What is cancer of the blood?

Blood contains various different types of cell, each of which has its own role to play. Red blood cells (erythrocytes) transport oxygen to the different parts of the body and organs; platelets (thrombocytes) are important for blood clotting. The white blood cells (granulocytes and lymphocytes) are responsible for the body's own defence system. All blood cells – white and red blood cells and platelets – originate from a common source in the bone marrow, the haematopoietic (blood-forming) stem cell. The process of blood formation is known as haematopoiesis (see diagram). The first cells to develop from stem cells are called myeloid and lymphatic stem cells, which then develop into the various blood cells and are subsequently released into the bloodstream.



Blood cells, just like other cells in the body, can mutate at any stage of their development, leading to cancer. Rudolf Virchow coined the term "leukaemia" for cancer of the blood. The word comes from the Greek and means "white blood", since a significantly increased number of white blood cells is often found in the blood of patients with a specific type of blood cancer.

Year	Estimated number of new cancers (all ages)	Male	Female	Both sexes
2012		2426	2119	4545
	ages < 65	2038	1687	3725
	ages >= 65	388	432	820

2020		2711	2413	5124
	ages < 65	2227	1845	4072
	ages >= 65	484	568	1052
Demographic change		285	294	579
	ages < 65	189	158	347
	ages >= 65	96	136	232

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Population forecasts were extracted from the *United Nations, World Population prospects, the 2012 revision.* Numbers are computed using age-specific rates and corresponding populations for 10 age-groups.

nternational Agency for Research on Cancer

Leukaemia





International Agency for Research on Cancer





Leukaemia is classified as either myeloid or lymphatic leukaemia depending on which cells in the blood have mutated. It is subdivided further into "acute" or "chronic" leukaemia, depending on the stage of development of the malignant cells.

Acute leukaemia usually starts suddenly and can occur at any age. Immature, malignant cells accumulate in the bone marrow and cause a decrease in normal blood formation.

The course of chronic leukaemia is slower. This form is seen most often in older people. Chronic leukaemia is frequently discovered by accident, during a routine blood test, for example.

Leukaemia is classified accordingly into four different types:

- acute lymphoblastic leukaemia (ALL)
- acute myeloid leukaemia (AML)
- chronic lymphocytic leukaemia (CLL)
- chronic myeloid leukaemia (CML)

What are the causes of and risk factors for developing leukaemia?

Cancer of the blood may develop if the normal maturation process of the white blood cells (leukocytes) in the bone marrow or lymphatic system is interrupted as a result of faulty switching of certain genes.

Instead of mature white blood cells, which are fully developed and hence functional, the white blood cells formed are immature to a greater or lesser extent.

These cells are not usually functional, and typically have the ability to multiply rapidly and in an uncontrolled manner.

This means that they increasingly suppress normal haematopoiesis in the bone marrow, with the result that healthy white blood cells, red blood cells and platelets are no longer formed in the numbers required.

This may manifest itself as follows:

- Anaemia due to a lack of functional red blood cells
- Increased susceptibility to infection due to a lack of functional white blood cells
- Increased susceptibility to bleeding due to a reduced number of platelets

These symptoms can also occur in other diseases and therefore the doctor can only make a diagnosis with the aid of more detailed investigations.

What role does lifestyle play in leukaemia?

The causes of leukaemia are not yet fully understood. Suspected triggers are radiation, chemotherapeutic substances (e.g. used to treat another cancer previously), chemicals or smoking. Experts estimate that some 10 per cent of leukaemia cases are caused by cigarette smoking.

It has been shown that some people are at greater risk of developing acute leukaemia, such as children with certain hereditary diseases, in particular trisomy 21 (Down's syndrome).

How is leukaemia diagnosed?

A number of investigations are undertaken in order to make an accurate diagnosis:

- Physical examination including liver and spleen (both of these may be enlarged in leukaemia) and lymph nodes
- Blood test
- Bone marrow aspiration a sample of bone marrow is taken under local anaesthetic. Because the blood is formed in the bone marrow, changes can be detected here. Investigations of the cell chromosomes (cytogenetic testing) are also carried out using the bone marrow cells. The result of these tests may be crucial for diagnosis and treatment, and also for the prognosis.
- Cerebrospinal fluid aspiration (lumbar puncture) Some forms of leukaemia can affect the nervous system, which is why this investigation is sometimes carried out. It involves taking a sample of cerebrospinal fluid from the lumbar spine with a syringe and examining it.

