LEUKEMIA & LYMPHOMA SOCIETY OF CANADA®

Acute Promyelocytic Leukemia APL

WHAT YOU NEED TO KNOW

You or your loved one has been diagnosed with acute promyelocytic leukemia (APL). What does it mean and how will it affect you?

This fact sheet will help you:

- Learn about APL and how it is diagnosed
- Get an overview of treatment options
- Understand what happens next

What is leukemia?

Leukemia is a cancer of the blood and bone marrow. Bone marrow is the soft, spongy material inside bones. Blood cells are formed in the bone marrow. When you have leukemia, cancerous blood cells form and choke out healthy blood cells.

There are four main types of leukemia. Each of these main types is further classified into subtypes. APL is a subtype of acute myeloid leukemia (AML).

About APL

- APL is a unique subtype of acute myeloid leukemia (AML).
- Cells in the bone marrow that produce blood cells do not develop or work normally.
- It begins with with a genetic change to the DNA of a single blood-forming cell.
- APL cells have a very specific abnormality that involves chromosomes 15 and 17 (part of your genetic material).
- The bone marrow produces too many immature white blood cells. (called promyelocytes), and they start to build up
- It most often appears in people between the ages of 20–50.
- Affects males and females equally.

Signs and symptoms

Many people with APL feel unwell. This feeling comes from the bone marrow not producing enough normal blood cells and accumulating too many cancer cells. It is also due to the common coagulation problems unique to this type of leukemia. You may experience:

- Fatigue and a pale complexion
  - When your red blood cell count is low (anemia)
- Bleeding, bruising easily, and tiny red spots on your skin (petechiae)
  - When your platelet count or coagulation factors are low
- Weight loss and loss of appetite
  - When you are eating less or using more energy
- Bone or joint discomfort
  - When your white blood cells build up, causing your bone marrow to expand
- Pain or a full feeling below the ribs on the left side
  - When your spleen or liver becomes enlarged, causing your abdomen to swell
- Headache, confusion, and changes in your vision
  - When your APL affects your central nervous system (brain and spinal cord) or if you have a bleed in your brain

APL is considered one of the most curable forms of acute adult leukemia.

With treatment, many people with APL have a good quality of life.

After your diagnosis

With your diagnosis, your doctor can determine the right treatment for you. Your test results help your doctor predict how APL will likely progress and how you may respond to treatment.

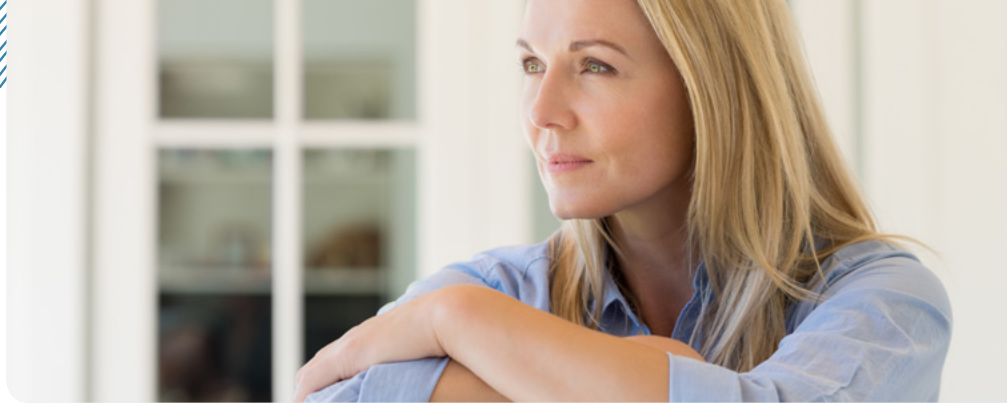
Name of test	Description
Medical history and physical exam	The doctor reviews past illnesses, injuries, and symptoms. They examine your lungs, heart, and other organs.
Bone marrow aspiration and biopsy	These two tests look at bone marrow cells for anything unusual with your chromosomes. The tests are usually done at the same time.
Complete blood count (CBC)	This test measures the number of red blood cells, white blood cells, and platelets in a sample of blood to find out if the counts are high or low.
Karyotyping	This test looks at your chromosomes to spot abnormalities.
Fluorescence *in situ* hybridization (FISH)	This test looks at genes and chromosomes in cells to detect APL cells.
Polymerase chain reaction (PCR)	This test looks at a sample of blood or marrow for specific changes in the structure or function of your genes.
Coagulation status tests	This test checks for blood clots. It is important to treat clots to prevent deep-vein thrombosis, pulmonary embolism, and stroke.
Next generation sequencing	This technology screens for other mutations.

