McCance: Pathophysiology, 6th Edition

Chapter 28: Alterations of Hematologic Function in Children

Key Points – Print

SUMMARY REVIEW

Fetal and Neonatal Hematopoiesis

1. After 2 weeks of gestation, circulating erythrocytes play a major role in delivering oxygen to the tissues.
2. Erythropoiesis in the liver and, to a lesser extent, in the spleen and lymph nodes reaches a peak at about 4 months.
3. By the fifth month of gestation, hematopoiesis begins to occur in the bone marrow, and by the time of delivery it is the only significant site of hematopoiesis.
4. A biochemically distinct type of hemoglobin is synthesized during fetal life, including Gower 1, Gower 2, and Portland.

Postnatal Changes in the Blood

1. Blood cell counts tend to rise above adult levels at birth and then decline gradually throughout childhood.
2. The immediate rise in blood cell counts is the result of increased hematopoiesis during fetal life, trauma of birth, and cutting of the umbilical cord.
3. The active rate of fetal erythropoiesis is observed in the large numbers of reticulocytes in the peripheral blood of the full-term neonate.
4. Erythrocyte values are age dependent, and values in males and females are apparent in adolescence.
5. The lymphocyte count is high at birth, and continues to rise in some healthy infants during the first year of life.
6. Platelet counts in full-term neonates are comparable to platelet counts in children and adults.

Disorders of Erythrocytes

1. Iron deficiency anemia is the most common blood disorder of infancy and childhood; the highest incidence occurs between 6 months and 2 years of age.
2. HDN results from incompatibility between the maternal and the fetal blood, which may involve differences in Rh factors or blood type (ABO). Maternal antibodies enter the fetal circulation and cause hemolysis of fetal erythrocytes. Because the immature liver is unable to conjugate and excrete the excess bilirubin that results from the hemolysis, icterus neonatorum, or kernicterus or both can develop.

3. Kernicterus, which may result from other causes as well, results in increased breakdown of red blood cells or decreased liver output of enzymes.
4. Infections of the newborn, often acquired by the mother and transmitted to the infant, may result in hemolytic anemia.
5. G6PD deficiency is an inherited enzyme deficiency in erythrocytes that results in a disruption of a common pathway of glycolysis, shortening erythrocyte life span.
6. Hereditary spherocytosis is the most common of the hereditary hemolytic states in which there is no abnormality of hemoglobin. The basic defect is an undefined abnormality of the proteins or spectrins of the erythrocyte membrane in which affected cells are unduly permeable to sodium and acquire a characteristic structure.
7. SCD is a genetically determined defect of hemoglobin synthesis, inherited by an autosomal recessive transmission; it causes a change in the shape of a red blood cell that results in decreased oxygen or hydration. This disease is most common among Africans, blacks, and those of Mediterranean descent.
8. The thalassemias are a heterogeneous group of hereditary hypochromic anemias of varying severity. Basic genetic defects include abnormalities of messenger ribonucleic acid (mRNA) processing or deletion of genetic materials, resulting in a decrease in the chains for hemoglobin.

Disorders of Coagulation and Platelets

1. Hemophilia is a condition characterized by impairment of the coagulation of blood and subsequent tendency to bleed. The classic disease is hereditary and limited to males, being transmitted through the female to the second generation. Many similar conditions attributable to the absence of various clotting factors are recognized.
2. von Willebrand disease is a dominantly inherited disease characterized by a vascular abnormality that produces a prolongation of bleeding time and by decreased levels of clotting factor VIII. The platelets in von Willebrand disease have decreased adhesiveness because the plasma factor is absent.
3. Disorders of congenital hypercoagulability and thrombosis include protein C deficiency, protein S deficiency, neonatal purpura fulminans, and antithrombin III deficiency.
4. The acquired antibody-mediated hemorrhagic diseases include ITP, autoimmune neonatal thrombocytopenia, and autoimmune vascular purpura.
5. ITP, the most common of the childhood thrombocytopenic purpuras, is a disorder of platelet consumption in which antiplatelet antibodies bind to the plasma membranes of platelets. This results in platelet sequestration and destruction by mononuclear phagocytes at a rate that exceeds the ability of the bone marrow to produce them.
6. Autoimmune neonatal thrombocytopenia is an antibody-mediated disorder that occurs in either autoimmune or alloimmune form.

7. The autoimmune vascular purpuras (allergic purpuras) are caused by the body's responses to allergens in the blood.

Leukemia and Lymphoma

1. The childhood leukemias include, in order of their rate of incidence, ALL, ANLL, and the very rare chronic granulocytic leukemia (CGL).
2. Although the cause of childhood leukemia is not known, it is probably the result of multiple interactions between hereditary or genetic predisposition and environmental influences.
3. Acute lymphoblastic leukemia is a potentially curable disease, with more than 75% to 80% of cases cured.
4. The lymphomas of childhood are non-Hodgkin lymphoma and Hodgkin lymphoma.
5. The origin of non-Hodgkin lymphoma is unknown. Factors that have been implicated include defective host immunity, a viral agent, chronic immunostimulation, and genetic predisposition.
6. Non-Hodgkin lymphoma has a favorable prognosis, with a 70% to 80% cure rate.
7. The risk of Hodgkin lymphoma is associated in part with infectious diseases, immune deficits, and genetic susceptibility.
8. Hodgkin lymphoma is a readily curable disease with long-term cure rates of 90% to 95%.