

media fact sheet

ACUTE PROMYELOCYTIC LEUKEMIA: a subtype of acute myelogenous leukemia

What is leukemia?

Leukemia is a cancer that begins in the soft, inner part of the bones (bone marrow), and in most cases moves into the blood. It can then spread to other parts of the body including the lymph nodes, liver, spleen, central nervous system, and testes.

What is acute myelogenous leukemia (AML)?

Acute myelogenous leukemia (AML) is a cancer that originates in bone marrow cells and is characterized by the uncontrolled growth of myeloid cells, white blood cells other than lymphocytes. In AML, these abnormal cells eventually crowd out the healthy blood cells needed to fight infections. "Acute" means that the leukemia develops quickly, and if not treated, may be fatal in as little as a few months.

12,330 new cases of AML are estimated to occur in the United States in 2010, mostly in older adults. The average age of a person with AML is 67 years.

What is acute promyelocytic leukemia (APL)?

Acute promyelocytic leukemia (APL) is a subtype of acute myelogenous leukemia. It is caused by a chromosome abnormality that rearranges or translocates specific parts of two chromosomes. This translocation prevents myeloid cells from developing into mature white blood cells, leading to an abundance of abnormal, immature cells. The cancerous cells eventually crowd out the healthy blood cells needed for the body to function normally.

Approximately ten percent of adult AML patients will have the APL subtype. The median age of onset of acute promyelocytic leukemia is approximately 40 years of age.

What are the signs and symptoms of APL?

The signs and symptoms of APL are nonspecific and can include fever; easy bruising and prolonged bleeding; nose bleeds and bleeding in the mouth; fatigue; skin rash or lesion; bone pain; shortness of breath; paleness; and in females, abnormal menstrual cycles. The disease is often associated with anemia and low levels of platelets. White blood cell levels

may be low, high or normal.

What causes leukemia?

Most cases of leukemia have no apparent cause. However, exposure to radiation, various chemicals or toxins such as benzene, and some chemotherapy drugs are thought to cause certain kinds of leukemia. Genetic abnormalities may also play a role in the development of leukemia, as well as immunosuppression following organ transplantation and certain blood disorders.

How is APL diagnosed?

A physical examination may show evidence of anemia, paleness and bleeding. Less commonly, an enlarged spleen and liver or enlarged lymph nodes may be found. A comprehensive blood count may show anemia and low platelet count, while the white blood cell count can be high, low or normal. A bone marrow aspiration may show evidence of leukemic cells. Ultimately APL is diagnosed using cytogenetic testing to identify the specific chromosomal translocation that characterizes the disease.

How is APL treated and what is the prognosis?

The treatment of APL differs from that for all other forms of AML. Most APL patients are treated with chemotherapy together with retinoid-based therapy, which causes the leukemic cells to mature into normal white blood cells.

Current first-line treatments produce long-term remission in 70 to 80 percent of patients. However, research indicates that approximately 20 to 30 percent of patients with APL (approximately 400 patients per year) will relapse from first-line therapy. In patients who relapse after remission, treatment may include the use of arsenic therapy.

Inducing molecular remission is a major treatment goal for APL, thought to be a target for achieving long-term survival.