

Hemolytic disorders and congenital anomalies



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Lowdermilk: Maternity & Women's Health Care, 10th Edition Chapter 36:

Hemolytic Disorders and Congenital Anomalies Key Points - Print This section

discusses key points about hemolytic disorders. · Hyperbilirubinemia is a

condition in which the total serum bilirubin level is increased. It is

characterized by jaundice, a yellow discoloration of the skin, mucous

membranes, and sclerae. Jaundice primarily results from accumulation of

unconjugated bilirubin, which is a product of hemoglobin breakdown. ·

Although physiologic jaundice is common and usually benign, pathologic

jaundice is serious and can lead to acute bilirubin encephalopathy, which is

associated with acute and chronic neurologic damage. · Pathologic

hyperbilirubinemia can result from various causes. The most common causes

are hemolytic diseases of the newborn, which usually occur when the blood

groups of the mother and neonate are different. ABO and Rh factor

incompatibilities are the most likely hemolytic diseases. · Rh incompatibility

occurs when an Rh-negative mother has an Rh-positive fetus who inherits

the Rh-positive gene from the father. o Severe Rh incompatibility results in

marked fetal hemolytic anemia. The fetus compensates by producing

excessive immature erythrocytes, which causes erythroblastosis fetalis. o

Erythroblastosis fetalis leads to anemia, cardiac decompensation,

cardiomegaly, hepatosplenomegaly, and generalized edema. Fetal or

neonatal death can result. · ABO incompatibility is more common than Rh

incompatibility, but usually results in milder anemia. ABO incompatibility

occurs if the fetal blood type is A, B, or AB, and the maternal blood type is O.

· The nurse should determine women's blood type and Rh factor prenatally

and should take steps to prevent sensitization in Rh-negative women. · In Rh-

negative and Coombs' test—negative women, injection of Rho(D)

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immunoglobulin confers passive immunity and minimizes the risk of isoimmunization. · Neonatal exchange transfusion with type O, Rh-negative red blood cells serves to treat anemia and acidosis and to remove bilirubin, maternal antibodies, and fetal red blood cells that are beginning to hemolyze. This section presents critical points about congenital anomalies. · A congenital anomaly is a defect that is present at birth and can be caused by genetic or environmental factors or both. · Major congenital defects are the leading cause of death in term neonates. The most common major anomalies that cause serious neonatal problems are congenital heart disease, neural tube defects, cleft lip or palate, clubfoot, and developmental dysplasia of the hip. · Minor anomalies can be part of a characteristic pattern of malformations. That means they can point to the presence of a more serious anomaly and aid in its diagnosis. Common minor anomalies include a lack of the helical fold of the pinna, low-set ears, alterations in hair pattern or texture, absent philtrum, or a hairy patch or birthmark over the vertebral column. · Congenital anomalies include: o Congenital heart disease, such as ventricular septal defect and tetralogy of Fallot o Neural tube defects (such as anencephaly and spina bifida) and other central nervous system anomalies o Gastrointestinal defects, such as laryngeal web, congenital diaphragmatic hernia, cleft lip and palate, tracheoesophageal fistula, and imperforate anus o Musculoskeletal abnormalities, such as developmental dysplasia of the hip, clubfoot, and polydactyly o And genitourinary anomalies, such as hypospadias, epispadias, bladder exstrophy, and ambiguous genitalia. · Current technology permits prenatal diagnosis of many congenital anomalies and disorders. The nurse must be prepared to support the family's decision regarding these tests and their findings. · The most widespread use

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of postnatal testing for genetic disease is the routine screening of newborns for inborn errors of metabolism. · For an infant with a congenital disorder, the curative and rehabilitative problems are often complex and require multidisciplinary care. · Parents often need special instructions before they take home a high risk infant. For example, they may need to know how to meet their infant's nutritional requirements, provide cast care, or perform home phototherapy. · For parents of infants with an abnormal condition, supportive care begins at birth (or at the time of diagnosis) and continues for years.