LEUKEMIA & LYMPHOMA SOCIETY®
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The ALL Guide

Information for Patients and Caregivers

Acute Lymphoblastic Leukemia

Christopher, ALL survivor

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Publication Update

The ALL Guide
Information for Patients and Caregivers

The Leukemia & Lymphoma Society wants you to have the most up-to-date information about blood cancer treatment. See below for important new information that was not available at the time this publication was printed.

- In August 2017, tisagenlecleucel (Kymriah®) was FDA approved treatment of patients up to 25 years of age with B-cell precursor acute lymphoblastic leukemia (ALL) that is refractory or in second or later relapse. Tisagenlecleucel is a CD19-directed genetically modified autologous T cell immunotherapy.

- In August 2017, the Food and Drug Administration (FDA) approved inotuzumab ozogamicin (Besponsa™) for the treatment of adults with relapsed or refractory B-cell precursor acute lymphoblastic leukemia (ALL).

- In April 2017, the Food and Drug Administration (FDA) approved methotrexate (Xatmep™) for the treatment of pediatric patients with acute lymphoblastic leukemia (ALL) as a component of a combination chemotherapy maintenance regimen.

For more information, contact an Information Specialist at (800) 955-4572 or infocenter@lls.org.

A Message from Louis J. DeGennaro, PhD

President and CEO of The Leukemia & Lymphoma Society

The Leukemia & Lymphoma Society (LLS) is the world's largest voluntary health organization dedicated to finding cures for blood cancer patients. Since 1954, we have invested more than $1 billion in research specifically targeting blood cancers to advance therapies and save lives. We will continue to invest in research for cures, programs and services to improve the quality of life for people with acute lymphoblastic leukemia (ALL).

We know that understanding ALL can be tough.

We are here to help and are committed to providing you with the most up-to-date information about ALL, your treatment and your support options. We know how important it is for you to understand your health information. We want you to be able to use that information, along with the help of members of your healthcare team, on your pathway toward good health, remission and recovery.

Our vision is that one day all people with ALL will be cured or be able to manage their disease and have a good quality of life.

Until then, we trust the information in this *Guide* will help you along your journey.

We wish you well.

Louis J. DeGennaro, PhD
President and Chief Executive Officer
The Leukemia & Lymphoma Society

Inside This Guide

This LLS Guide about ALL is for information only. LLS does not give medical advice or provide medical services.

Introduction

Acute lymphoblastic leukemia (ALL) is a type of blood cancer. Other names for ALL are "acute lymphocytic leukemia" and "acute lymphoid leukemia." ALL is the most common type of leukemia in children.

Advances in the treatment of ALL have resulted in improved remission rates. The number of patients who have gone into remission or have been cured increases each year.

- About 6,590 people in the United States were expected to be diagnosed with ALL in 2016.
- About 71,898 people in the United States are living with or are in remission from ALL as of 2012.

Want more information?

You can view, print or order the free LLS booklet *Acute Lymphoblastic Leukemia* at www.LLS.org/booklets. Or, contact our Information Specialists for a copy.

Resources and Information

LLS offers free information and services for patients and families affected by blood cancers. This section of the *Guide* lists various resources available to you. Use this information to learn more, to ask questions and to make the most of your healthcare team.

For Help and Information

Consult with an Information Specialist. Information Specialists are master's level oncology social workers, nurses and health educators.

They offer up-to-date information about disease, treatment and support. Language services are available. For more information, please

- Call: (800) 955-4572 (Monday through Friday, 9 a.m. to 9 p.m. ET)
- Email: infocenter@LLS.org
- Live chat: www.LLS.org/informationspecialists.

Free Information Booklets. LLS offers free education and support booklets that can either be read online or ordered. For more information, please visit www.LLS.org/booklets.

Telephone/Web Education Programs. LLS offers free telephone/Web education programs for patients, caregivers and healthcare professionals. For more information, please visit www.LLS.org/programs.

Sign Up for an e-Newsletter. Read the latest disease-specific news, learn about research studies and clinical trials, and find support for living with blood cancer. Please visit www.LLS.org/signup.

Community Resources and Networking

Online Blood Cancer Discussion Boards and Chats. Online discussion boards and moderated online chats can help cancer patients reach out, share information and provide and receive support. For more information, please visit www.LLS.org/discussionboard and www.LLS.org/chat.

LLS Community. LLS Community is an online social network and registry for patients, caregivers, and supporters of those with blood cancer. It is a place to ask questions, get informed, share your experience, and connect with others. To join visit www.LLS.org/community.

LLS Chapters. LLS offers community support and services in the United States and Canada including the *Patti Robinson Kaufmann First Connection Program* (a peer-to-peer support program), in-person support groups and other great resources. For more information about these programs or to contact your chapter, please

- Call: (800) 955-4572
- Visit: www.LLS.org/chapterfind.

Other Helpful Organizations. LLS offers an extensive list of resources for patients and families. There are resources that provide help with financial assistance, counseling, transportation, locating summer camps and other needs. For more information, please visit www.LLS.org/resourcedirectory.

Clinical Trials (Research Studies). New treatments for patients are under way. Many are part of clinical trials. Patients can learn about clinical trials and how to access them. For more information, please call (800) 955-4572 to speak with an LLS Information Specialist who can help conduct a clinical-trial search.

Advocacy. The LLS Office of Public Policy (OPP) engages volunteers in advocating for policies and laws that encourage the development of new treatments and improve access to quality medical care. For more information, please

- Call: (800) 955-4572
- Visit: www.LLS.org/advocacy.

