

Nursing care of a child with acute lymphoblastic leukemia

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ABSTRACT

Every year a very large number of children in the world suffer from acute lymphoblastic leukemia, and for years there has been a steady increase in the number of new cases. Acute lymphoblastic leukemia accounts for 75% of leukemia cases in the world. Lymphoblastic leukemia is a cancer disease that originates in B or

T cell lymphocytes, which expansion takes place in blood and in the bone marrow.

The etiology of the disease is not fully understood because it consists of several factors

conditioning its formation. The most important element is the early detection and taking actions resulting in effective disease control through treatment and care of the patient.

The nursing process should allow the patient to be involved in and accept the ongoing cancer process, and medical personnel, family and specialists in such fields as psychology and psychiatry should participate.

Keywords: Acute lymphoblastic leukemia, cancer, care.

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ACUTE LYMPHOBLASTIC LEUKEMIA

The incidence of cancer in children up to the age of 15 varies between 110-150 cases/1 million children. This means that from 1/600 to 1/450 children will face this disease. Cancer is one of the main causes of deaths of children right after accidents, injuries, and poisonings [1-3]. It is estimated that leukemia accounts for about 26% of cancers in children in Poland. In most cases, it is acute lymphoblastic leukemia, which accounts for 75% of leukemias. The peak incidence is in the period between the age of 2 and 5, and the rate of ALL (acute lymphoblastic leukemia) in boys is

30% greater than in girls. Over the last 20 years, the incidence of acute lymphoblastic leukemia has increased by 0.9% per year [3-7]. The morbidity rate of ALL in different regions of the world is varied. There are geographical differences in the incidence of this cancer. The highest incidence rate is observed in the USA, Germany, Australia and Costa Rica. In the majority of European countries, the incidence remains at an average level. The lowest one occurs in China, India, Japan, New Zealand and Brazil. The highest morbidity rate in children in the USA is observed in Caucasian, while much lower in children of African Americans [3,6,7,8].

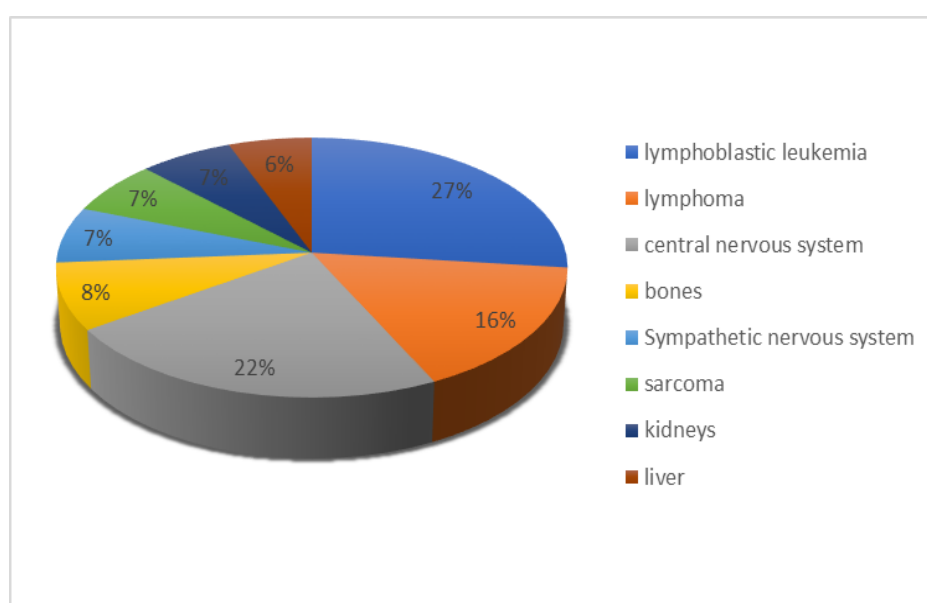


Figure 1. The incidence of specific cancers in children [7]

The development of ALL occurs as a result of leukemia transformation in the B or T lymphocyte system cells. The causes of mutations are not fully explained; however, it is suspected that genetic, viral, environmental and immunological abnormalities have a significant impact on the disease. Leukemia is often accompanied by congenital chromosomal disorders, including Down syndrome, Bloom syndrome, neurofibromatosis, Klinefelter syndrome, Swachman syndrome, ataxia, Langerhans cell histiocytosis, albinism, neurocutaneous syndromes, immune dysfunctions, gastrointestinal syndrome, chronic inflammatory bowel disease and other syndromes, severe combined immunodeficiency. The main risk factors include immune system dysfunctions. Moreover, it is believed that males, age from 2 to 5 and a higher socio-economic status may predispose to the

occurrence of ALL [6,8]. Environmental factors also play an important role in the initiation of cancer. Caucasian children and children living in highly developed countries are more exposed to the risk of leukemia [7].

