

Practice Exam Questions



CPHON

Certified Pediatric Hematology Oncology Nurse



EXAMAIDES

PASS YOUR EXAM AT FIRST TRY

Total Question: 165 QAs

Question No: 1

Common fears experienced by infants hospitalized for cancer or blood disorders include all of the following EXCEPT:

- A. separation from parents.
- B. loud noises.
- C. bright lights.
- D. slow movements.

Answer: D

Explanation: Infants have many fears, including separation from parents, strangers, loud noises, sudden movements, animals, and heights. Any of these can be exacerbated by hospitalization and disease.

Hospitalized infants need caring, nutrition, and support from caregivers. Thus, parents are encouraged to spend as much time as possible with their child, maintaining feeding and tactile stimulation as close to normal as possible. Examinations should not be prolonged, especially if they cause discomfort for the patient. Having a parent hold the child during examinations, if possible, is comforting to the infant. Assessment of the developmental level, using charts and guidebooks, is an essential nursing task for sick infants.

Question No: 2

Hospitalized preschoolers require all of the following EXCEPT:

- A. assessment of normal behavior.
- B. encouragement to play.
- C. separation from other children.
- D. brief, simple explanations of treatment.

Answer: C

Explanation: Preschoolers in the 4-6-year-old age-group have a sense of their identity and less fear of strangers than younger children. They have a more complete understanding of body parts and functions as well as the concepts of appropriate behavior and social interaction. Obtaining the pattern of normal routines and behavior from the parents is quite helpful in distinguishing hospital or disease-acquired regressive behaviors. Preschoolers should be encouraged to play, by themselves and with other children, barring dangerous exposures; however, viewing other children in pain is discouraged since it generates fear and anxiety. It is usually possible to explain medical procedures and treatments in an age-appropriate way to preschoolers that may reassure the anxious child.

Question No: 3

Teenagers with cancer or other serious diseases often fear all of the following EXCEPT:

- A. loss of body image.
- B. rebellious behavior.
- C. rejection by peers.
- D. loss of independence.

Answer: B

Explanation: Teenagers with serious diseases often fear the loss of peer acceptance more than the disease itself. They tend to exaggerate other's poor opinion of them and often become embarrassed by minor flaws.

Privacy is especially important. Risky or rebellious behavior may follow a serious diagnosis often in an effort to deny their disease and be more inviting to their peer group. They are caught in the classic conflict of the desire for independence and the loss of control of their own bodies. A poor self-image is often a problem. The nurse should encourage frank discussions of their care. Self-care as much as possible is encouraged. However, the nurse should explain that dependence on others during an illness is a sign of strength and maturity.

Question No: 4

Nursing interventions for the hospitalized toddler age-group (1-3 years of age) include:

- A. limiting mobility to prevent injury.
- B. assisting with toilet training.
- C. assessing the patient without assistance of a parent.
- D. enforcing standardized sleep patterns to ensure rest.

Answer: B

Explanation: Nursing interventions in toddlers should start with an assessment of the child's normal behavior and routine as well as an evaluation of developmental stages. A conference with the parents or other guardian is always advisable. Such items as fears, sleep patterns, security items and stage of toilet training are important in the evaluation of the patient and offer clues as to management of an ill child in an unfamiliar, even scary environment, such as the hospital or outpatient clinic. Mobility should be encouraged and the child's particular sleep patterns from home should be followed if possible. Evaluation of the influence that the disease and its treatment play in modifying behavior and attitude is especially important. This is of particular concern if there is disease of the nervous system (e.g., stroke, intracerebral infection or bleeding) or treatment induced abnormalities (e.g., drugs that influence the brain, intrathecal therapy, radiation effects). Pain is, of course, a major issue and must be addressed individually.

Question No: 5

The psychosocial factor LEAST likely to influence school-age children with cancer is:

- A. absence from school.
- B. loss of peer-group acceptance.
- C. inability to carry out previously acquired skills.
- D. separation from parents.

Answer: D

Explanation: School-age children have already begun to develop some independence from their parents, at least for certain periods of time, therefore they would handle separation from parents better than younger children. This may change in a hospital environment so that the child may regress to a needier stage and may be unable or unwilling to perform previously acquired skills and habits.

Absence from school and loss of peer group contact and acceptance are strong influences in modifying behavior. There are programs for school re-entry and maintenance of self-esteem and socialization that are useful despite changes in physical appearance (e.g., hair loss, prosthetic devices) or habits (e.g., enuresis, emotional control). Positive reinforcement of normal behavior and parent counseling are helpful in re-establishing usual patterns of acceptable behavior.