Additional Help for Specific Populations

Información en Español (LLS information in Spanish). For more information, please visit www.LLS.org/espanol.

Language Services. Let your doctor know if you need a language interpreter or other resource, such as a sign language interpreter. Often, these services are free.

Children's Concerns. Parents of a child with ALL may want to talk to members of their child's healthcare team about how to find enough time for everything, pay for treatment and best help their children. For more information, see the free LLS booklet *Coping With Childhood Leukemia and Lymphoma.*

The Trish Greene Back to School Program for Children With Cancer. This program helps doctors, nurses, parents and school personnel work together for a smooth return to school for children with cancer. For more information, contact your LLS chapter or call (800) 955-4572.

World Trade Center Survivors. People involved in the aftermath of the 9/11 attacks and subsequently diagnosed with a blood cancer may be able to get help from the World Trade Center (WTC) Health Program. People eligible for help include

- Responders
- Workers and volunteers who helped with rescue, recovery and cleanup at the WTC-related sites in New York City (NYC)
- Survivors who were in the NYC disaster area and those who lived, worked or were in school in that area
- Responders to the Pentagon and the Shanksville, PA crashes.

For more information, please

- Call: WTC Health Program at (888) 982-4748
- Visit: www.cdc.gov/wtc/faq.html.

Depression. Treating depression has benefits for cancer patients. Seek medical advice if your mood does not improve over time, for example, if you feel depressed every day for a two-week period. For more information, please

- Call: National Institute of Mental Health (NIMH) at (866) 615-6464
- Visit: NIMH at www.nimh.nih.gov, enter "depression" in the search box.

Feedback. To provide your opinion of this *Guide* please visit: www.LLS.org/publicationfeedback.

Part 1

Understanding ALL

Leukemia is the general term for a number of different types of blood cancer. ALL is one of four main types of leukemia.

About Marrow, Blood and Blood Cells

The general descriptions below may help you understand the information in the rest of this *Guide*.

Marrow is the spongy center inside of bones where blood cells are made.

Blood cells are made in the bone marrow. They begin as stem cells. Stem cells become red blood cells, white blood cells and platelets in the marrow. Then the red blood cells, white blood cells and platelets enter the blood.

Platelets form plugs that help stop bleeding at the site of an injury.

Red blood cells carry oxygen around the body. When the number of red blood cells is below normal, the condition is called **anemia**. Anemia may make you feel tired or short of breath. It may make the skin look pale.

White blood cells fight infection in the body. There are two major types of white blood cells: germ-eating cells (neutrophils and monocytes) and infection-fighting **lymphocytes** (B cells, T cells and natural killer [NK] cells).

Plasma is the liquid part of the blood. It is mostly water. It also has some vitamins, minerals, proteins, hormones and other natural chemicals in it.

Normal Blood Cell Count Fast Facts

The ranges of blood cell counts below are for adults. They may be a little different from lab to lab and for children and teens.

Red blood cell (RBC) count

- Men: 4.5 to 6 million red cells per microliter of blood
- Women: 4 to 5 million red cells per microliter of blood

Hematocrit (the part of the blood made up of red cells)

- Men: 42% to 50%
- Women: 36% to 45%

Hemoglobin (amount of the red-cell pigment that carries oxygen)

- Men: 14 to 17 grams per 100 milliliters of blood
- Women: 12 to 15 grams per 100 milliliters of blood

Platelet count

- 150,000 to 450,000 platelets per microliter of blood

White blood cell (WBC) count

- 4,500 to 11,000 white cells per microliter of blood

Differential (also called **diff**)

- Shows the part of the blood made up of different types of white cells.
- The types of white cells counted are neutrophils, lymphocytes, monocytes, eosinophils and basophils.
- Adults usually have about 60% neutrophils, 30% lymphocytes, 5% monocytes, 4% eosinophils and less than 1% basophils in their blood.

About ALL

ALL is a type of cancer that begins in the bone marrow. A person can get ALL at any age. The overall risk of getting ALL is small for children and adults. However, ALL remains the most common type of cancer in children under age 20.

Causes of ALL. ALL starts with a change to a single cell in the bone marrow. Doctors do not know what causes most cases of ALL. There is no way to prevent ALL. You can't catch ALL from someone else.

Signs and Symptoms. Many ALL signs and symptoms are also caused by other types of illness. Most people with these signs and symptoms do not have ALL.

A sign is a change in the body that the doctor sees in an exam or a test result.

A symptom is a change in the body that the patient can see or feel.

A person with ALL may have

- Aches in legs, arms or hips
- Black-and-blue marks for no clear reason
- Enlarged lymph nodes
- Fever without obvious cause
- Pale-looking skin
- Pinhead-sized red spots under the skin called **petechiae** (peh tee' key uh)
- Prolonged bleeding from minor cuts
- Shortness of breath during physical activity
- Tiredness or no energy
- Vomiting
- Unexplained weight loss.

Diagnosis

It is important for patients to receive the right diagnosis. ALL is diagnosed with blood and bone marrow tests.

Blood Cell Counts. The doctor orders a test called a **CBC (complete blood count)**. This test counts the numbers of red blood cells, white blood cells and platelets. Usually, patients with ALL have lower-than-expected red blood cells and platelets.

Blood Cell Examination. The cells are stained (dyed) and looked at through a microscope. This test is called a **blood smear**. A person with ALL usually has too many leukemic blast cells in the blood. Blast cells are immature (young) cells that do not function like normal mature cells. The blood smear sample can also be used for cytogenetic analysis and immunophenotyping, explained below.

