Certified Pediatric Hematology Oncology Nurse (CPHON) Practice Exam (Sample)

Study Guide

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Questions

1. At what age do symptoms of Diamond-Blackfan Anemia typically present?

A. At birth

B. Between 6 months and 1 year

C. After the age of 2

D. In adolescence

2. Chronic neutropenia in children with Schwachman-Diamond Syndrome increases the risk for which of the following conditions?

A. Aplastic anemia

B. Rheumatoid arthritis

C. Bone demineralization

D. Asthma

3. Which is an accurate estimate for the incidence of Diamond-Blackfan Anemia?

A. 1,000 cases per year worldwide

B. 400 cases per year worldwide

C. 2,000 cases per year worldwide

D. 800 cases per year worldwide

4. What is the typical classification of Hb Sbeta0 Thalassemia?

A. Usually mild

B. Usually severe

C. Carrier state

D. Moderate

5. Schwachman-Diamond Syndrome is characterized by defects in which type of immune cells?

A. Red blood cells

B. Neutrophils only

C. B and T cells

D. Platelets

6. What approach is recommended if a medication is suspected to cause AIHA?

A. Increase the dosage

B. Switch to an alternative medication

C. Stop the medication

D. Consult a specialist

7. Leg ulcers in SCD patients typically result from:

A. Poor perfusion of the skin

B. Infection from insect bites

C. Autoimmune responses

D. Nutritional deficiencies

8. Which symptom might indicate a bleeding disorder associated with Schwachman-Diamond Syndrome?

A. Chronic cough

B. Bloody emesis

C. Joint pain

D. Rashes

9. Which complication of Sickle Cell Disease is characterized by a sudden decrease in blood flow to the brain?

A. Acute Chest Syndrome

B. Cerebral Vascular Accident (Stroke)

C. Spleen Sequestration

D. Aplastic Crisis

10. What effect does expanding marrow have in patients with untreated beta thalassemia?

A. Increased energy levels

B. Malocclusion of teeth

C. Reduced risk of infections

D. Enhanced immune response

Answers

1. B
2. C
3. B
4. B
5. C
6. C
7. A
8. B
9. B
10. B

Explanations

1. At what age do symptoms of Diamond-Blackfan Anemia typically present?

A. At birth

B. Between 6 months and 1 year

C. After the age of 2

D. In adolescence

Diamond-Blackfan Anemia (DBA) is a rare type of macrocytic anemia that is often characterized by insufficient red blood cell production due to a failure of the bone marrow to produce these cells adequately. The timing of symptom onset is a critical aspect of understanding and diagnosing this condition. Typically, symptoms of Diamond-Blackfan Anemia present between 6 months and 1 year of age. During this period, infants may exhibit signs of anemia such as pallor, fatigue, and growth retardation, primarily due to the body's inability to produce enough red blood cells. Identifying DBA at this early age is crucial because timely diagnosis and intervention can significantly impact the management of the condition, including potential transfusions or the administration of corticosteroids. Although some symptoms might be traced back to birth or later stages of childhood, the most characteristic and definitive presentation occurs within the 6 months to year timeframe. This understanding assists healthcare providers in recognizing the disorder early and initiating appropriate care for affected children.

2. Chronic neutropenia in children with Schwachman-Diamond Syndrome increases the risk for which of the following conditions?

A. Aplastic anemia

B. Rheumatoid arthritis

C. Bone demineralization

D. Asthma

Chronic neutropenia, particularly in conditions such as Schwachman-Diamond Syndrome, leads to multiple challenges due to impaired immune function. Children with this syndrome often have a compromised ability to fight infections because neutrophils, a type of white blood cell essential for combating infections, are diminished. This persistent low level of neutrophils can also affect other aspects of health, including the maintenance of bone integrity. Bone demineralization is often a consequence of prolonged periods of infection or inflammation, which is common in children with neutropenia. The focus on chronic inflammation and the need for the body to rely on alternative mechanisms to compensate for the lack of neutrophils can result in a decrease in bone density over time. Therefore, bone demineralization is a concern in these patients as they may lead to an increased risk of fractures and other skeletal issues. While aplastic anemia, rheumatoid arthritis, and asthma are significant health concerns, they are not directly attributable to the chronic neutropenic condition associated with Schwachman-Diamond Syndrome in the same way that bone demineralization is.

