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Hepatic Hemangioendothelioma

Hepatic hemangioendothelioma (HHE) is a rare, benign tumor that appears during the first six-months of life. Considered the most common vascular tumor of the liver in children is associated with a high mortality rate. HHE can be associated with congestive heart failure, anemia, thrombocytopenia (Kasabach-Merritt syndrome), hepatomegaly and cutaneous hemangiomas. Prenatal diagnosis has been associated with hydrops fetalis. Postnatal diagnosis is established with US, CT-Scan and MRI. Alpha-fetoprotein levels should be obtained to differentiate from hepatoblastoma. Mortality results from high-output cardiac failure secondary to arteriovenous shunting within the tumor (up to 50% of the cardiac output can be diverted), respiratory compromise, hepatic failure, intraperitoneal hemorrhage and consumptive coagulopathy. The younger the age at diagnosis, the more severe the cardiac symptoms. Natural history of asymptomatic HHE is spontaneous involution. Symptomatic lesions need aggressive management. Radiotherapy and chemotherapy have not shown consistently good results. Steroid and alpha-interferon are used as initial treatment to inhibit proliferation of endothelial and smooth muscle cells. Symptomatic solitary lesions can be managed with resection. Severe bilobar disease might need hepatic artery embolization or transplantation. Hepatic artery ligation or embolization should not be done in patients with shunting from the portal vein to the hepatic vein and minimal systemic arterial collateral circulation since it can result in hepatic necrosis.

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Candidemia

Candida species (*Albicans, Parapsilosis, Tropicalis and Krausei*) systemic infection has

steadily increased in the neonatal intensive care units during the past years. Associated factors for this type of infection are: prolonged use of broad-spectrum antibiotics, parenteral hyperalimentation, intravenous fat emulsions and placement of a central-venous catheters (CVC). Fungal infections are particularly common when TPN is administered through CVC. Candida can be cultured from the skin, urine, blood and mouth of affected patients. Fever, not-doing-well, and abdominal distention are the most common presentations. Infants who are found to have systemic candidiasis should be treated by removing all factors that predispose to systemic candidiasis (eg., indwelling catheters, broad-spectrum antibiotics) as persistent fungemia, morbidity and mortality are associated with attempts to maintain the CVC in the presence of Candidemia. Early initiation of systemic antifungal therapy (amphotericin, fluconazole) is imperative, along with searching for additional foci of disease. Endophthalmitis, venous thrombosis and endocarditis are complications of CVC associated Candidemia. Once the disease is recognized mortality rates are 20% in infants.

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Congenital Lobar Emphysema

Congenital lobar emphysema (CLE) is an unusual lung bud anomaly characterized by massive air trapping in the lung parenchyma that nearly always occurs in infancy and affects males more commonly (2:1). Lobar over distension causes compression of adjacent lung tissue, mediastinal shift and decrease in venous return. When this occurs persistent progressive respiratory distress (dyspnea, tachypnea, wheezing, cough and cyanosis) develops requiring lobectomy. Asymptomatic CLE exists, more commonly beyond infancy and associated with an acute viral respiratory infection. Lobar hyperinflation, flat diaphragms and retrosternal air, mediastinal shift in simple films suggests the diagnosis. CT scan depicts the abnormal anatomy (lung herniation) and the morphology of the remaining lung. V/Q scans confirm the non-functioning nature of the affected lobe. Upper and middle right lobes are more commonly affected. Etiology centers in a combination of bronchial (flap/valve) obstruction with congenital cartilage dysplasia. Most common associated defect is cardiovascular (VSD, PDA). Symptomatic patients nearly always

require lobectomy. Asymptomatic children do not benefit from surgical treatment but need close follow-up. Prenatally diagnosed cases need referral to surgery centers.

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