- Cytogenetic analysis — Each cell in the body has chromosomes that carry genes. Genes give the instructions that tell each cell what to do. The test called **cytogenetic analysis** examines the chromosomes of the ALL blast cells.
- Immunophenotyping — The test called **immunophenotyping** is used to identify cells based on the types of proteins on the cell surface to find out if the ALL cells are B cells or T cells. Most people have B-cell ALL.

Bone Marrow Tests. Other tests will be done to make sure the diagnosis of ALL is correct. Tests called **bone marrow aspiration** and **bone marrow biopsy** are done. These tests take a close look at the ALL cells. They also find out the percentage of ALL cells in the bone marrow.

The doctor uses information from all of the tests to decide

- The type of drug therapy a patient needs
- How long treatment will last.

The type and length of treatment may also depend on the patient's age.

How are the Blood and Bone Marrow Tests Done?

Blood Test — A small amount of blood is taken from the patient's arm with a needle. The blood is collected in tubes and sent to a lab.

Bone Marrow Aspiration — A sample of cells is taken from the marrow.

Bone Marrow Biopsy — A very small amount of bone filled with marrow cells is taken from the body.

Both bone marrow tests are done with a special needle. Some patients are awake for the procedure. They get medication first to numb the part of the body that will be used to get the sample of cells. Some patients are given a drug that makes them sleep during this procedure. The sample of cells is usually taken from the patient's hip bone.

Blood and marrow tests may be done in the doctor's office or in a hospital. A bone marrow aspiration and biopsy are almost always done at the same visit.

Blood and marrow tests are also done during and after treatment. The tests are repeated to see if treatment is destroying the ALL cells.

Want more information?

You can view, print or order the free LLS booklet *Understanding Lab and Imaging Tests* at www.LLS.org/booklets. Or, contact our Information Specialists for a copy.

Part 2

Treatment

Finding the Right Doctor

Choose a doctor who specializes in treating ALL. He or she should know about the most up-to-date treatments. This type of specialist is usually called a **hematologist-oncologist**. Or, your local cancer specialist can work with a leukemia specialist.

Ways to Find an ALL Specialist

- Ask your primary care doctor.
- Contact your community cancer center.
- Reach out to doctor and/or health plan referral services.
- Call our Information Specialists at 800.955.4572.
- Use online doctor-finder resources, such as
  - The American Medical Association's (AMA) "DoctorFinder."
  - The American Society of Hematology's (ASH) "Find a Hematologist."

Want more information?

You can view, print or order the free LLS fact sheet *Choosing a Blood Cancer Specialist or Treatment Center* at www.LLS.org/booklets. Or, contact our Information Specialists for a copy.

Ask Your Doctor

Talk with the doctor and ask questions about how he or she plans to treat your ALL. This will help you to

- Be actively involved in your care
- Make decisions.

This *Guide* includes questions to ask your doctor about ALL treatment (see pages 38-40).

It may be helpful to write down the answers to your questions and review them later. You may want to have a caregiver, family member or friend with you when you talk to your doctor. This person can listen, take notes and offer support. Some people like to audio record information from the doctor and then listen to the recording later on.

People with ALL who are unsure about their treatment options are encouraged to get a second opinion.

Want more information?

For a list of Healthcare Question Guides about second opinions and other topics that you can print, go to www.LLS.org/whattoask. Or, contact our Information Specialists for copies.

Treatment Planning

There are three stages of treatment for ALL

- Induction therapy
- Consolidation therapy (also called **intensification**)
- Maintenance therapy.

Consolidation and maintenance are post-remission therapies. These are described beginning on page 19.

The goal of treatment for ALL is to cure the disease.

- Many children with ALL are cured.
- The number of adults with ALL in remission (no sign of the disease after treatment) has increased.
- The length of remission in adults has improved.

Many children with ALL are treated in clinical trials. Adults with ALL should talk to their doctors about clinical trials. A clinical trial may be a good treatment choice for you or your child. Clinical trials are explained beginning on page 25.

Some things that may affect the outcome of your ALL treatment are

- Your subtype of ALL
- The results of your lab tests
- Your age and general health
- Your medical history, including whether you were treated before with chemotherapy
- Whether you have
  - A serious infection at the time of diagnosis
  - ALL in your central nervous system
  - ALL that has not responded to treatment or has relapsed.

The bone marrow examination gives the doctor important information for treatment planning.

Pretreatment Considerations. Infertility is a concern for adults of childbearing age and parents of children diagnosed with ALL. Ask the doctor for information that may lessen the risk for infertility.

Want more information?

You can view, print or order the free LLS fact sheet *Fertility Facts* at www.LLS.org/booklets. Or, contact our Information Specialists for a copy.

About ALL Treatments

Treatment for patients with ALL (or relapsed ALL) may include

- Chemotherapy
- Stem cell transplantation
- New approaches under study (called **clinical trials**).

A patient may receive different drugs from those described in this *Guide*. This may still be considered proper treatment.

Speak to your doctor to find out what treatment is best for you.

Our Information Specialists can help you plan questions to ask your doctor about treatment.

Induction Therapy. Induction therapy is the first round of treatment with chemotherapy. Most ALL patients need to start induction chemotherapy right away. Induction therapy is done in the hospital.

Patients are often in the hospital for four to six weeks.

The aims of induction therapy are to

- Kill as many ALL cells as possible
- Get blood counts back to normal
- Get rid of all signs of the disease for an extended period of time.