Question No: 6

In a child with cancer or chronic hematologic disease, family system assessment should include all of the

following except:

- A. occupation and education of family members.9:46 AM
- B. an analysis of family dynamics and coping mechanisms.
- C. cultural and religious traditions.
- D. protecting family members from information that may be painful to hear.

Answer: D

Explanation: A detailed analysis of the family structure and dynamics is essential to the support of a pediatric patient with a serious illness. Often truthful and direct answers to questions about the disease and its care go a long way to reassure parents and siblings. Answer choices A, B, and C are all important variables in evaluating and supporting family members attitudes and coping mechanisms. This is especially true for situations in which aggressive treatment is required, relapse occurs, or end-of-life care is necessary. It is often the nurse who must explain the medical diagnosis and the treatment rationale to family members. In such stressful situations, misinformation may be devastating to patients. Today, many families will get their medical information from external sources (e.g., books, magazines, Internet), and it is critical that they have a competent and trustworthy source that can assist in the understanding and management of a complex situation.

Question No: 7

All of the following factors impact the need for social support in pediatric patients with cancer or blood disease EXCEPT:

- A. parents' work obligations.
- B. physician's personality or specialty.
- C. medical insurance and family finances.
- D. residential location.

Answer: B

Explanation: Numerous factors must be taken into account in achieving social support for the family. Often this requires the assistance of social services or other outside religious or charitable agencies.

Single-parent families, number and care of siblings, work schedules, and other outside obligations as well as medical insurance and family finances are obvious factors that require inquiry and remediation, if necessary. Plans for transport to and from the hospital or physician's office should be established. Accessibility of ambulance services for emergencies is also a consideration.

Physicians rarely play much of a role in securing social support networks; it is usually up to the nurse or social worker to coordinate and request assistance from appropriate sources.

Question No: 8

Recent polls have indicated that the percentage of children younger than 18 in the United States without health insurance is:

- A. 8.5%.
- B. 15%.
- C. 25%.
- D. 40%.

Answer: A

Explanation: Socioeconomic status greatly influences medical insurance coverage in the United States. Much family coverage is obtained through parental employment or federal and state government programs, such

as Medicaid; however, there are a significant number of children who are uninsured (8.5%). It should be noted that children with serious diseases, though cured, may be denied medical coverage based on "pre-existing conditions." This situation will change if health care reforms are passed by Congress. The nurse must inquire about insurance coverage or the ability to pay for expensive medical care and be cognizant of possible solutions.

Question No: 9

Breaches of professional boundaries between a nurse and patient or family include all of the following EXCEPT:

- A. sharing feelings of sympathy or grief.
- B. using terms of endearment.
- C. non therapeutic physical contact.
- D. accepting gifts.

Answer: A

Explanation: There are numerous occasions when the pediatric nurse is tempted to violate professional standards with respect to a patient or the family. This is particularly true in hematology- oncology pediatric nursing where terminal diseases and harsh treatments of children are common. However, there are many situations that must be avoided: calling the patient dear, honey, or sweetie are examples of unacceptable terms of endearment; self-disclosure of personal matters; giving or receiving gifts; favoring one patient's care over another; intimate relationships with a patient or family member; and many others. Expressing sympathy or grief to the family or patient is permissible if the nurse is not overtly personal.

Question No: 10

In preparing a child or teenager for return to school, which of the following is LEAST indicated?

- A. a conference with the school nurse regarding the patient's condition.
- B. education of teachers regarding the patient's condition and requirements.
- C. a conference with cafeteria staff regarding limitations in the patient's diet.
- D. education of classmate's regarding the patient's condition in simple terms.

Answer: C

Explanation: Assessing the patient's previous school history, such as home schooling, frequent absences or exclusion from school, or extracurricular activities, is important in planning the often-difficult return to school. A conference with the school nurse about physical limitations and the medication schedule is usually helpful. Classroom teachers should also be advised regarding management of the affected child and his or her interaction with schoolmates. Is there a likelihood of learning disabilities due to the disease treatment? Will there be a need for absences for doctor's appointments or scheduled treatments? Should there be a tutor for "catch-up" or ongoing education? Education of classmates via simple explanations or graphic aids is often advisable to minimize taunting or exclusion of the patient. Generally, cafeteria personnel are not responsible for knowing every student's dietary restrictions, but including the element of dietary restrictions in the education of the patient, family, school nurse, and teacher would be beneficial.

