Acute Lymphoblastic Leukemia in Children

Deborah Cagen, ARNP, MSN, CPON®, and Michelle Franco, ARNP, MSN, CPN

1. Acute lymphoblastic leukemia (ALL), the most common malignancy in children, accounts for what percentage of all cancer diagnosed in children who are younger than age 15?
   a. 10%
   b. 15%
   c. 25%
   d. 40%

2. Bobby Sullivan recently was diagnosed with ALL. After a family conference with the physician, Bobby’s parents approach the nurse and ask for clarification on Bobby’s long-term survival. Which of the following age groups has the most favorable prognosis?
   a. 1–4 years
   b. 5–10 years
   c. 11–14 years
   d. 15–19 years

3. Later, Bobby’s parents approach the nurse and ask how their seven-year-old child acquired leukemia. Which of the following causes would be the most appropriate response?
   a. Exposure to a rare virus
   b. Environmental influences
   c. Mutations in their child’s cells
   d. Radiation exposure

4. A four-year-old boy is admitted to the pediatric oncology unit for evaluation of a possible diagnosis of ALL. In addition to a bone marrow aspiration (BMA) and comprehensive blood work, which additional diagnostic test should be used to confirm the diagnosis of ALL?
   a. Lumbar puncture (LP)
   b. Bone scan
   c. Computed tomography (CT) of the head
   d. Testicular biopsy

5. The definitive diagnosis of ALL is confirmed when the bone marrow reveals at least what percentage of lymphoblasts?
   a. 10%
   b. 20%
   c. 25%
   d. 35%

6. The most appropriate laboratory diagnostic study to establish subtypes (lineages) of ALL is
   a. Cytochemistry stains.
   b. Cerebrospinal fluid (CSF) cytology.
   c. Immunophenotyping.
   d. Cytogenetic analysis.

7. The rationale for treating the central nervous system (CNS) in patients with ALL is based on which premise?
   a. The CNS is considered a pharmacologic sanctuary site.
   b. More than 50% of children have detectable CNS disease at diagnosis.
   c. Intrathecal chemotherapy is more efficacious than radiation therapy, with fewer side effects.
   d. Intrathecal chemotherapy should be given only as salvage therapy.

8. Which cytogenetic abnormality is a poor prognostic indicator in children with ALL?
   a. Trisomies 4 and 10
   b. Translocation (4; 11)
   c. Hyperdiploidy
   d. TEL AML-1 gene

9. Mrs. Smith approaches the nurse and begins crying, stating, “I caused my child’s leukemia because I gave her bad genes.” Which response would be most appropriate?
   a. Gently reassure the mother that ALL is not considered an inherited disorder.
   b. Comfort the mother and encourage her to obtain genetic testing if she decides to have another child.
   c. Refer Mrs. Smith for individual counseling.
   d. Encourage Mrs. Smith to attend the unit support group.

10. In addition to fatigue, bone pain, and bleeding, which symptom is seen commonly in childhood ALL?
    a. Fever
    b. Blurred vision
    c. Behavioral changes
    d. Enlarged, very painful lymph nodes

**Answers**

**Question 1:** The correct answer is choice c, 25%. ALL is the most common malignancy in children, accounting for nearly 25% of all cancers diagnosed in children younger than 15. About 20% of all cases of ALL occur in adults. In 2003, approximately 3,600 adults and children will be diagnosed with ALL and 1,400 people with ALL will die (American Cancer Society, 2003; National Childhood Cancer Organization, 2003; Westlake & Bertolone, 2002). Therefore, choices a, b, and d are incorrect.

**Question 2:** The correct answer is choice a, 1–4 years. Five-year survival rates have been shown to be the highest, 85%, for the 1–4 age group. Survival in children with ALL is related strongly to age at diagnosis (Westlake & Bertolone, 2002).

Deborah Cagen, ARNP, MSN, CPON®, is a clinical nurse specialist in the Pediatric Intensive Care Unit and Michelle Franco, ARNP, MSN, CPN, is a clinical nurse specialist in the Hematology/Oncology Unit, both at Miami Children’s Hospital in Florida.