Names of Some Induction Therapy Drugs for ALL

- Daunorubicin (Cerubidine®) or doxorubicin (Adriamycin®) by central line, port or PICC
- Asparaginase *Erwinia chrysanthemi* (Erwinaze®) or pegaspargase (PEG-L-asparaginase; Oncaspar®) by injection
- Vincristine (Oncovin®) by central line, port or PICC
- Dexamethasone or prednisone (corticosteroids) by mouth
- Methotrexate by injection into the spinal fluid
- Cytarabine (cytosine arabinoside, ara-C, Cytosar-U®) by injection into the spinal fluid
- 6-mercaptopurine (6-MP) (Purinethol® given by mouth as a tablet and Purixan® given by mouth as a liquid)

Chemotherapy and other drugs. Chemotherapy and some other types of drugs kill or damage cancer cells. Several types of drugs are used to kill ALL cells. Each drug type works in a different way. Combining the drugs can make the treatment work better.

The first round of chemotherapy usually does not get rid of all the ALL cells. Most patients will need more rounds of treatment. Usually the same drugs are used for the added rounds.

Some drugs are given by mouth. Other drugs are given through a central line, port or PICC. Central lines, ports or PICCs can be used to give medications, nutrition and blood cells. They can be used to take blood samples, too. Central lines, ports and PICCs can stay in place for weeks or months. Talk to your doctor about the best one for you or your child to use.

Central Lines, Ports and PICCs

Central Line — A thin tube that is put under the skin and into a large vein in the chest. The central line stays firmly in place. "Catheter" is another word for central line.

Port — This is a small device that is used with a central line. The port is placed under the skin of the chest. After the site heals, no dressings and no special home care are needed. The doctor or nurse can give medicines or nutrition or take blood samples. He or she puts a needle through the skin into the port. A numbing cream can be put on the skin before the port is used.

PICC or PIC Line (short for "percutaneously inserted central venous catheter") — A PICC is inserted through a vein in the arm.

Some ALL Drugs

These are some of the standard ALL drugs used.
Some drugs under study in ALL clinical trials are also listed.

Drug Types	Drug Names
Antimetabolites	clofarabine (Clolar®), cytarabine (cytosine arabinoside; ara-C, Cytosar-U®), 6-mercaptopurine (Purinethol® or Purixan®), methotrexate, 6-thioguanine (Tabloid®), nelarabine (Arranon®)
Antitumor Antibiotics	daunorubicin (Cerubidine®), doxorubicin (Adriamycin®), mitoxantrone (Novantrone®), idarubicin (Idamycin®)
DNA Repair Enzyme Inhibitor	etoposide (VP-16; VePesid®)
DNA-Damaging Agent	cyclophosphamide (Cytoxan®)
Drugs That Prevent Cells from Dividing	vincristine (Oncovin®) liposomal vincristine (Marqibo®)
Enzymes That Prevent Cells From Surviving	asparaginase *Erwinia chrysanthemi* (Erwinaze®), pegaspargase (PEG-L-asparaginase; Oncaspar®)
Tyrosine Kinase Inhibitors	imatinib mesylate (Gleevec®), dasatinib (Sprycel®), nilotinib (Tasigna®), ponatinib (Iclusig®)
Corticosteroids	prednisone, prednisolone, dexamethasone
Immunotherapies	alemtuzumab (Campath®) rituximab (Rituxan®), blinatumomab (Blincyto®), inotuzumab ozogamicin, chimeric antigen receptor (CAR) T-cell therapy

Information about side effects begins on page 26.

Want more information?

You can view, print or order the free LLS booklet *Understanding Side Effects of Drug Therapy* at www.LLS.org/booklets. Or, contact our Information Specialists for a copy.

Central Nervous System (CNS) Leukemia

Patients may have ALL cells in the lining of the spinal cord and brain. A spinal tap (also called a **lumbar puncture**) is used to check the spinal fluid for ALL cells. Every ALL patient gets treatment to prevent CNS leukemia. Even if ALL cells are not found in the spinal fluid, patients are still treated.

The spinal cord and brain are hard to reach with chemotherapy that is given through a vein in the arm. So, spinal fluid is removed and chemotherapy is injected right into the spinal canal. Drugs, such as methotrexate, are used.

Radiation therapy may be given to the spine or brain. Sometimes both chemotherapy and radiation therapy are used. In some practices, doctors are not using cranial radiation for pediatric patients except in patients who have CNS leukemia or CNS relapse. When radiation therapy is not used, the chance of long-term effects decreases.

Spinal taps are done from time to time throughout treatment. This test checks to see if the ALL cells are being killed.

Post-Remission Therapy

When there is no sign of ALL, this is called a **remission**. Unfortunately, more treatment is usually needed even after an ALL patient is in remission. Some ALL cells may remain. These are not found by common blood or marrow tests. This part of ALL treatment is called **post-remission therapy**. (Post-remission means after remission.) Chemotherapy is used in post-remission therapy for ALL. Stem cell transplantation may be part of post-remission therapy for some ALL patients. Post-remission therapy consists of consolidation (intensification) therapy and maintenance therapy.

- Consolidation therapy is usually given in cycles for four to six months.
- Maintenance therapy is usually given for about two years.

For most people, the drugs used during consolidation and maintenance therapy are different than the drugs used during induction therapy.

The doctor considers many things when deciding a treatment plan. This includes ALL subtype, doses and length of consolidation and maintenance therapy. The doctor also asks

- Did the induction therapy kill the ALL cells?
- Does the patient have changes to the chromosomes of the ALL cells?

Some types of ALL are usually treated with higher doses of drugs during induction, consolidation and maintenance therapy. These types include T-cell ALL, ALL in infants and ALL in adults.