Question No: 11

A 5-year-old child is admitted for chemotherapy and expresses fear of pain. In this situation, a nurse would:

- A. arrange an explanation of the procedure by the physician.
- B. minimize the amount of pain involved.
- C. try to divert the child 's fear with games and activities.

D. arrange for the patient to socialize with patients undergoing similar painful treatments.

Answer: C

Explanation: Reassuring young children who are ill is often quite difficult. The nurse should not be tempted to divert this task to others, such as parents, other nurses, or physicians. Honesty is often the best solution, especially when expressed in a kind and caring way. Minimizing the pain of an unpleasant procedure leads to a lack of trust and may complicate future cooperation. Simple brief explanations are best, taking into account the child's level of understanding and poor sense of time.

Often a child's fears may be diverted or overridden with games or favorite activities, such as reading, singing, or drawing. Socialization is generally a good idea, but the patient should not be allowed to see other children in pain. Self-care that is appropriate for age and disease limitations should be supported.

Question No: 12

Cultural or spiritual factors that may interfere with effective treatment or communication include which of the following EXCEPT:

- A. language barriers.
- B. religious beliefs.
- C. ethnic traditions.
- D. education level.

Answer: D

Explanation: Cultural and spiritual factors may influence an understanding of a disease and its prognosis or the acceptance of treatment by the patient or family. These may include ancestry, ethnic group, minority status, and religious beliefs. The population of the United States is now quite diverse, and many languages and modes of behavior or beliefs are encountered in a hospital environment. If there is a language barrier, it is important to be certain that there is a competent translator available early in the hospital stay. Beliefs may be based on religion (e.g., Jehovah's Witnesses, Christian Scientists, and faith healers), ethnicity, or ancestry (e.g., belief in Asian or Native American healing methods) or a desire for nontraditional or complementary practices (e.g., vitamins, strange foods, diets). Cultural or spiritual factors do not correlate with education level therefore should not be considered or assumed. Respect for these beliefs is essential, but a careful explanation is indicated if there is potential for harm.

Question No: 13

Adolescents and young adults should be counseled on matters of heredity, sexuality, and life goals, including all of the following EXCEPT:

- A. limitations on ambition or life planning.
- B. birth control, HIV, and protection from sexually transmitted diseases.
- C. reproductive decisions in patients with inherited diseases (e.g., hemoglobinopathy, hemophilia).
- D. the influence of cancer or its treatment on fertility and reproductive capacity.

Answer: A

Explanation: Certain blood diseases and cancers are influenced by heredity. While detailed analyses and reproductive advice should be left to a specialist in genetic counseling, adolescent and young adult patients should have some awareness of the effect their disease may have on their future offspring.

Many adolescent and young adult patients with cancer will be cured, but treatment with radiation or chemotherapy may diminish fertility. Techniques for sperm and ovary preservation are now common and should be explained to patients when appropriate. Genetic background is important in assessing susceptibility

to some cancers and in response to therapy. This will only increase as progress is made toward "personalized medicine." It is unwise to discourage planning for life goals or ambitions since long-term cures are increasingly common.

Question No: 14

In preparing a child for a painful procedure, the nurse should:

- A. tell the child that the procedure will not hurt.
- B. return an infant or young child to the crib for the procedure.
- C. explain how long the procedure will last with a time reference the child understands.
- D. avoid having the parents present since they will likely interfere or distract the nurse, physician, or technician.

Answer: C

Explanation: Preparing a pediatric patient for psychologically unsettling or painful procedures, such as diagnostic procedures (e.g., blood tests, x-rays, scans, physical examinations), is important. The procedure should be explained, and if age appropriate, the patient should be informed as to why the exam is necessary. The crib should be a place of calm and safety so procedures and examinations should be done elsewhere. Medical slang should be avoided. Young children are often inquisitive about the duration of a procedure; this may be explained with reference to a familiar item, such as the duration of a TV show, a class in school, or the length of the lunch period. Parents should also be prepared for the procedure and encouraged to be present.

Question No: 15

Internal sources of support for children with cancer and their families include which of the following?

- A. literature and lectures from disease-specific organizations.
- B. patient and survivor web sites.
- C. community support groups.
- D. on unit physical therapy.

Answer: D

Explanation: On unit physical therapy is the only example of an internal source of support for children with cancer and their families. The remaining sources are examples of external sources of support that are available outside of the hospital. Numerous web sites are also available for all aspects of pediatric cancers, including the National Cancer Institute and the American Cancer Society.