**Key Words:** acute lymphoblastic leukemia; children; abnormalities, cytogenetic

Digital Object Identifier: 10.1188/03.CJON.604-606
Choices b, 5–10 years, and c, 11–14 years, are incorrect because a relationship exists between age at diagnosis and outcome. Children older than 10 have a relatively poor prognosis compared to children in the younger age group of 1–4 years.

Choice d, 15–19 years, also is incorrect. Adolescents with ALL fare poorly because they have a lower remission induction rate than younger children. Adolescents tend to have other poor risk factors, including T cell immunophenotyping (Margolin, Steuber, & Poplack, 2002).

Question 3: The correct answer is choice c, mutations in their child’s cells. ALL is believed to originate from a mutation in a single hematopoietic lymphoid progenitor cell, capable of indefinite self-renewal, and passed on to all of the person’s descendants (Westlake & Bertolone, 2002).

Choice a, exposure to a rare virus, is incorrect, although some researchers have had an intense interest in correlating childhood leukemia and viruses because of the fact that a child’s immune system is developing and, perhaps, may be more vulnerable to the oncogenic effects of particular viruses. No direct association between childhood or maternal viral infections and ALL has been documented (Margolin et al., 2002).

Choice b, environmental influences, is incorrect because little is known about the cause of cancer in children and adolescents. Given the fact that the duration of exposure to potential environmental carcinogens is directly proportional to age, very little evidence suggests that cancers in young people are related to known environmental carcinogens. Environmentally related cancers appear to take considerably longer than one to two decades to manifest in most people (Reaman & BLEVER, 2002).

Choice d, radiation exposure, is also incorrect, although controversy surrounds this issue. Ionizing radiation has the potential to cause leukemia; however, the actual percentage of leukemia cases directly attributed to radiation is small (Margolin et al., 2002).

Question 4: The correct answer is choice a, LP. The workup for childhood ALL includes a comprehensive physical examination, complete blood count with differential, BMA, chemistry panel, uric acid, lactate dehydrogenase, chest x-ray, and LP. Although only 3% of children have detectable CNS involvement at diagnosis, obtaining a baseline understanding of CNS involvement is considered standard practice (Westlake & Bertolone, 2002).

Choice b, bone scan, is incorrect. The most definitive test to make the diagnosis of leukemia is BMA. In most circumstances, BMA provides sufficient material to establish the diagnosis. Bone scans, serum liver panels, and uric acid levels are not traditionally a part of the ALL workup (Margolin et al., 2002).

Choice c, CT of the head, is incorrect. This study usually is part of the workup for acute myeloid leukemia to rule out a chroma (a type of tumor associated with myeloid leukemia formed by the accumulation of white blood cells).

Choice d, testicular biopsy, is incorrect. Routine testicular biopsy was performed in the past because the testes were considered a sanctuary disease site, but the procedure is no longer recommended routinely. The test is reserved to document relapse or residual disease for patients with persistently enlarged testes (Westlake & Bertolone, 2002).

Question 5: The correct answer is choice c, 25%. Although more than 5% lymphoblasts in the bone marrow usually is suggestive of leukemia, a minimum of 25% blast cells is required before the diagnosis in confirmed (Margolin et al., 2002; Westlake & Bertolone, 2002). Therefore, choices a (10%), b (20%), and d (35%) are incorrect.

Question 6: The correct answer is choice c, immunophenotyping. To establish the correct diagnostic subtype of ALL, immunophenotyping of lymphoblasts is necessary. Monoclonal antibodies are used to identify cell surface antigens of hematopoietic cells associated with B cell, T cell, and myeloid lineages of malignant cells. The two lineages identified in ALL are B cell and T cell (Westlake & Bertolone, 2002).

Choice a, cytochemistry stains, is incorrect. These stains subdivide the cells according to the French-American-British system. ALL is divided into three subtypes: L1, L2, and L3. The L1 morphology subtype is the most common, accounting for 84% of children with ALL, and has the best prognosis.

Choice b, CSF cytology, is incorrect. The purpose of a lumbar puncture is to determine whether lymphoblasts are present in the CNS and does not establish ALL subtypes. Only 3% of patients have detectable CNS involvement at diagnosis (Westlake & Bertolone, 2002).