An allogeneic stem cell transplant may be a good treatment for some ALL patients. Allogeneic transplants are explained beginning on page 22.

Some Consolidation and Maintenance Therapies

- Vincristine (Oncovin®) by intravenous (IV) infusion
- Prednisone or dexamethasone by mouth
- Mercaptopurine (Purinethol®) by mouth
- Methotrexate by mouth, IV or injection into the spinal fluid
- Cytarabine (cytosine arabinoside, ara-C, Cytosar-U®) by IV or injection into the spinal fluid
- Asparaginase *Erwinia chrysanthemi* (Erwinaze®) or PEG-L-asparaginase (Oncaspar®) by injection
- Radiation therapy to the head and sometimes to the spine

Ph-Positive ALL

About one out of four adults with ALL has a type called **Ph-positive ALL**. A small number of children (two to four out of 100 children) with ALL have Ph-positive ALL.

Induction Therapy. Ph-positive ALL may be treated with imatinib mesylate (Gleevec®), dasatinib (Sprycel®), nilotinib (Tasigna®) or ponatinib (Iclusig®). Other chemotherapy drugs will be used as well. Gleevec, Sprycel, Tasigna and Iclusig are given by mouth.

Post-Remission Therapy. During consolidation and maintenance therapy, Gleevec, Sprycel, Tasigna or Iclusig is given with other drugs. Usually people with Ph-positive ALL stay on Gleevec, Sprycel, Tasigna or Iclusig after maintenance chemotherapy is completed.

Relapsed and Refractory ALL

Most children with ALL are cured of their disease. Some patients (children or adults) have a remission after treatment, but then ALL cells return later (called **relapsed ALL**). Patients may have ALL cells in the marrow even after treatment (called **refractory ALL**).

Patients with relapsed ALL may be

- Treated with the same drugs as newly diagnosed patients
- Given different drugs
- Given an allogeneic stem cell transplant if they have a matched donor. Information about allogeneic transplantation begins on page 22.

For patients with refractory ALL

- Drugs that were not used in the first round of treatment may be given
- An allogeneic stem cell transplant may also be a part of treatment.

For children with relapsed or refractory ALL

- The drug clofarabine (Clolar®) is approved by the FDA to treat children (from age 1 to 21) if the usual treatments do not work.
- Treatment with Clolar followed by an allogeneic transplant may result in a cure.

The FDA has approved these drugs for relapsed and refractory patients.

- Liposomal vincristine (Marqibo®) is approved for adult patients with Ph chromosome-negative ALL who have relapsed two or more times. It is also approved for patients whose leukemia has progressed following two or more regimens of therapy.
- Nelarabine (Arranon®) is approved for patients with relapsed T-cell ALL.
- Blinatumomab (Blincyto®) is approved to treat patients with Ph chromosome-negative precursor B-cell ALL for patients whose cancer relapsed or was refractory.

Stem Cell Transplantation

Your doctor will talk with you about whether stem cell transplantation is a treatment option for you.

Allogeneic Stem Cell Transplantation. An allogeneic transplant uses stem cells from a donor. The donor may be a brother or sister. Or the donor can be an unrelated person. This person's stem cells "match" the patient's. Stem cells may also come from a cord blood unit (the blood in the umbilical cord after a baby's birth). The goals of an allogeneic transplant are to

- Restore the body's ability to make normal blood cells after high-dose chemotherapy
- Cure the patient of his or her ALL by killing any remaining ALL cells.

Allogeneic transplants may be done in the hospital. First, the patient is given high-dose chemotherapy and/or radiation therapy. Stem cells are collected from a donor. The donor stem cells are given to the patient through an intravenous (IV) line or central line. The donor stem cells go from the patient's blood to the bone marrow. They help start a new supply of red blood cells, white blood cells and platelets.

An allogeneic transplant is a high-risk procedure. Doctors are working to make allogeneic transplants safer. An allogeneic transplant may be a choice for an adult ALL patient if

- He or she is not doing well with other treatments
- The expected benefits of an allogeneic transplant exceed the risks
- There is a stem cell donor.

The upper age limit for an allogeneic transplant depends on the treatment center.

A stem cell transplant is usually not considered for a child unless

- Doctors have determined that the child's type of ALL is not likely to respond well to chemotherapy

- Chemotherapy has not worked well
- The ALL has returned (relapsed).

A reduced-intensity allogeneic transplant uses lower doses of chemotherapy than a standard allogeneic transplant. Some older and sicker patients may be helped by this treatment. Many centers use 70 years as the upper age limit for a reduced-intensity allogeneic transplant. Your doctor will talk to you about whether a reduced-intensity allogeneic transplant is a treatment option for you.

A reduced-intensity allogeneic transplant may be a choice for an ALL patient if

- He or she is not doing well with other treatments
- A standard allogeneic transplant is not a choice because of the patient's age or overall health
- The expected benefits of a reduced-intensity allogeneic transplant exceed the risks
- There is a stem cell donor.

Autologous Stem Cell Transplantation. An autologous transplant uses the patient's own stem cells.

Your doctor will talk with you about whether an autologous transplant is a treatment option for you. It is not commonly used to treat ALL. This is because of the high relapse rate following this type of transplant.

Want more information?

You can view, print or order the free LLS booklets *Blood and Marrow Stem Cell Transplantation* and *Cord Blood Stem Cell Transplantation Facts* at www.LLS.org/booklets. Or, contact our Information Specialists for copies.