Information about clinical trials and new or experimental treatments are easily obtained from an Internet search. Many organizations maintain lecture series and telephone counseling. Most major libraries have volumes on pediatric cancer. Social service organizations are also of value in arranging financial, ambulatory, or psychological support. Community support groups are helpful and available in local or nearby communities. Child psychiatrists or psychologists may be useful in assisting survivors with coping mechanisms and returning to former activities. Clinical depression is increasingly diagnosed in children and adolescents and the experience of having cancer or a serious blood disorder may promote or bring out psychiatric or psychosocial disorders.

Question No: 16

Teenagers with cancer should be encouraged to carry out all the following EXCEPT:

- A. take an active role in the management of their disease.
- B. retain and develop social relationships.
- C. avoid seeking information about their disease and its complications.

D. maintain a positive body image.

Answer: C

Explanation: One of the most important aspects of psychosocial support for teenagers is to encourage them to take an active role in the management of their disease. This includes direct discussion with caregivers, responsibility for taking medications, and keeping appointments on time, following instructions about how to watch for and communicate drug side effects, or signs and symptoms of the underlying disease. They should also retain friendships and participate in former activities (or develop new interests) to the extent possible. Maintenance of a positive body image is crucial for this age-group and should be emphasized by parents and caregivers. Such visible physical side effects of disease or treatment, such as alopecia, abnormal fat distribution, swollen glands, or facial lesions, may be addressed cosmetically. It is important not to hide the details of the disease or prognosis and to encourage the patient to seek information about the condition. Questions or concerns may then be discussed directly with caregivers.

Question No: 17

The most common malignancy in children is:

- A. brain tumor.
- B. leukemia.
- C. lymphoma.
- D. Ewing's sarcoma.

Answer: B

Explanation: Leukemia is the most common malignancy in children with a preponderance of acute lymphoblastic leukemia. Acute myelocytic leukemia is less common, accounting for only about 20% of cases. Even less common are the promyelocytic and myelomonocytic leukemias and myelodysplasia, a preleukemic condition found mostly in adults. The chronic leukemias are far less common in the pediatric age-groups. Brain tumors are the commonest solid malignant tumor in children, but even benign tumors can considerably damage a developing brain. Lymphomas are also fairly common with non-Hodgkin's lymphoma more frequently seen in the younger age-groups while Hodgkin's lymphoma is commoner in the 15- 17-year-old age-group. Ewing's sarcoma, a type of bone tumor found in children, is far less common than the others mentioned above.

Question No: 18

A 16-year-old boy presents with pain in the knee area and a mild limp of several weeks duration. He has otherwise been active and well, and there is no history of trauma. He is afebrile and his complete blood count is normal. There is no calf tenderness or inflammation. Plain radiographs reveal periosteal new bone formation and destruction of preexisting cortical bone in the distal femur. A soft-tissue mass is present. The most likely diagnosis is:

- A. a popliteal cyst.
- B. osteogenic sarcoma.
- C. acute osteomyelitis.
- D. deep vein thrombosis.

Answer: B

Explanation: Osteogenic sarcoma is the most common malignant bone tumor in the pediatric- adolescent age-group. Boys tend to be older than girls at diagnosis, possibly because of their later skeletal maturation. The symptoms and laboratory findings described are typical for this tumor. Plain x-rays of the painful area

should be the first diagnostic procedure and usually show bone destruction and a soft tissue mass. Computed tomography scan of the chest to search for possible metastatic disease is important. Treatment involves limb-sparing surgery or amputation along with multidrug chemotherapy. Cure rates as high as 50%-70% have been reported for non metastatic disease. A popliteal cyst may cause pain and swelling behind the knee, but there is no evidence of bone destruction. Osteomyelitis is unlikely because there is no history of leg injury, fever, or signs of inflammation at the painful site. Deep vein thrombosis is also unlikely because there is no calf tenderness, leg swelling, injury, or prolonged inactivity.

Question No: 19

In sickle cell anemia, the most dominant hemoglobin (Hb) type is:

- A. Hb A.
- B. Hb S.
- C. Hb F.
- D. Hb A1C.

Answer: B

Explanation: Sickle cell anemia, found predominantly in African Americans, is inherited as an autosomal recessive trait. Normal hemoglobin (Hb A) has two polypeptide chains (alpha and beta) in which the sixth position on the beta chain contains glutamic acid. In Hb S, the sixth position is replaced by the amino acid valine. This causes the red blood cells to become adherent to each other and form discoid (sickle) shapes. Normal red cells are biconcave and pliable; sickle red cells are far less flexible and tend to obstruct small blood vessels. The sickle phenomenon may be provoked by hypoxia, infection, dehydration, or fever. In patients with sickle cell anemia, there is an increased level of Hb F, or fetal hemoglobin, that does not cause sickling and partially compensates for the predominant Hb S. Glycosylated hemoglobin, or Hb A1C, is measured to diagnose diabetes and plan for its treatment.