Choice d, cytogenetic analysis, is incorrect. Although intense focus has been placed on cytogenetic analysis, cytogenetics assist in subtyping leukemia but do not define lineage. Aberrations in chromosomal number (ploidy) and/or structure (translocations, deletions, rearrangements) occur in more than 90% of childhood ALL cases, and many are of prognostic significance (Hockenberry, WILSON, WINKELSTEIN, & KLINE, 2003).

Question 7: The correct answer is choice a, the CNS is considered a pharmacologic sanctuary site. Preventive CNS therapy is based on the premise that the CNS provides a sanctuary site where leukemic cells, undetected at diagnosis, reside protected from the action of systemic chemotherapy by the blood-brain barrier (Westlake & Bertolone, 2002).

Choice b, more than 50% of children have detectable CNS disease at diagnosis, is incorrect. Only 3% of patients have detectable CNS involvement at diagnosis (Westlake & Bertolone, 2002).

Choices c, intrathecal chemotherapy is more efficacious than radiation, and d, intrathecal chemotherapy should be given only as salvage therapy, are incorrect. Fifty percent or more of children eventually will develop overt CNS leukemia unless specific treatment is provided such as intrathecal chemotherapy or cranial irradiation. For this reason, all children receive CNS prophylactic therapy. Before the 1980s, children with ALL received cranial or spinal irradiation. Because of a concern regarding the late effects of radiation therapy, this treatment modality is reserved for high-risk patients or those with confirmed CNS disease (Hockenberry et al., 2003).

Question 8: The correct answer is choice b, translocation (4; 11), which is the mixed lineage leukemia gene rearrangement. This is associated with high-risk and unfavorable outcomes in infants and children with ALL. Choices a, c, and d are incorrect. Trisomies 4 and 10, hyperploidy, and TEL-AML1 gene are all favorable prognostic factors (Westlake & Bertolone, 2002).

Question 9: The correct answer is choice a, gently reassure the mother that ALL is not considered an inherited disorder. One of the key risk factors for childhood ALL is chromosomal aberrations. Estimates suggest that 60%–70% of children with ALL have cytogenetic abnormalities that will direct their treatment plan. Parents need to be reminded that abnormal chromosomal results refer to aberrations in the cancer cell and are not an inherited disorder (Robinson, 2001).

Choice b, comfort the mother and encourage her to obtain genetic counseling, is incorrect. Established guidelines indicate when genetic testing should be performed. Direct testing and linkage analysis are the two approaches in identifying a genetic risk of a predisposing cancer mutation. Direct testing in pediatrics is recommended for those with a familial history of retinoblastoma, Wilms tumor, and Li-Fraumeni syndrome (a cancer predisposition syndrome associated with leukemia, melanoma, soft tissue sarcoma, and cancer of the breast, colon, pancreas, or...
brain). Predictive tests for other pediatric cancers are in development (Ruccione, 2002).

Choices c, refer Mrs. Smith for individual counseling, and d, encourage Mrs. Smith to attend the unit support group, are incorrect. These interventions do not completely address Mrs. Smith’s need for education. In this particular situation, she is confirming that she has a knowledge deficit as to the etiology of her child’s leukemia.

**Question 10:** The correct answer is choice a, fever. Symptoms such as fatigue, bone pain, bleeding, and fever are the most common symptoms and clinical findings in children with ALL. Fever is the most common finding at diagnosis and is usually the result of the leukemic process. It generally resolves within 72 hours from the start of induction therapy (Westlake & Bertolone, 2002).

Choices b, blurred vision, and c, behavioral changes, are incorrect. These are signs and symptoms of CNS involvement and rarely are observed at the time of initial diagnosis (Margolin et al., 2002).

Choice d, enlarged, very painful lymph nodes, also is incorrect. Lymphadenopathy is a manifestation of extramedullary leukemic spread. The enlarged lymph nodes usually are painless or minimally tender and may be localized or generalized. Lymph nodes that are cancerous typically are firm, fixed, and nonpainful (Margolin et al., 2002).

**Author Contact:** Deborah Cagen, ARNP, MSN, CPON®, can be reached at Deborah.Cagen@mch.com.

**References**