ALL Treatment in Young Adults

Older adolescents and adults younger than 40 are often called **young adults**. Traditionally, treatment for this group has been similar to adult treatment protocols. However, a number of cancer centers are using pediatric protocols to treat young adult patients. Speak to your doctor or call an Information Specialist about the different clinical trials that may be available to you.

ALL Treatment in Adults

Today, cures are possible for some adults with ALL. These include those who may have other serious health problems. But treatment results in adults are not as good as treatment results in children. ALL in adults is more resistant to treatment than ALL in children. New and better treatments for ALL in adults are needed.

A number of cancer centers are using pediatric protocols to treat younger adult patients. Speak to your doctor or call an Information Specialist about the different clinical trials that may be available to you.

Part 3

About Clinical Trials

There are new treatments under study for ALL patients of all ages.

New treatments are studied in clinical trials. Clinical trials are also used to study new uses for approved drugs or treatments. For example, changing the amount of the drug might be more effective. Or, giving the drug along with another type of treatment might work better. Some clinical trials combine drugs for ALL in new sequences or dosages.

There are clinical trials for

- Newly diagnosed ALL patients
- Patients who do not have a good response to treatment
- Patients who relapse after treatment.

A carefully conducted clinical trial may provide the best available therapy.

Ask your doctor if treatment in a clinical trial is right for you. Some drugs being studied in clinical trials are listed in the chart on page 17. You can call our Information Specialists for information about clinical trials.

Want more information?

You can view, print or order the free LLS booklets *Understanding Clinical Trials for Blood Cancers* and *Knowing All Your Treatment Options* at www.LLS.org/booklets. Or, contact our Information Specialists for copies.

Part 4

Side Effects and Follow-Up Care

Side Effects of ALL Treatment

The term side effect describes the way that treatment affects healthy cells.

The aim of treatment for ALL is to kill the ALL cells. Treatment for ALL also affects healthy cells. Side effects of ALL treatment may be severe, but they usually go away once treatment ends. Ask your doctor about the side effects to expect from your treatment.

ALL treatment may affect your blood cell counts.

- The number of red blood cells may decrease (anemia). Red blood cell transfusions (red blood cells that are donated and given to the patient) may be needed to increase red blood cell counts.
- Patients may also have a drop in the number of platelets. A platelet transfusion may be needed to prevent bleeding if a patient's platelet count is very low.
- A big drop in the number of white blood cells may lead to an infection. These infections are usually treated with antibiotics.

Patients at home should not delay in seeking medical attention if any signs of infection develop. A rise in temperature to 101°F or higher or the onset of chills may be the only sign of infection in a patient with a very low white blood cell count. Patients with an infection may also have

- Coughing
- Sore throat
- Pain when urinating
- Frequent loose bowel movements.

Growth factors are sometimes given to increase white blood cells. G-CSF (Neupogen® and Neulasta®) and GM-CSF (Leukine®) are drugs that increase the number of white blood cells.

Growth factors are only given to children in certain cases. Scientists are studying which children with ALL are most likely to be helped by treatment with growth factors to prevent infection.

To lower the risk of infection

- The patient, the patient's visitors and the medical staff need to wash their hands often and well.
- The patient's central line must be kept clean.
- Patients should take good care of their teeth and gums.

The doctor may talk about the absolute neutrophil count, or ANC. This is the number of neutrophils (a type of white blood cell) a person has in his or her body to fight an infection.

Other Treatment Side Effects. Chemotherapy affects the parts of the body where new cells form quickly. This includes the inside of the mouth and bowel, and the skin and hair. The side effects listed here are common during chemotherapy

- Mouth sores
- Diarrhea
- Hair loss
- Rashes
- Nausea
- Headache
- Vomiting.

Not all patients have these side effects. Treatment to prevent or manage nausea, vomiting, diarrhea and other side effects can help patients feel more comfortable.

Chemotherapy may cause the amount of uric acid to increase in the blood of some ALL patients. Some patients also have a buildup of uric acid from the disease itself. Uric acid is a chemical made in the body. A high level of uric acid can cause kidney stones.

Patients with high uric acid levels may be given

- A drug called allopurinol (Aloprim®, Zyloprim®) by mouth
- A drug used called rasburicase (Elitek®), which is given by IV.

Information about long-term and late effects of treatment begins below.

Want more information?

You can view, print or order the free LLS booklet *Understanding Side Effects of Blood Therapy* at www.LLS.org/booklets. Or, contact our Information Specialists for a copy.

Long-Term and Late Effects

Long-term effects are medical problems that last for months or years after treatment ends. Fatigue is an example. Late effects are medical problems that do not show up until years after treatment ends. Heart disease is an example.

Not everyone who is treated for ALL develops long-term or late effects. It depends on the patient's age, overall health and treatment.

Children who are treated for ALL may have

- Growth problems
- Fertility problems (ability to have children later on)
- Bone problems

- Heart problems
- Learning problems.

Adults who are treated for ALL may have

- Fertility problems
- Thyroid problems
- Trouble concentrating
- Persistent fatigue.

Patients should talk with their doctors about

- Any long-term or late effects that may be related to their treatment
- When a child's learning skills should be checked.

Want more information?

You can view, print or order the free LLS fact sheets *Long Term and Late Effects of Treatment for Childhood Leukemia or Lymphoma Facts* and *Long Term and Late of Effects of Treatment in Adults Facts* at www.LLS.org/booklets. Or, contact our Information Specialists for copies.

Follow-Up Care

Medical follow-up is important for every ALL patient. Follow-up care helps the doctor to see if more treatment is needed. Children and adults should see their primary care doctor for follow-up care. They should also see an oncologist (cancer specialist) for follow-up care. Patients should talk to the doctors about how often to have follow-up visits. They can ask what tests they will need. They can also find out how often to have the tests. It is important for patients to get a record of all cancer treatment they received. This is so their doctors can follow up on specific late effects that may be associated with their treatment.

