Adult Acute Lymphocytic Leukemia (ALL): Types and Treatment

This article is a more specific discussion of ALL. Please be sure to read Leukemia: The Basics first, so you have a basic understanding of leukemia.

Acute lymphoid/lymphoblastic leukemia (ALL) is a blood cancer that affects your white blood cells. If your provider thinks you have ALL, you will have tests done to find out which subtype (classification) of ALL you may have. These tests may include:

- A blood smear and a bone marrow biopsy and aspiration.
- A lumbar puncture (spinal tap) to see if there are any leukemia cells in your spinal fluid.
- Flow cytometry and cytogenetic tests that look for chromosomal changes.
- Imaging tests like CT scans of your chest, abdomen (belly), and pelvis, and an MRI of your head or spinal cord. Younger men may need a testicular ultrasound as ALL can also cause scrotal/testicular masses.

Unlike other cancers, ALL is not staged. It is classified based on chromosomal and genetic abnormalities (changes) in the cancer cells. The classification also helps your provider decide what kind of treatment you need for your ALL. ALL is classified using the World Health Organization (WHO) system. There are 3 categories: precursor B cell ALL, precursor T cell ALL, and mature B cell ALL (also called Burkitt lymphoma).

Your care team may call your leukemia by its chromosomal or genetic abnormalities. The "t" stands for translocation. The p, q and tell where the abnormality is on the chromosome. These are the different classifications of ALL:

- B Lymphoblastic leukemia/lymphoma, not otherwise specified, NOS
- B lymphoblastic leukemia/lymphoma with recurrent genetic abnormalities.
  - B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2), BCR-ABL1 (Philadelphia Chromosome).
  - B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged.
  - B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22) TEL-AML1.
  - B lymphoblastic leukemia/lymphoma with hyperdiploidy >50.
  - B lymphoblastic leukemia/lymphoma with hypodiploidy.
  - B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32) IL3-IGH.
  - B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3) TCF3-PBX1.
- T Cell lymphoblastic leukemia/lymphoma.

How is ALL treated?

Treatment for ALL depends on many things, like your age, overall health, and testing results. Your treatment may include some or all of the following:

- Chemotherapy.
- Targeted therapy.
- Immunotherapy.
- Stem cell transplant.

Chemotherapy, Targeted Therapy, and Immunotherapy

Chemotherapy is the main treatment for ALL. The goal of chemotherapy is remission. Remission means there are less than 5% blast cells found in the bone marrow.
Chemotherapy for ALL can be complex. ALL is often treated for a longer period of time (2-3 years) in three phases. The three phases are:

- **Induction:** The goal of this first phase of treatment is to get you into remission. This means as many cancer cells as possible have been destroyed through chemotherapy. In remission, your team does not see leukemia cells in your bone marrow when they look at it under a microscope. There are also no blast cells in your blood tests and your blood counts have returned to normal.

- **Consolidation:** This is the second phase of treatment. Sometimes it is also called “intensification.” During this phase, you will get higher doses of chemotherapy than you did during induction. These medications are given in combinations and may include some of the same medications you received during induction. The goal is to kill any remaining leukemia cells and to keep you in remission.

- **Maintenance:** This is the third phase of treatment. The goal of maintenance is to keep you in remission. You will receive lower-dose chemotherapy treatment in cycles. Maintenance therapy is not given for Burkitt’s lymphoma leukemia, as this subtype has a high cure rate with induction and consolidation therapy alone.

What chemotherapy medications you receive depends on your age and the classification/sub-type of ALL you have.

The Philadelphia Chromosome is a genetic abnormality that was first identified in chronic myelogenous leukemia (CML) but is also seen in over 20% of adult ALL cases. (Read more about the Philadelphia Chromosome) Tyrosine Kinase Inhibitors (TKIs) are a type of targeted therapy used in the treatment of Ph+ ALL. They include imatinib, ponatinib, nilotinib, bosutinib, and dasatinib. These medications are used alone or in combination with medications including cyclophosphamide, vincristine, daunorubicin, dexamethasone, cytarabine, methotrexate, pegaspargase, etoposide, blinatumomab, and prednisone.

Ph- ALL also uses a combination of several medications for induction. These include nelarabine, pegaspargase, cytarabine, cyclophosphamide, methotrexate, l-asparaginase, and rituximab. In patients with relapsed or refractory Ph-negative ALL, chemotherapy medications used could include those mentioned above as well as blinatumomab, inotuzumab ozogamicin, clofarabine, fludarabine, idarubicin, etoposide and mitoxantrone.