The treatment of acute lymphoblastic leukemia is based mainly on the standard of treatment of leukemia, which is chemotherapy. A cancer diagnosis causes anxiety and shock both in the patients and the family. Therefore, it is medical staff who should be responsible for providing patients with comprehensive care and psychological support for them and their families. Such conditions can be provided by creating and professional functioning of an interdisciplinary therapeutic team. Proper care allows to improve the quality of life of a cancer patient [1,6,8].

CARCINOGENESIS OF ACUTE LYMPHOBLASTIC LEUKEMIA

In 2008, the World Health Organization introduced the ALL classification based on cell biology and human development [9]. This classification was introduced in order to implement the best treatment and determine the complications. Acute lymphoblastic leukemia was included in the group of "tumors of lymphocyte progenitors" where B-lineage and T-lineage cells were isolated. The process of tumor formation, or carcinogenesis, takes a long time and is a multi-stage process [8].

The first phase of cancer development is called initiation and it is the emergence of a single mutation under the influence of a carcinogen. In the case of genetically determined ability of the body to eliminate carcinogens and DNA repair, there is no damage to the cells. In the opposite situation, when the body has no ability to fight such factors, a mutation occurs in the DNA. Already at this stage there may be adverse changes in the tissues, despite the fact that the affected cell may look normal. Typically, tissue hypertrophy also occurs at this time [10-12].

The next phase is promotion, or multi-stage clonal development. During this time, mutated cells multiply preventing destruction. Cells lose their functional capability, which results in an increased activity of enzymes and growth factors as well as suppression of the production of proper enzymatic proteins. In this phase, proliferation also occurs, i.e. the rapid growth and spread of cancer cells. Mutated cells begin to show invasiveness and lose connection with healthy cells. During this phase, there are also disturbances in the number and structure of chromosomes, and cells acquire pre-cancerous features [10-12].

Conversion is the third stage of cancer development, also known as the asymptomatic phase, in which mutation and multiplication of affected cells occur. At this stage, irreversible changes and formation of cancer cells occur [10,11,12].

The last phase, known as progression, is characterized by local invasion and the formation of so-called metastasis. Cancer cells get into blood and lymph vessels and spread to other organs. During the progression, the full symptoms of cancer are observed [11,12].

ALL symptoms are due to proliferation of the leukemic clone in the marrow and displacement of healthy cells, as well as the emission of lymphoblasts into blood. Early signs of cancer in children are non-specific. Initial symptoms result from dysfunctions of the bone marrow in which erythrocytes, thrombocytes and leukocytes are produced. There are also symptoms that arise as the result of acute extramedullary leukemic infiltration

[13]. General symptoms include: fever, apathy, fatigue, paleness, lack of appetite, pain in bones and joints [1].

Symptoms resulting from affected bone marrow and the inhibition of hematopoiesis are mainly anaemia, neutropenia and thrombocytopenia. Paleness, fatigue, tachycardia and dyspnoea are observed in anemia. Neutropenia is characterized by fever, mouth ulcers and infections. Symptoms associated with thrombocytopenia include: ecchymosis, bleeding from mucous membranes, sallowing, bruising. A large group of symptoms resulting from immunodeficiency disorders are: painful mouth sores, mouth ulcers, herpes, angina, and periodontal changes. The susceptibility to bacterial and fungal infections also increases. Symptoms indicating that the lymphatic tissue is affected include enlargement of lymph nodes, splenomegaly and hepatomegaly [1]. A significant group are the symptoms resulting from the infiltration of organs by leukemic cells, and for example when the central nervous system is infected, there is an increase in intracranial pressure which results in morning nausea and vomiting, headaches, bradycardia, papilloedema, and the sixth nerve palsy. Among the symptoms of brain infiltration focal symptoms such as hemiplegia, cranial nerve palsies, convulsions, ataxia, dysmetria and hypotonia are distinguished [1]. Moreover, the symptoms of the affected central nervous system may be hypothalamic syndrome, which manifests as a polyphagia with continuous weight gain, hirsutism and behavioral disorders. The affected urogenital system is most often manifested by painless enlargement of testicles in men and ovaries in women. During ultrasound examination infiltration of kidneys may be diagnosed. It may be manifested by hematuria, hypertension and renal insufficiency. In case of affected gastrointestinal tract, bleeding may occur, however, gastrointestinal leukemia infiltrations are clinically invisible until the terminal stage. Dysfunction of the musculoskeletal system is manifested in the form of bone pain and spine lesions that can cause vertebral collapse with spinal cord compression and paresis of the limbs. Skin symptoms most often occur in newborns, where infiltrations within salivary glands can occur. In case of affected heart, changes include infiltrations and intramyocardial or pericardial haemorrhages. When the lungs are affected, leukemia cell proliferation and haemorrhages also occur [1,9,13].