Follow-up care includes physical exams and blood tests. Sometimes bone marrow tests are also needed. As treatment progresses, the doctor may advise longer periods of time between follow-up visits. This will happen if a patient

- Continues to be free of signs of ALL
- Does not need medical care for any long-term or late effects.

To find a survivorship clinic and other resources for child and adult survivors, contact our Information Specialists.

Tracking Your ALL Tests

These tips may help you save time and learn more about your health. Ask your doctor why certain tests are being done and what to expect.

- Discuss test results with your doctor.
- Ask for and keep copies of lab reports in a file folder or binder. Organize the reports in date order.
- Find out if and when follow-up tests are needed.
- Mark upcoming appointments on your calendar.

Take Care of Yourself

- Keep all appointments with the doctor.
- Discuss how you feel with the doctor at each visit. Ask any questions you may have about side effects.
- People with ALL may have more infections than other people. Follow the doctor's advice for preventing infection.
- Eat healthy foods each day. It is okay to eat four or five smaller meals instead of three bigger ones.
- Contact the doctor about tiredness, fever or other symptoms.
- Do not smoke. Patients who smoke should get help to quit.
- Get enough rest and exercise. Talk with your doctor before starting an exercise program.
- Keep a healthcare file with copies of lab reports and treatment records.
- Have regular cancer screening. See your primary care doctor to keep up with other healthcare needs.
- Talk with family and friends about how you feel. When family and friends know about ALL and its treatment, they may worry less.
- Seek medical advice if you feel sad or depressed and your mood does not improve over time. For example, if you feel sad or depressed every day for a two-week period, seek help. Depression is an illness. It can and should be treated even when a person is being treated for ALL. Treatment for depression has benefits for people living with cancer.

Medical Terms

Absolute Neutrophil Count (ANC). The number of neutrophils (a type of white blood cell) that a person's body has to fight infection.

Anemia. A decrease in the level of hemoglobin in the blood.

Antibiotics. Drugs that are used to treat infections caused by bacteria or fungi. Penicillin is one type of antibiotic.

Blast cells. Early bone marrow cells. About 1 to 5 percent of normal marrow cells are blast cells. In ALL, up to 20 percent of marrow cells may be blast cells.

Bone marrow aspiration. A procedure to remove and examine marrow cells to see if they are normal. A liquid sample of cells is taken from the marrow. Then, the cells are looked at under a microscope. **Bone marrow biopsy.** A procedure to remove and examine marrow cells to see if they are normal. A very small amount of bone filled with marrow cells is taken from the body. Then, the cells are looked at under a microscope.

Central line. Special tubing the doctor puts into a large vein in the upper chest. This prepares a patient for chemotherapy treatment. The central line is used to give the patient chemotherapy drugs and blood cells. It also can remove blood samples. Also called **indwelling catheter**.

Central nervous system prophylaxis. Treatment directed to the lining of the spinal cord and brain. ALL cells often collect here.

Chemotherapy or drug therapy. Treatment with chemical agents to treat ALL and other cancers.

Chromosomes. Any of the 23 pairs of certain basic structures in human cells. Chromosomes are made up of genes. Genes give the instructions that tell each cell what to do. The number or shape of chromosomes may not be normal in blood cancer cells.

Clinical trials. Careful studies done by doctors for new drugs or treatments, or studies done for new uses of approved drugs or treatments. The goal of clinical trials for blood cancers is to improve treatment and quality of life and to find cures.

Combination chemotherapy or drug therapy. The use of two or more drugs together to treat ALL and other cancers.

Consolidation therapy. Added treatment given to a cancer patient after the disease is in remission. It usually includes chemotherapy drugs not used during induction therapy. Also called **intensification therapy.**

Cytogenetic analysis. The examination of the chromosomes of ALL cells. This gives doctors information about how to treat patients. Cell samples can be taken from blood or bone marrow.

Diagnose. To detect a disease from a person's signs, symptoms and test results. The doctor diagnoses a patient.

FDA. The short name for the **U.S. Food and Drug Administration**. Part of the FDA's job is to assure the safety and security of drugs, medical devices and the U.S. food supply.

FISH. The short name for **fluorescence in situ hybridization**, a test to measure the presence in cells of a specific chromosome or gene. This test can be used to plan treatment and to measure the results of treatment.

Hematologist. A doctor who treats blood cell diseases.

Hemoglobin. The part of the red cell that carries oxygen.

Immune system. Cells and proteins in the body that defend it against infection.

Immunophenotyping. A lab test that can be used to identify the type of ALL cells.

Induction therapy. The initial treatment with chemotherapy (or radiation therapy). The aim of induction therapy is to kill a maximum number of blood cancer cells so as to induce a remission (absence of signs or effects of the disease).

Intensification therapy. Another name for **consolidation therapy**.

Leukemia. A cancer of the marrow and blood.

Lymph nodes. Small bean-shaped organs around the body that are part of the body's immune system.

Maintenance therapy. Chemotherapy given to ALL patients after induction and consolidation therapy to help destroy the remaining ALL cells. Maintenance therapy is given for about two years.

Marrow. The spongy material in the center of bones where blood cells are made.

Oncologist. A doctor who treats patients with cancer.

Pathologist. A doctor who identifies disease by studying cells and tissues under a microscope.