Question No: 20

All of the following statements about Ewing's sarcoma are true EXCEPT:

- A. it is far more common in Caucasian than African-American children.
- B. it is of neural crest origin.
- C. it is relatively resistant to radiation therapy.
- D. plain x-rays of bone often reveal a "moth-eaten" or "onion skin" appearance.

Answer: C

Explanation: Ewing's sarcoma is a prominent member of a class of childhood cancers that arises mostly in bone; it is radiosensitive but may also be sensitive to chemotherapy. The tumor is of neuroectodermal origin and is one of a class of these tumors, some of which arise in soft tissue. It is second to osteogenic sarcoma as the most frequent childhood bone tumor. Pain at the tumor site is the usual presentation, often present for weeks to months and described as increasing in severity.

Plain x-rays reveal a moth-eaten appearance of bone at the site of the tumor, and there may also be an onion-skin appearance to the area due to periosteal reaction. Metastatic disease to the lungs may be present in which case pulmonary radiation may be curative. The overall cure rate is 60%-70% with distal lesions having a better prognosis. Metastatic disease is curable in 18%-30% of cases.

Question No: 21

Childhood cancer differs from adult cancer in all of the following ways EXCEPT:

- A. it is far less common than in adults.
- B. lymphomas and sarcomas are commoner than carcinomas.
- C. the prognosis is considerably better than for adults.
- D. the disease is usually localized at diagnosis.

Answer: D

Explanation: Childhood cancer tends to be distinct from adult disease in several important features. It is far less common (< 1 % of all cancers). Leukemias, lymphomas, and sarcomas predominate in children, and carcinomas predominate in adults. There is a higher incidence of cancer in white American children, whereas African-Americans have a higher rate in the adult forms. Prognosis for the childhood cancers is better than for adults (e.g., a 70%-90% 5-year survival rate for children as compared to a less than 60% rate for adult cancers). This may stem from the higher incidence of leukemias and lymphomas in the pediatric age-group, diseases for which aggressive therapy has been quite successful. Another difference is that childhood disease is more likely to be metastatic at diagnosis, again possibly related to the predominance of leukemia and lymphoma, while adult disease tends to be localized at the time of diagnosis.

Question No: 22

The mother of a 3-year-old boy discovers, while giving him a bath, that he has a nontender abdominal mass. He has not complained of pain, and he has not lost weight; there have been no changes in bowel function, fevers, or hematuria. His pediatrician confirms the presence of a smooth, non tender, left abdominal mass extending to the midline. The rest of the examination is normal. An abdominal ultrasound reveals a solid left renal mass but no other abdominal lesions. A computed tomography scan shows a normal right kidney. A chest x-ray is negative. The most likely diagnosis is:

- A. Wilms' tumor (nephroblastoma).
- B. metastatic carcinoma.
- C. renal cell carcinoma.
- D. renal cyst.

Answer: A

Explanation: Wilms' tumor represents about 6% of childhood cancers. It is usually diagnosed in children around 3-4 years of age. The usual presentation is an asymptomatic abdominal mass with few other clinical findings. Abdominal ultrasound with Doppler is the best initial diagnostic test. The tumor may spread locally or hematogenously to the liver or lungs. The preferred treatment is surgical excision, including nephrectomy, renal hilar structures, and partial ureterectomy, followed by chemotherapy with or without radiation therapy. The current cure rate exceeds 85%. The tumor may occur bilaterally in 5%-10% of patients. Metastatic carcinoma to the kidney is very rare in children, and there often will be evidence of a primary tumor. Pediatric renal cell carcinoma may present with an abdominal mass, but pain, hematuria, and constitutional symptoms are more common as are distant metastases to the lung, liver, or bone. A renal cyst may also be a cause of an abdominal mass, but ultrasound findings should be diagnostic.

Question No: 23

Cancer cell proliferation differs from healthy cell replication in all of the following ways except:

- A. evading apoptosis (programmed cell death).
- B. tissue invasion and metastasis.
- C. sustained angiogenesis.
- D. presence of telomeres.