Diagnosis of acute lymphoblastic leukemia should be performed in specialized haematological units. Diagnosis is based on the results of morphological examinations of blood and bone marrow. According to the World Health Organization, ALL is diagnosed when the percentage of leukemia cells in the bone marrow is

≥20% [9]. Laboratory tests helpful in diagnosis of ALL include blood cell morphology, bone marrow aspiration, molecular and immunophenotype tests, cytogenetic, trepanobiopsy, haemostasis test, ECG, cardiac assessment, cerebrospinal fluid test, and other laboratory test as well as and imaging (abdominal ultrasound, computed tomography, magnetic resonance imaging of the central nervous system and chest x-ray).

TREATMENT OF ACUTE LYMPHO-BLASTIC LEUKEMIA

ALL treatment should be conducted by specialized and interdisciplinary medical staff in oncohaematological hospitals. Treatment planning depends on prognostic factors that are associated with the risk of recurrence during or after treatment. Determination of risk factors allows to select a treatment method that will enable to achieve optimal results [14]. Baseline prognosis is based on cytogenetic characteristics, clinical and laboratory characteristics, and changes in gene expression [9].

After recognizing and determining the risk factors and general condition of the patient, he/she must immediately be placed in a hospital private room to protect against infection. It is necessary to treat possible complications and improve the overall condition of the patient. Time, from the diagnosis to treatment implementation, adversely affects the final results of treatment [11].

The treatment of ALL is based on the standard of chemotherapy. The choice of protocol according to which chemotherapy will be carried out depends on the immunophenotype and the risk group. When administering cytostatic medications, supportive therapy is also very important. It should include the prevention of infections, their elimination, treatment of bleeding disorders and anaemia. In addition, it is recommended to administer recombinant hematopoietic stimulants. Prevention of the central nervous system is also very important [8,9].

The first stage of chemotherapy is steroidal prophylaxis and it covers 7 days during which the patient is given glucocorticosteroids. The goal of this phase is to obtain not too rapid breakdown of the tumor cells so that the tumor lysis syndrome does not occur. At this stage, it is also possible to determine drug resistance of leukemic cells. If there is a bad response to the applied measures, we are dealing with an active multi-drug resistance mechanism, which is synonymous with the occurrence of unfavorable prognostic factors [1,9]. The next step is induction, which refers to reducing cancer cells to the amount impossible to detect by typical hematological methods. The number of tumor cells is usually reduced from 1012 to <109. The goal is to achieve complete remission. Over the

next 3-6 weeks after the treatment induction, bone marrow regeneration and expected remission occurs [1,9].

The third stage is consolidation, which takes place after achieving complete remission, despite the fact that the number of leukemic cells can reach 109. Such a high number of cells may be the cause of relapse; therefore, consolidation treatment is used to exclude the so-called minimal amount of cytostatic medications [1,9].

The final stage is a post-consolidation treatment aimed at maintaining remission and preventing recurrence. In patients in good general condition belonging to a high risk group and having a donor, allogeneic stem cell transplantation is recommended [1,9].

