



Portal Vein Thrombosis

Extrahepatic non-cirrhotic portal vein thrombosis (PVT) is the second most frequent cause of portal hypertension in the world. In extrahepatic portal vein obstruction forward hepatopedal flow from the superior mesenteric vein and splenic vein is impeded due to obstruction of the portal vein which results in development of prehepatic portal hypertension and cavernomatous replacement of the portal vein. The causes of PVT can be split into four groups: direct injury to the portal vein and consequent thrombus formation as it occurs in omphalitis or umbilical catheterization; congenital malformation of the portal system associated with other cardiovascular disorders; indirect factors that predispose to thrombus formation in the portal system (sepsis, dehydration, hypercoagulability); and idiopathic. Children with PVT present with esophageal varices and bleeding, symptoms of secondary hypersplenism (splenomegaly) and growth retardation. Anomalies associated with PVT include heart and large vessel malformation and biliary (cholelithiasis). With time the child develops collateral circulation around the PVT along with coagulation disorders, ascites and biliary cirrhosis. Diagnosis of PVT is established with Doppler ultrasound and angio CT Scan. Variceal bleeding is managed with medication or endoscopic sclerotherapy. Persistent bleeding following endoscopic treatment, prominent splenomegaly with symptomatic hypersplenism, growth retardation, and symptomatic portal biliopathy may need surgical portosystemic shunt decompression the mesenteric-portal Rex shunt if the umbilical portion of the intrahepatic left PV and the superior mesenteric veins are patent. Otherwise, a splenorenal shunt is warranted.

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Congenital Hyperinsulinism

Congenital hyperinsulinism (CH) is the most frequent cause of severe, persistent hypoglycemia in neonates and young infants leading to seizures, developmental delay, and permanent brain damage. The inappropriate oversecretion of insulin is responsible for profound hypoglycemia which require aggressive treatment to prevent severe and irreversible brain damage. CH is a heterogeneous disorder with two histopathological lesions, diffuse and focal which are clinically indistinguishable. Focal CH is characterized by a sporadic somatic islet-cell hyperplasia. Diffuse CH corresponds to a functional abnormality of insulin secretion in the whole pancreas and involves several genes with different transmissions. Most cases are caused by mutations in genes coding for either of the two subunits of the beta-cell K(ATP) channel (ABCC8 and KCNJ11). In the diffuse form the hyperinsulinism is due to a recessive mutation of both alleles of these genes. To differentiate the focal from the diffuse form 18 F DOPA PET and CT scan is needed. Initial management of CH consist of diazoxide therapy. Focal lesions are effectively treated by limited pancreatic resection while diffuse lesions which are unresponsive to drug or dietary treatment require extensive laparoscopic or open pancreatectomy. Pancreatic beta-cell dysfunction persists following subtotal pancreatectomy of diffuse CH.

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Dermatopathic Lymphadenopathy

Adenopathies are a source of concern for both physicians and patients during the pediatric age. Diagnosis can be suggested by fine needle aspiration cytology, but other times complete lymph node removal is warranted. Most adenopathies in children are benign in nature. Dermatopathic lymphadenopathy (DPL) is characterized by non-neoplastic lymph node enlargement with reactive process and is generally caused by chronic inflammatory skin disease. DPL is usually caused by viral, bacterial, or other specific infections (tattoo). The morphologic features helpful in the diagnosis of DPL on fine needle aspiration cytology are melanin-laden macrophages with variable pigment; large, histiocytic clusters with blood vessels at the center; characteristic histiocytes, with elongated vesicular nuclei, nuclear grooves, crumpled and convoluted nuclei and pseudonucleoli; and absence of or very few tingible body macrophages. DPL is a harmless cause of enlarged lymph nodes rarely difficult to differentiate from mycosis fungoides or lymphoma.

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