Answer: D

Explanation: Cancer cells differ from normal cells in several important ways, often with a genetic cause but occasionally resulting from a viral infection. Cancer cells have a nearly unlimited ability to replicate. In normal cellular mitosis, a small portion of the telomere (the replicative DNA at the end of a chromosome) is lost and ultimately accumulation of these losses leads to cell senescence and cellular death. Cancer cells have a telomerase enzyme that prevents this loss and allows almost indefinite cellular replication. Apoptosis refers to the intrinsic programming of cellular death; cancer cells may avoid this and continue grow. Angiogenesis refers to the ability of tumors to stimulate blood vessel proliferation and thus sustain their nutritional and oxygen supply. Tissue invasion and capacity to spread to distant anatomical sites is a characteristic property of most cancers and is often the lethal mechanism of action.

Question No: 24

A 4-year-old child was found to have a painless mass in his neck. According to his mother, he recently complained of feeling bad and had lost weight; she also reported that her child had experienced intermittent fevers over the past 4-6 months. Scalp nodules were present as was a posterior mediastinal mass on chest x-ray. Biopsy of the neck mass revealed neuroblastoma. All of the following statements about this disease are true EXCEPT:

- A. urinary catecholamines may be elevated.
- B. bone marrow aspiration and biopsy are a necessary part of the evaluation.
- C. this tumor is rare in children over 10 years of age.
- D. metastatic disease is uncommon at the time of diagnosis.

Answer: D

Explanation: Neuroblastoma is the most common extracranial pediatric solid tumor, accounting for 8%- 10% of childhood cancers. The tumor is rare in children over 10 years old. Metastatic disease is present in 75% of patients at the time of diagnosis. The most common findings are masses in the neck, posterior mediastinum, or abdomen. Constitutional symptoms and weight loss over a period of months are often noted. The tumor arises from neural crest cells of the sympathetic nervous system, and urinary catecholamines are elevated in most cases. It also may secrete vaso- and neuroactive peptides that can lead to gastrointestinal or neurologic symptoms. Bone marrow aspiration and biopsy often detect metastatic disease, and radionuclide skeletal scintigraphy can be helpful in staging. Localized disease may be surgically resected with good results or treated with radiation therapy if inoperable. Combination chemotherapy is the mainstay of treatment since the disease is most often widespread at the time of diagnosis.

Question No: 25

An 8-year-old boy is experiencing right ocular proptosis with facial swelling and vision loss. Magnetic resonance imaging reveals a large retro-orbital mass displacing the globe and invading the adjacent soft tissue. Pathologic diagnosis of the lesion is rhabdomyosarcoma.

All of the following statements about this disease are true EXCEPT:

- A. this tumor may arise in any soft tissue, but the extremities are then most common site.
- B. the 5-year survival rate is over 70%.
- C. surgery, radiation therapy, and chemotherapy are often used in treatment protocols.
- D. combination chemotherapy with vincristine, dactinomycin, and sometimes cyclophosphamide has proved successful.

Answer: A

Explanation: Rhabdomyosarcoma is the most common soft tissue sarcoma in the pediatric age-group, representing about 3% of childhood cancers. It can arise in almost any tissue in the body but the head and neck are most frequently affected, followed by the genitourinary system and the extremities. Surgical excision is the mainstay of localized disease. About 15%- 20% of patients have distant metastatic disease at the time of diagnosis, and another 10%- 15% have localized nodal involvement with tumor. Patients with orbital and nonparameningeal head and neck tumors tend to do better (favorable sites) as compared to patients with parameningeal, prostate/bladder, and extremity lesions (unfavorable sites). Patients with the so-called embryonal histologic subtype also do better than those with the alveolar subtype. Radiation therapy is often used for residual postsurgical disease, and chemotherapy with two or three drugs is used because of the assumption that all patients with this tumor have micro metastatic disease.

Question No

26. All of the following statements about acute lymphoblastic leukemia (ALL) are true EXCEPT:

- A. there is an increased incidence of ALL in children with Down syndrome.
- B. the presence of a Philadelphia chromosome is a favorable prognostic sign.
- C. slow response to remission-induction therapy is a poor prognostic sign.
- D. the 5-year survival of children with ALL is now over 80%.

Answer: B

Explanation: Children with Down syndrome (trisomy 21) have a 10- 20-fold increased incidence of leukemia, both acute lymphoblastic leukemia and acute myelocytic leukemia. Other genetic defects associated with an increased risk are ataxia telangiectasia, Fanconi anemia, Wiskott-Aldrich syndrome, pure red cell aplasia, and neurofibromatosis. The presence of a so-called Philadelphia chromosome, a reciprocal translocation between chromosome 9 and 22, confers a poor prognosis to the leukemic patient. In general, patients who do not respond promptly to remission-induction therapy with clearing of all leukemic cells and infiltrates have a poor prognosis as well and may be candidates for allogenic bone marrow transplantation. The 5-year survival rate is now over 80% with several different three-phase treatment protocols.

