurse medicates her with an antiemetic medicine and the child and family get some much needed sleep. A sign is attached to the hospital room door directing visitors to the nurse's station before entering.

### **Case Study One: Supportive Care**

Children and adolescents are comforted by close physical proximity to their significant caregivers, and every effort should be made to include the family into the child's plan of care. Family centered care is a favorable goal that will help the child and family to cope effectively with the diagnosis.

Lily is now three weeks post initial chemotherapy and is experiencing neutropenia (a decreased white blood cell count and low neutrophils), a fever of 103 degrees, mouth sores, oral pain, mucositis, decreased platelet counts, and a lack of appetite.

Lily's supportive care includes:

1. Three broad spectrum intravenous antibiotics.
2. Anti-fungal mouth care and oral regime for mouth sores.
3. Platelet transfusions for a platelet count less than 50,000.
4. Pain control, a mild narcotic based pain medication is administered for oral pain.

With her pain controlled, Lily starts to take fluids and eat solids and is able to take her medicine. She is discharged three days later as her temperature returns to normal and her white count increases.

### **Case Study One: Prognosis and Recovery**

Although Lily's diagnosis and cell type (ALL) is low risk and her prognosis is favorable, the treatment is aggressive and will require support and understanding from the healthcare team.

A follow-up bone marrow test at the outpatient clinic reveals Lily's marrow to be free of leukemia cells. She is responding to treatment. Lily has achieved remission. She has no signs or symptoms of leukemia. Treatment is completed.

Lily's parents are happy, hopeful, yet fearful. Will the leukemia come back? These parental feelings are normal. Lily will need frequent follow-up with the Hematology team in the years ahead to watch for relapse.

Lily has a 90% chance of staying cancer free. Lily doesn't understand the details but she is just happy, feels well, and calls out to the staff as she leaves the clinic, *"No sticks today! Bye-Bye!"* Good-bye Lily!

### **Case Study Two**

Jaycee, a fifteen year old high school football player is brought to his family doctor for complaints of a lemon size painless *"lump on his neck."* Jaycee and his mother describe the lump as appearing quickly *"in the last two weeks"*. The nurse practitioner takes a careful medical and family history. Jaycee describes a weight loss of fifteen pounds, night sweats, and feeling *"tired"* for the past month. He has a cough and inspiratory wheeze. He claims he did not want to bother his parents and he hoped to finish the football season and participate in the state playoff games. He is sent to the local children's hospital emergency room where a chest x-ray and CT scan reveal a large mediastinal mass surrounding his heart. A lymphoma is suspected.

### **Case Study Two: Admission to the Oncology Unit**

Jaycee is admitted to the oncology unit. A biopsy of his enlarged cervical lymph nodes is planned. The coach and sixteen of Jaycee's fellow football team members arrive on the Hematology Oncology Unit crowding into his room to offer their support. Jaycee assures them he is fine and will be back playing football next week. His parents retreat to the visitor waiting room exhausted and overwhelmed. The nursing staff starts to orient the patient and family to the unit. They watch Jaycee carefully for signs of cardiac and respiratory distress. The hematologist is concerned about the large chest mass. Jaycee is placed on cardiac monitoring.

### **Test Your Knowledge**

How can the staff help support Jaycee and his parents during the initial diagnosis stage?

***The correct answer is: Provide educational information. Offer physical comforts such as coffee, blankets, and a quiet place to rest. Explore the needs of the family and determine how the parents feel they can best be supported.***

### **Case Study Two: Risk Factors and Symptoms**

Jaycee's risk factors for lymphoma include his age and his gender. He is male and a fifteen year old adolescent. Jaycee has presented with the most common early sign of Hodgkin lymphoma, a painless swelling of the lymph nodes. He has also reported a history of weight loss and night sweats. Jaycee admits to the medical staff during the patient history that he had felt *"sick"* for several weeks, and felt he might have *"mono."* Jaycee delayed seeking treatment so he could continue to play football. The size of the lymph node in his neck caused him to show the swollen area to his mother.

Jaycee's mother immediately made an appointment with their family doctor. As Jaycee awaits his lymph node biopsy in the morning he asks his mother, *"When can I play football? Can antibiotics get rid of the lump in my neck?"*

How can the healthcare team help Jaycee cope with his diagnostic tests and diagnosis? Would age appropriate educational materials dealing with lymphoma help? Let him talk to another teen patient with cancer? Tell him he cannot play football? Assess Jaycee's perception of why he is in the hospital?

### **Case Study Two: Treatment and Support**

Jaycee undergoes a biopsy of the cervical lymph node as well as a bone marrow biopsy and spinal tap. A review of the slides from the biopsy reveals Reed-Sternberg cells consistent with Hodgkin Lymphoma. Jaycee undergoes additional diagnostic imaging and is diagnosed with Stage III Hodgkin Disease.

Jaycee receives chemotherapy and radiation treatment on an outpatient basis. He returns via ambulance to the emergency department (ED) complaining of chest pain, rapid heart rate, and difficulty breathing. Jaycee's parents are distraught asking, *"What is happening?"* As Jaycee is rolled into the trauma room he exclaims; *"Am I dying? Help me!"* The ED staff quickly perform a complete physical assessment. After determining Jaycee's vital signs and oxygen saturations are within normal limits, chest x-rays and CT scans are taken.

### **Case Study Two: The Psychosomatic Impact of Cancer**

All of Jaycee's imaging reports are normal and his lymphoma is noted to be shrinking in his chest. The doctors diagnose him with anxiety and a panic attack. The hospital psychologist comes and talks to Jaycee. They set up follow up appointments to discuss the stressors affecting cancer patients and effective coping strategies such as deep breathing, positive imagery, and meditation. Jaycee states he feels better now and the chest pain is gone.

### **Case Study Two: Remission**

The lymphoma is in complete remission. He does not like to talk about his treatment; however he volunteers as a camp counselor at Oncology Camp and wants to help other children diagnosed with cancer. His parents are relieved and cautiously optimistic. Jaycee will follow-up yearly with the long term follow-up team. He has a good chance of maintaining his remission and be cured of lymphoma. Jaycee is graduating from high school and going to college in the fall. Jaycee is disease free. *"See you next year"* he calls out to the staff.

### **Conclusion**

The outlook for children and adolescents diagnosed with childhood leukemia and lymphoma has never looked brighter. Modern medical advances in cancer research, treatment, and long term follow up has improved survival rates dramatically. With appropriate diagnosis and care from the healthcare provider, the majority of pediatric patients presenting with leukemia and/or lymphoma will achieve long term remissions.

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