Tisagenlecleucel is a CAR-T immunotherapy that may be used in the treatment of both Ph+ and Ph- refractory ALL. In this treatment, the patient’s own immune cells are used to treat their cancer.

For all subtypes of all, consolidation therapy uses some similar medications, but is more variable in its schedule, depending upon the particular subtype of ALL. Consolidation is given over a period of 4 to 8 months. If you are a candidate for stem cell transplant, then the transplant would likely be done in place of consolidation and maintenance therapy.

Maintenance therapy consists of mercaptopurine (6-MP), methotrexate, vincristine, and prednisone, given over a period of 2 to 3 years. Studies found no benefit to increasing this time but did find outcomes to be worse if the maintenance period was shorter.

**Measurable or Minimal Residual Disease Testing**

Measurable or minimal residual disease (MRD) testing is used to see if the cancer treatment is working and to guide further treatment plans. MRD tests use highly sensitive methods to look for any remaining cancer cells that cannot be seen in routine tests. It is recommended that MRD testing be done in adults with ALL after the completion of induction chemotherapy. Your team may want you to have this test again at other times during your treatment.

**Central Nervous System (CNS) Prophylaxis**

Less than 10% of patients have CNS involvement of leukemia (leukemia found in the spinal fluid) at the time of diagnosis. But, 50-75% of patients will develop this by 1 year if they don’t have preventive therapy. The most effective therapy to prevent CNS disease in adults with ALL is intrathecal chemotherapy. This involves giving chemotherapy directly into the spinal canal. This can be achieved by a lumbar puncture (spinal tap), or through a catheter called an Ommaya reservoir that is surgically implanted in the head. This catheter allows for multiple, repeat intrathecal infusions without needing multiple lumbar punctures. The number of intrathecal infusions given depends on the subtype of ALL and the risk of CNS disease that subtypes have.
Stem Cell Transplant

The use of **stem cell transplant** for ALL is not completely clear. It is most often used early in therapy for patients with high-risk subtypes of ALL in the first remission. Current clinical trials are evaluating the best time for transplant (first or second remission, before maintenance therapy, etc.) and trying to determine which patients will benefit most from transplant.

**Complications & Concerns of Leukemia and Treatment**

Leukemia puts you at a higher risk of infection (because your white blood cell count is low) and bleeding (because your platelet count is low). Treatment of leukemia should help your abnormal blood counts, but your counts may get worse before they get better. You will likely get blood and platelet transfusions, antibiotics, and will need to be extra careful to avoid infection or bleeding.

**Hand washing** is the best way to prevent infection. You and your visitors should wash your hands often. You may also have some restrictions on eating some types of fresh fruit and vegetables or receiving fresh flowers or plants while in the hospital. (See the gift guide for ideas on what to send a patient with these restrictions) This may sound odd, but these items can bring in bacteria and may put you at higher risk of infection. Ask any sick family members to hold off on visiting until they feel better.

In most cases, some type of infection or fever will happen. If it does you will have some tests done to look for where the infection is in your body. These tests can include blood, urine, and stool cultures, and a chest x-ray. Antibiotics may be started or adjusted if they are already being given.

Over the course of their treatment, you may need either blood (for low hemoglobin levels) or platelet (for low platelet counts) transfusions. If you have a low hemoglobin count (also called **anemia**), you may feel fatigued, short of breath, or look pale. A low platelet count (also called **thrombocytopenia**) can lead to bleeding. This can be as small as gums bleeding when brushing the teeth or a nosebleed, or dangerous bleeding, such as a stroke. Be careful and try to avoid bumping into things. Don’t shave with a razor (an electric razor is okay to use with caution) and avoid any activities that increase the risk of bleeding or bruising. Patients should always inform their healthcare team if they have symptoms of anemia or thrombocytopenia.

**Clinical Trials**

You may be offered a clinical trial as part of your treatment plan. To find out more about current clinical trials, visit the [OncoLink Clinical Trials Matching Service](https://www.oncolink.org).

**Making Treatment Decisions**

Your care team will make sure you are included in choosing your treatment plan. This can be overwhelming as you may be given a few options to choose from. Friends and family can help you talk through the options and the pros and cons of each, but they cannot make the decision for you. You need to be comfortable with your decision – this will help you move on to the next steps. If you ever have any questions or concerns, be sure to call your team.

---

OncoLink is designed for educational purposes only and is not engaged in rendering medical advice or professional services. The information provided through OncoLink should not be used for diagnosing or treating a health problem or a disease. It is not a substitute for professional care. If you have or suspect you may have a health problem or have questions or concerns about the medication that you have been prescribed, you should consult your health care provider.