APL treatment

It is important to diagnose APL quickly so you can start treatment. This will help you avoid serious and potentially life-threatening complications. Your treatment is focused on controlling symptoms and decreasing the risk of complications. APL is considered one of the most curable forms of acute adult leukemia.

Treatment is aimed at:

- Targeting the chromosome abnormality
- Bringing blood cell counts to normal or near normal levels
- Decreasing any symptoms



Types of treatment

Drug therapy is the main form of treatment. Many people with APL take two or more drugs.

Drug type	Description
All-trans retinoic acid (ATRA)	targets the chromosomal abnormality. It is an effective treatment: often, a remission follows (meaning no evidence of leukemia in the body).
Arsenic trioxide (ATO)	is a type of chemotherapy drug that uses chemicals to kill cancer cells.
Anthracyclines	are a type of chemotherapy drug that uses chemicals to kill cancer cells.
Antimetabolites	are a type of chemotherapy drug that uses chemicals to prevent cancer cells from growing.

If you are **low risk**, you may have a less intensive treatment plan and often only need ATRA and ATO. If you are at a **higher risk**, the addition of chemotherapy may be needed.

Ask your doctor if you can take part in clinical trials of treatments. Clinical trials test new drugs and treatments before they are approved.

Factors that affect treatment

Discuss your treatment options with your doctor. Make sure you understand the benefits and risks of each approach. Your treatment plan is based on:

- Your age and your overall health
- Whether you are considered low risk or high risk, based on your white blood cell count

Treatment side effects

When you begin your treatment for APL, you may experience mild to severe side effects, depending on your age, your overall health, and your treatment plan. Most side effects disappear once your treatment ends. New drugs and therapies can help control most side effects. Speak to your doctor if you are having side effects.

Common side effects	You may experience side effects such as: • Difficulty breathing, fluid building up around the heart or lungs, and episodes of low blood pressure. This group of symptoms, known as differentiation syndrome, is from your immature cells becoming mature, which is quickly induced by your treatments. • An increase in heart rate that can lead to fainting or seizures, called QT interval prolongation, from ATO treatments. • Bleeding, if abnormalities in your blood coagulation are not properly corrected.

Childhood APL	APL in children and teenagers is similar to APL in adults. • Children are more likely to have higher-risk symptoms, including a high white blood cell count. • Children and teens are treated with similar therapies. • Outcomes are similar to outcomes for adults. Recent studies have shown that very young children are at a higher risk of relapse (cancer returning). • It is important to monitor cardiac (heart) function during treatment.
Long-term or late effects of treatment	Medical follow-up is important after treatment for APL. You may need blood tests, bone marrow tests, or molecular tests to determine if you need further treatment. Your medical team should provide you with a care plan listing the frequency of follow-up visits and the tests you will have at those visits. • **Long-term side effects** are not common but may last for months or years after treatment ends. Examples might be headaches, confusion, and personality changes if a brain bleed took place. • **Late effects** are medical problems that do not show up until years after treatment ends. See your doctor to get follow-up care for possible early detection of heart disease and secondary cancers.

Living with APL can be hard. Seek medical help if you feel "down" or "blue" or don't want to do anything and your mood does not improve over time. These could be signs of depression, an illness that should be treated even when you're undergoing treatment for APL. Treatment for depression has important benefits for people living with cancer. Remember, you are not alone.

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