PCR. The short name for **polymerase chain reaction,** a sensitive lab test that can measure the presence of cancer cell-markers in the blood or marrow. PCR is used to detect remaining cancer cells that cannot be detected by other tests.

Plasma. The liquid part of the blood.

Platelet. A type of blood cell that helps prevent bleeding. Platelets cause plugs to form in the blood vessels at the site of an injury.

Post-remission therapy. The treatment given to ALL patients after induction therapy. Post-remission therapy may have two parts: consolidation (or intensification) and maintenance.

Radiation therapy. Treatment with x-rays or other high-energy rays.

Refractory ALL. ALL that has not responded to initial treatment. Refractory disease may be a disease that is getting worse or staying the same even after treatment (stable disease).

Relapse or recurrence. When disease comes back after it has been successfully treated.

Remission. No sign of the disease and/or a period of time when the disease is not causing any health problems.

Signs and symptoms. Changes in the body that show the presence of disease. A sign is a change that the doctor sees in an exam or a lab test result. A symptom is a change that a patient can see or feel.

Stem cell. A type of cell found in bone marrow that makes red blood cells, white blood cells and platelets.

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Questions to Help You **Choose a Specialist**

Asking questions will help you take an active role in managing your (or your child's) care. If you do not understand any part of the information your healthcare provider gives you, ask him or her to explain it in another way.

Doctor's name _____

Date of appointment or call _____

1. What is your board certification and licensing? Are you a member of any professional societies?

_____

_____

2. How much experience do you have treating patients who have my disease?

_____

_____

3. Is your hospital, university, center or clinic accredited and experienced in treating blood cancers?

_____

_____

4. How long would I usually have to wait for appointments or return of my phone calls?

_____

_____

5. Will there be nurses, social workers and case managers available to help me with support needs and quality-of-life concerns?

6. Do you know of other oncologists who specialize in treating blood cancers? Would you recommend that I speak to any of them?

7. What types of things should I call you about? What types of things should I call my family doctor about?

8. How should I contact you when I have questions?

9. How do I contact you at night? On weekends? On holidays?

To print additional copies of this question guide, or to print copies of question guides on other topics, go to www.LLS.org/whattoask. You may also request that copies be sent to you by contacting our Information Specialists at (800) 955-4572.

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Questions to Ask Your **Healthcare Provider About Treatment**

Asking your healthcare provider questions at any phase of your treatment will help you take an active role in managing your (or your child's) care. If you do not understand any part of the information your healthcare provider gives you, ask him or her to explain it in another way.

Doctor's name _____

Date of appointment or call _____

1. What are my (my child's) treatment options? What is the goal of the treatment?

_____

_____

2. What are the FDA-approved treatments, and are there treatments being studied in clinical trials (study treatments), for my (my child's) diagnosis?*

_____

_____

3. What are the benefits and risks of the treatment(s) available to me (my child)? What are the expected side effects?

_____

_____

4. Is there one treatment option (FDA-approved or study treatment) that you recommend over the others? Please explain.

_____

_____

5. If I (my child) enroll(s) in a clinical trial, who will be in charge of my (my child's) treatment?

6. When do you think I (my child) will need to begin treatment?

7. How long will I (my child) be treated and how many treatments will be needed?

8. Will I (my child) need to be hospitalized for all or part of the treatment?

9. What kind of testing will be done to monitor my (my child's) disease and treatment? How often will testing be needed?

10. If I am treated at an out-patient clinic or at the doctor's office, will I be able to drive/get myself home after treatments or will I need someone to assist me?

11. What are the risks if I don't (my child doesn't) get treatment?

12. How will I know if the treatment is effective? What options are available if the treatment is not effective?

_____

_____

13. How do I find out if my insurance will cover the costs of my (my child's) treatment or the study treatment? Who can help answer any medical questions my insurance company or health plan asks?

_____

_____

14. If I do not have insurance coverage, how can the healthcare team help me (my child) get treatment? Is there someone I need to speak to for assistance?

_____

_____

15. If I'm (my child is) getting a study treatment, will I be responsible for paying any treatment-related costs, such as tests, travel or the clinical trial drug(s)?

_____

_____

16. Will the healthcare team continue to check on me (my child) after the treatment is over? If so, for what period of time?

_____

_____

17. I (My child) would like to continue some type of lifelong follow-up care in order to be monitored for long-term and late effects of treatment. Can I (my child) follow up with you?

_____

_____

*For definitions of an FDA-approved treatment and a clinical trial (study treatment), visit www.LLS.org or contact an Information Specialist.

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REACH OUT TO OUR **INFORMATION SPECIALISTS**

The Leukemia & Lymphoma Society's (LLS) Information Specialists provide patients, families and healthcare professionals with the latest information on leukemia, lymphoma and myeloma.
Our team consists of master's level oncology social workers, nurses and health educators who are available by phone Monday through Friday, 9 am to 9 pm (ET).

Co-Pay Assistance
LLS's Co-Pay Assistance Program helps blood cancer patients cover the costs of private and public health insurance premiums, including Medicare and Medicaid, and co-pay obligations. Support for this program is based on the availability of funds by disease.
For more information, call 877.557.2672 or visit www.LLS.org/copay.

For a complete directory of our patient services programs, contact us at
800.955.4572 or **www.LLS.org**
(Callers may request a language interpreter.)

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For more information, please
contact our Information Specialists
800.955.4572 (Language interpreters
available upon request)
www.LLS.org

or:

National Office
3 International Drive, Suite 200
Rye Brook, NY 10573

Our Mission:

Cure leukemia, lymphoma, Hodgkin's disease and myeloma, and improve the quality of life of patients and their families.

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