How is leukaemia treated?

The aim of all leukaemia therapies is to destroy the leukaemia cells and restore normal blood formation. Treatment for leukaemia differs depending on the form of the disease and is carried out by specialist doctors.

For patients with acute leukaemia, it is important that treatment is immediate and very intensive. The treatment is designed to be more or less intensive depending on the form of the leukaemia and the condition of the individual patient, so as to maximise its effectiveness while minimising side effects.

In contrast to chronic leukaemias, some patients with acute leukaemias can be permantely cured by chemotherapy alone. For many patients, a stem cell transplant, from the bone marrow for example, therefore represents the only chance of a cure. In what is known as allogeneic bone marrow transplant, the diseased bone marrow of the patient is replaced by healthy bone marrow from a donor. Alternative options for obtaining stem cells are to take them from the donor's bloodstream or from umbilical cord blood.

Chronic leukaemia generally progresses more slowly and is often discovered during routine examinations. As with acute leukaemias, chronic forms are treated with drugs that inhibit the growth of the malignant cells.

The aim of chemotherapy is to kill cancer cells throughout the body by means of medicines that inhibit cell growth (cytostatics). Cytostatic drugs are effective against rapidly-growing cells. Cancer cells typically grow very rapidly. The type and scope of chemotherapy depend in each individual case on the type of leukaemia and the extent to which the disease has advanced.

Further information - diagnosis and treatment

The classification of leukaemias is based on the morphological and immunological characteristics of the leukaemic cells.

Cytogenetic and molecular biology characteristics have also become increasingly important in recent years.

A tentative diagnosis can often be made just from the blood count and differential blood count, but precise classification normally requires bone marrow aspiration.

Treatment of acute myeloid leukaemia (AML) is undertaken in several stages:

- 1. Induction therapy: Two to three different drugs are administered in blocks over several days. These blocks are repeated depending on the treatment regimen being used. Induction therapy is designed to kill the leukaemic cells and restore normal blood formation. Once this has been accomplished the patient is said to be in remission.
- 2. Consolidation therapy: To prevent the leukaemia recurring (relapse). The number of leukaemic cells is reduced still further.
- 3. In some cases, a course of maintenance therapy follows, which is also designed to minimise the risk of relapse.

The treatment of acute lymphoblastic leukaemia (ALL) also consists of several stages which last for a total of about two to three years:

- 1. Induction therapy with several different drugs to achieve remission (see AML)
- 2. Consolidation therapy to stabilise the remission
- 3. Re-induction therapy with similar drugs to those used in the induction; this block is necessary to reduce the risk of relapse
- 4. Maintenance therapy to further reduce the risk of relapse

Chronic myeloid leukaemia (CML) usually progresses in three phases:

- 1. The first phase is the chronic phase, which is characterised by slow cell division. Most cases are discovered during this phase, when the disease can easily be treated.
- 2. The second or accelerated phase involves accelerated cell division and deterioration of the blood count.
- 3. The third phase is the "blast crisis", which resembles acute leukaemia.

Typical of CML (in all phases of the disease) is the presence of a specific gene change, known as the Philadelphia chromosome, in the leukaemic cells. This results in formation of a new enzyme (called a "tyrosine kinase") which is constantly active and results in uncontrolled proliferation of the leukaemic cells. It is precisely this enzyme that is the starting point for treatment of CML, with new drugs that inhibit the tyrosine kinase and hence the proliferation of leukaemic cells. The drugs must be taken daily and on a permanent basis, since this is the only way to control the disease.

Treatment of chronic lymphocytic leukaemia (CLL) depends on the clinical picture. In a few cases, no chemotherapy is required and progress is simply monitored closely ("watch and wait"). In more aggressive cases, drug treatment is necessary.