During the illness, the child's psyche is a very important aspect. Psychoemotional factors have a large impact on pain and adaptation of the patient to the new situation which is the disease [15]. In case of children, they handle treatment and aggressive therapy better, despite the more rapid course of the disease, however, the responses to treatment and hospitalization are more divergent. Older children close up and show aggression. They have the need to move and contact with their peers, while the disease process prevents them from doing so. Small children react with fear of the unknown and pain. Then, the presence of parents plays a very important role in order to build a sense of security and help find themselves in a new situation. Teenagers show a worse response to treatment and the disease. Emotional swings associated with adolescence are natural for this stage of life. The necessity to feel acceptance among peers and the physical appearance plays a big role. Teenagers want to make choices themselves and have influence on the treatment process, when there is often a situation in which this autonomy is undermined, for example by medical personnel. Young people want to emphasize their independence, and when they fail, they rebel and close up [16].

An aspect worth paying attention to is the negative impact of cancer on the development, personality and education of the child's sphere of life. The disease exposes the child to pain in the physical, mental and social spheres. A sudden diagnosis disturbs the current life of both the child and the family, causes fear and stress. A child torn out of his/her environment must adapt to life according to new rules. Patients are exposed to negative feelings and emotions from the beginning of treatment, they must undergo processes that are very painful. The disease and its therapy also expose the child to changes in the mental sphere. At this stage, psychological help may be necessary because pathological reactions may occur, which is why it is very important to look after the child by

an interdisciplinary team, or a group of people acting together, for whom the immediate goal is the fastest recovery of the child. The team includes: pediatric oncologists, pediatric surgeons, nurses, clinical psychologists, therapists, priests, teachers, volunteers and parents. Only the whole team can provide proper medical care and support to the child and parents [17].

A child suffering from ALL requires intensive care of medical personnel. They play a significant role in the care, therapy and education of the patient and the family. Their task is also a careful observation and establishing a friendly contact with the child, which facilitates future cooperation. People, who from the beginning of the hospitalization take an active part in preparing the child for diagnostic tests or further treatment, are nurses and doctors. Their duty is to follow the principles of asepsis and antisepsis, to inform the child and parents about planned procedures, preparation and care during and after the tests. Such support allows to minimize anxiety during subsequent stages of the therapy [11].

The staff also plays an important role during treatment. Children need close observation in order to avoid complications such as those after administering cytostatics. In addition, the staff supervise if the administration of medicines is safe and correct. It is extremely important to correctly insert the intravenous cannula and to care of the injection site in order to avoid infection. The cooperation of medical personnel with ill children and their families allows the identification of needs and their appropriate satisfaction [11,17].

Nurses and doctors also play the role of educator of a child and the parents. These tasks include compliance with the sanitary regime, care for hygiene and cleanliness of the environment. It is very important to inform about the food requirements so that it is fresh and unprocessed. Another issue is the adaptation of the home for the child during breaks in chemotherapy. The home should provide hygienic conditions similar to those prevailing in the clinic. The child should not be visited by ill people. The medical staff should also inform parents about the possibility of using family and care-related benefits [17].

SUMMARY

In Poland, every year about one in 15 to 25 thousand children, between 2 and 5 years of age became affected by lymphoblastic leukemia. Fortunately, more than 2/3 of children undergoing treatment can be completely cured. Knowledge of the problems of young patients and their families by medical personnel allows cooperation to minimize the consequences of treatment and hospitalization. Empathy allows proper implementation of the

treatment process and provides professional and comprehensive care. Thanks to the tips, knowledge and help, it is easier for patients to go through the painful process of therapy. Knowledge, experience and skills affect the safety during the therapeutic process and the final result of treatment. The nurse is one of the persons in the ward with whom the patients and their relatives, who take care of them during hospitalization, have continuous and close contact.

„As far as nursing care is concerned, from Cepuch et al. [18], outcomes of the therapy and quality of care result not only from nurses' knowledge about the disease itself and its influence on patient's functioning but also from carefully developed plan of actions directed toward the patient and considering patient's age, mental status, and the method of applied treatment”.

CONCLUSIONS

The following conclusions based on medical publications have been drawn:

- The most common causes of acute lymphoblastic leukemia are genetic, environmental and viral infections and also their consequences.
- Properly implemented treatment process and cooperation of the patients and their families, together with an interdisciplinary therapeutic team positively affect the final result of the treatment.
- Children need more support and attention from the parents and medical staff in the course of cancer to ensure a sense of security and to help find themselves in a new situation.
- Education in the field of a lifestyle of patients with cancer is extremely important.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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