3. Which is an accurate estimate for the incidence of Diamond-Blackfan Anemia?

A. 1,000 cases per year worldwide

B. 400 cases per year worldwide

C. 2,000 cases per year worldwide

D. 800 cases per year worldwide

Diamond-Blackfan Anemia (DBA) is a rare blood disorder characterized by the failure of the bone marrow to produce enough red blood cells. The estimated incidence of DBA is approximately 1 in 200,000 live births, leading to an overall incidence of around 400 new cases diagnosed each year worldwide. This statistic is derived from research and population studies documenting the prevalence and diagnosis rates of this specific anemia, making it a reliable figure in the medical literature. Understanding the incidence of DBA is important for pediatric oncology nursing, as it helps in providing appropriate care and resources for affected individuals.

4. What is the typical classification of Hb Sbeta0 Thalassemia?

A. Usually mild

B. Usually severe

C. Carrier state

D. Moderate

Hb Sbeta0 Thalassemia is classified as usually severe due to the significant reduction or absence of beta globin chains, which leads to a clinical presentation characterized by severe anemia and associated complications. The combination of sickle hemoglobin (Hb S) and the absence of beta globin results in aggressive sickling of red blood cells, leading to vaso-occlusive crises, splenic sequestration, and increased risk of infections. Patients with Hb Sbeta0 Thalassemia typically exhibit symptoms such as jaundice, fatigue, and growth impairment due to the severity of the anemia. Chronic hemolysis further exacerbates their condition, making regular transfusions and extensive medical management necessary to maintain hemoglobin levels and prevent complications. In contrast, other classifications such as usually mild or moderate do not accurately represent the debilitating nature of Hb Sbeta0 Thalassemia, and the carrier state does not involve the same symptomatic and clinical challenges faced by those with the condition. Therefore, the classification of Hb Sbeta0 Thalassemia as usually severe underscores the critical need for comprehensive care and intervention in these patients.

5. Schwachman-Diamond Syndrome is characterized by defects in which type of immune cells?

A. Red blood cells

B. Neutrophils only

C. B and T cells

D. Platelets

Schwachman-Diamond Syndrome is primarily characterized by defects in both B and T lymphocytes, which are crucial components of the adaptive immune system. These lymphocytes are responsible for orchestrating immune responses, with B cells producing antibodies and T cells playing a vital role in cell-mediated immunity. The defective functioning of these immune cells in Schwachman-Diamond Syndrome leads to a weakened immune response, making individuals more susceptible to infections. In this context, the other cell types mentioned do not represent the core issues associated with Schwachman-Diamond Syndrome. Red blood cells are primarily related to oxygen transport in the body, while platelets are involved in blood clotting. Neutrophils, as part of the innate immune system, are also important, but the key characteristics of Schwachman-Diamond Syndrome specifically relate to the impairment of immune functions linked to B and T lymphocytes. Therefore, understanding the involvement of these adaptive immune cells is crucial for comprehending the pathology and clinical implications of the syndrome.

6. What approach is recommended if a medication is suspected to cause AIHA?

A. Increase the dosage

B. Switch to an alternative medication

C. Stop the medication

D. Consult a specialist

In cases where a medication is suspected to cause Autoimmune Hemolytic Anemia (AIHA), the recommended approach is to stop the medication. This is because the first-line management of drug-induced AIHA involves the immediate cessation of the offending agent. Continuing the medication could exacerbate the hemolytic process and worsen the patient's condition. Discontinuing the medication helps to halt the immune response that is triggered by the drug, allowing the patient's immune system and red blood cell levels to stabilize over time. While switching to an alternative medication or consulting a specialist may eventually be necessary steps in a broader treatment plan, they are not the immediate actions taken in response to a suspected medication-induced adverse effect. Additionally, increasing the dosage could potentially intensify the hemolytic process and is not a safe or effective approach in this scenario. Stopping the medication directly addresses the root cause of the AIHA, making it the most suitable action.