Question No: 27

What coagulation factors are missing or reduced in hemophilia A?

- A. Factor VII.
- B. Factor VIII.
- C. Prothrombin.
- D. Factor IX.

Answer: B

Explanation: Hemophilia is an X-linked genetic disorder in which coagulation factors VIII (known as hemophilia A, 85%) or IX (known as hemophilia B, 10%- 15%) are missing or reduced. Due to its Xlinkage, the disease is most common in sons of female carriers. About 30% of persons with the disease have no family history, and presumably the missing coagulation factor is a result of a mutation. Excessive bleeding may occur after circumcision or minor trauma. Bleeding from the synovium into a joint is very common and may lead to chronic arthropathy if left untreated. Other areas of hemorrhage include muscles, oral cavity, urinary tract, or gastrointestinal tract.

Intracranial bleeding is less common (1 %- 2%). Both factors VIII and IX are protein coagulation factors in the intrinsic pathway of coagulation, and deficiency may prolong the partial thromboplastin time. Factor VII and prothrombin deficiency tend to prolong the prothrombin time and may be acquired or congenital.

Question No: 28

Which of the following is NOT considered to be a site of extramedullary (nonhematologic) leukemic infiltration?

- A. Central nervous system.
- B. Spleen.
- C. Lymph nodes.
- D. Bone marrow.

Answer: D

Explanation: Leukemic infiltrates may occur in many organs and occasionally are the presenting clinical finding. The term "extramedullary" refers to sites outside of the bone marrow, therefore this is not a correct option. Common sites of extramedullary leukemic infiltration are the liver, spleen, lymph nodes, and thymus. Testicular involvement is not unusual in acute lymphoblastic leukemia. Acute myelocytic leukemia may infiltrate the skin, oral cavity, head and neck area, or form myeloblastomas (chloromas) that resemble solid tumors and may be found in skin, soft tissue, bone, or other organs. Sometimes leukemia is asymptomatic and discovered on a routine blood count. The presence of leukemic cells or more than five white blood cells in the cerebrospinal fluid suggests central nervous system or meningeal involvement as does a cranial nerve palsy. This may result in a relapse from this location, and intrathecal chemotherapy is required.

Question No: 29

Which of the following statements about the incidence of Hodgkin's lymphoma in children in the 15-18-year-old age-group is true?

- A. It is higher than for non-Hodgkin's lymphoma.
- B. It is lower than for non-Hodgkin's lymphoma.
- C. Both Hodgkin's and non-Hodgkin's lymphoma occur with an equal incidence.
- D. It does not occur in this age-group.

Answer: A

Explanation: In children under 15 years of age, lymphomas of the non-Hodgkin's type occur more frequently. However, if one takes into account all lymphomas in children and adolescents under the age of 18, there is a slight predominance of the Hodgkin's type. Lymphomas of both the Hodgkin's and non-Hodgkin's types together represent the third most frequent malignancy in childhood.

There is some geographical distribution in the subtypes of non-Hodgkin's lymphoma, mostly the increased frequency of Epstein-Barr virus-associated Burkitt's lymphoma in equatorial Africa and Brazil, which is far more common than in the United States. The incidence is also increased in children with congenital and acquired immunodeficiency syndromes, including those with immunosuppressive treatment after organ transplantation.

Question No: 30

A staging workup of a child with non-Hodgkin's lymphoma reveals disease in the cervical lymph nodes and enlargement of the hilar lymph nodes on computed tomography scan of the chest. No mediastinal masses are seen, and the rest of the examination is negative. This clinical picture is consistent with:

- A. stage I.
- B. stage II.
- C. stage III.
- D. stage IV.

Answer: B

Explanation: Staging of non-Hodgkin's lymphoma is usually based on criteria developed by the St. Jude's system. This includes four stages I- IV. The stage of the disease has major implications for both prognosis and treatment. Some also include the absence (A) or presence (B) of symptoms such as fever, night sweats, pruritus, or significant weight loss, but this is used more often in Hodgkin's lymphoma. The total burden of disease is perhaps the most significant prognostic factor and may correlate with serum lactate dehydrogenase and serum interleukin-2 receptor levels as well as stage and histologic type. Generally, stage I disease is limited to a single nodal or extranodal area; stage II reflects two nodal or non-nodal areas on the same side of the diaphragm; stage III indicates disease on both sides of the diaphragm or any intra thoracic tumor or extensive intra-abdominal disease; and stage IV indicates involvement of the central nervous system or bone marrow.