7. Leg ulcers in SCD patients typically result from:

A. Poor perfusion of the skin

B. Infection from insect bites

C. Autoimmune responses

D. Nutritional deficiencies

Leg ulcers in patients with sickle cell disease (SCD) are primarily attributed to poor perfusion of the skin, which is a direct consequence of the vaso-occlusive crises that characterize this condition. In SCD, sickled red blood cells can obstruct blood flow in small blood vessels, leading to ischemia and decreased oxygen delivery to tissues. This impaired perfusion negatively affects the healing processes of the skin, making ulceration more likely to occur, especially in areas subjected to trauma or pressure. On the other hand, while insect bites can lead to infections that may complicate the condition of already existing ulcers, it is not the primary cause of leg ulcers in SCD patients. Autoimmune responses and nutritional deficiencies can certainly impact overall health and wound healing but are not considered typical direct causes of leg ulcers specifically in the context of sickle cell disease. Thus, poor perfusion stands out as the primary factor due to the inherent pathophysiological processes associated with SCD.

8. Which symptom might indicate a bleeding disorder associated with Schwachman-Diamond Syndrome?

A. Chronic cough

B. Bloody emesis

C. Joint pain

D. Rashes

Bloody emesis is a significant symptom that may indicate a bleeding disorder associated with Schwachman-Diamond Syndrome. This condition is characterized by exocrine pancreatic insufficiency, bone marrow dysfunction, and an increased risk for hematological abnormalities, particularly those affecting platelet function and coagulation pathways. In individuals with bleeding disorders, gastrointestinal bleeding can manifest as bloody emesis, serving as a potential indicator of underlying hemostatic issues. In contrast, while chronic cough, joint pain, and rashes may be noted in various conditions or syndromes, they are not specifically indicative of bleeding disorders. Chronic cough is often associated with respiratory infections or conditions, joint pain could result from various forms of arthritis or other musculoskeletal issues, and rashes might indicate dermatological conditions or systemic diseases but do not directly relate to bleeding abnormalities.

9. Which complication of Sickle Cell Disease is characterized by a sudden decrease in blood flow to the brain?

A. Acute Chest Syndrome

B. Cerebral Vascular Accident (Stroke)

C. Spleen Sequestration

D. Aplastic Crisis

A Cerebral Vascular Accident (Stroke) is indeed characterized by a sudden decrease in blood flow to the brain. In patients with Sickle Cell Disease, the abnormal sickle-shaped red blood cells can obstruct blood vessels due to their rigidity and tendency to clump together. This obstruction can lead to ischemia, resulting in stroke. The reduced blood flow can cause damage to brain tissue, leading to neurological deficits or even death if not promptly treated. The occurrence of stroke in sickle cell patients highlights the importance of early recognition and management strategies aimed at preventing such complications. These patients may be at a higher risk for both ischemic stroke, which is caused by an obstruction of blood flow, and hemorrhagic stroke, which results from blood vessel rupture. Understanding this complication underscores the need for regular monitoring and possible interventions, such as blood transfusions or hydroxyurea therapy, to reduce the frequency of sickle cell crises and to prevent vascular occlusions.

10. What effect does expanding marrow have in patients with untreated beta thalassemia?

A. Increased energy levels

B. Malocclusion of teeth

C. Reduced risk of infections

D. Enhanced immune response

In patients with untreated beta thalassemia, the expansion of bone marrow occurs as a compensatory mechanism due to chronic hemolytic anemia. The bone marrow attempts to produce more red blood cells to counteract the ineffective erythropoiesis and the destruction of red blood cells. This expansion can lead to a number of skeletal abnormalities, one of which is malocclusion of teeth. As the marrow expands, it causes changes in the facial bones and the dental arch, which can affect the alignment and position of the teeth, leading to malocclusion. This is a common clinical manifestation in individuals with significant marrow expansion due to the chronic nature of their disease. Therefore, malocclusion of teeth is a direct consequence of the skeletal changes associated with the expansion of bone marrow in untreated beta thalassemia patients. The other options do not accurately reflect the impacts of bone marrow expansion in this context. Increased energy levels may not be seen due to ongoing anemia, while the risk of infections is typically increased due to complications from the disease and its treatment. Similarly, an enhanced immune response is not a consequence of marrow expansion in this condition.