Question No: 31

Autoimmune hemolytic anemia is associated with all of the following EXCEPT:

- A. increased hemoglobin levels.
- B. positive Coombs' test.
- C. medications.
- D. systemic lupus erythematosus.

Answer: A

Explanation: Autoimmune hemolytic anemia is a disorder in which red blood cells are hemolyzed due to adherence of antibodies to the cell membrane. The normal life span of erythrocytes (100-120 days) is shortened. Hemolysis may occur intravascularly or extra-vascularly (most prominently in the spleen where phagocytosis of antibody or complement-coated red cells occurs). Most cases are idiopathic, but the disorder may also be associated with systemic lupus erythematosus, certain malignancies, infections, or medications (such as cephalosporins). Brisk intravascular hemolysis may cause fatigue, pallor, jaundice, and tachycardia, and hepatosplenomegaly may be prominent.

Hemoglobin is low, the reticulocyte count is elevated, and the direct Coombs' test is usually positive; that is, there is clumping or agglutination of red cells after washing and exposure to anti- human globulin, also known as Coombs' reagent.

Question No: 32

In the workup of a child suspected of having acute leukemia, which of the following tests would most accurately diagnose the disease?

- A. test for Epstein-Barr virus.
- B. blood smear.
- C. bone marrow aspiration.
- D. lumbar puncture.

Answer: C

Explanation: Sometimes infectious mononucleosis can masquerade as leukemia with a sore throat, fever, lymphadenopathy, and splenomegaly along with an abnormal white blood cell count and atypical lymphocytes on the blood smear. A test for Epstein-Barr virus is usually diagnostic, but occasionally there are false-positive and false-negative results. A pancytopenia may have several causes. A clearcut leukemic pattern on the peripheral blood smear may or may not be present. A lumbar puncture (spinal tap) may or may not show a leukemic infiltrate or abnormalities in glucose or protein content. The surest diagnostic procedure is a bone marrow examination in which leukemia can usually be differentiated from such disorders as

myelodysplasia, aplastic anemia, or leukemoid reaction.

Question No: 33

The most reliable diagnostic test for Hodgkin's lymphoma is:

- A. positive test for Epstein-Barr virus.
- B. the presence of Reed-Sternberg cells on a lymph node biopsy.
- C. the presence of enlarged cervical and axillary lymph nodes on physical examination.
- D. the presence of a mediastinal mass on a computed tomography scan.

Answer: B

Explanation: The hallmark diagnostic criterion of Hodgkin's disease is the presence of the eponymous Reed-Sternberg cells, almost always found on a lymph node biopsy. These are giant cells with polypoid or multiple nuclei and eosinophilic nucleoli. These represent a minority, only 1 %-5% of the cell population against a background of small lymphocytes and fewer other cell types such as granulocytes and fibroblasts. Immunochemical markers, especially positive for CD 15 and CD 30 are also helpful in distinguishing Hodgkin's from non-Hodgkin's lymphomas. There are four main histologic subtypes that are important in prognosis and treatment decisions. In addition to these, a so-called nodular lymphocyte- predominant form occurs with tightly packed nodules under lowpower microscopy.

Question No: 34

Brain tumors are the second most common neoplasm in children and the first among solid tumors. Of these, the most frequently diagnosed is:

- A. astrocytoma.
- B. medulloblastoma.
- C. ependymoma.
- D. craniopharyngioma.

Answer: A

Explanation: Brain tumors in the pediatric age-group may present aggressively or slowly and be hard to diagnose. Many symptoms are related to increased intracranial pressure, which typically causes morning headache, nausea, vomiting, ataxia, nystagmus, or diplopia. Cranial nerve palsies or seizures may be a presenting complaint. Sometimes symptoms are subacute, such as declining academic performance, cognitive deficits, and intermittent headaches. Bulging fontanelles and limited upward and downward deviation of gaze may occur in infants. Magnetic resonance imaging and magnetic resonance angiography are key imaging techniques to determine the size, location, and vascularity of the tumor. Positron emission tomography may also be used to determine the metabolic activity of the tumor. Metastatic disease is rare except for medulloblastomas, which may invade bone marrow or viscera via the central nervous system.

Question No: 35

Remission-induction therapy for an 8-year-old child with acute lymphoblastic leukemia may include all of the following except:

- A. intrathecal chemotherapy.
- B. asparagine.
- C. steroids.
- D. anthracyclines.

Answer: B