



# **PEDIATRIC SURGERY UPDATE ©**

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### **Caroli's Disease**

Caroli's Disease (CD) first described in 1958 by Jacques Caroli as communicating cavernous ectasia of the biliary tree is an uncommon cause of chronic, often lifethreatening hepatobiliary disease. CD is a rare condition characterized by non-obstructive saccular or fusiform dilatation of the intrahepatic bile ducts usually manifested in childhood, thought to be congenital and presumably of autosomal recessive hereditary character. Caroli described two types: Type 1- the rare isolated variety characterized by recurring episodes of cholangitis. The more frequently occurring Type 2 is associated with congenital hepatic fibrosis, and consequently there are also symptoms of portal hypertension. Both types may make their first appearance at a very early age. Diagnosis can be done non-invasively with hepatobiliary scintigraphy, ultrasound and echo-doppler. The threats of this condition are: cholestasis, cholangitis, intrahepatic lithiasis, hepatic failure, and cholangiocarcinoma. Treatment of the localized form includes lobectomy. In diffuse disease, treatment may be medical with antibiotics and sometimes bile solvents. In case of failure, transplantation may be entertained. Therapy using Ursodeoxycholic acid (10 mg/kg/day) is indicated for intrahepatic stones in Caroli's syndrome. Patients must be followed closely for many years to ensure that the intrahepatic ducts do not remain dilated and that cholangitis do not recur.

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### **Splenosis**

Splenosis refers to autotransplantation of individual fragments of splenic tissue left behind after either operative or traumatic removal of the spleen. Although rarely symptomatic, splenosis may cause intestinal obstruction since the splenules link adjacent loops of bowels to each other, kinking and obstructing them. Tomographic selective splenic scintigraphy with sulphur colloid and heatdamaged red cells is the most sensitive method to detect splenosis. Intramural lesions may also be detected in barium studies of patients after prior trauma or splenectomy. In large inoculum the splenotic tissue has been found to have the capacity to remove intranuclear inclusion bodies from circulating red cells, phagocyte old erythrocytes and confer some immune protection. Patient who undergo emergency splenectomy for trauma are at a much higher risk of developing splenosis than those splenectomized due to hematologic conditions. Spilled splenic tissue seeds the peritoneum, takes root and grow into vascularized splenules. Experimental evidence

(Folkman) suggests that the presence of a large amount of vascularized spleen inhibits the growth of other splenic tissue in mice, theory why patients who undergo partial splenectomy, splenography or are merely observed after splenic rupture have almost no splenosis. Management of splenosis is expectant.

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## Pancreas Development

The pancreas develops from an anterior and posterior anlage of the foregut early during gestation (28 days). The anterior bud leads to the liver and body and tail of the pancreas. The posterior diverticulum develops into the head of the pancreas. This bud rotates anteriorly and later fuses to achieve the relationship to the rest of the pancreas. Development of the pancreas in embryonic life requires a trophic stimulus from the associated mesenchyme. Under the influence of this mesenchyme the mature organ develops, being mainly composed of ductal, exocrine and endocrine cells. Exocrine and ductal pancreas are derived from the endoderm of the foregut. Recent evidence suggests that the endocrine cells derive also from the endoderm of the foregut as evidenced by the expression of the genes responsible for hormonal production. This challenges the theory that endocrine cells may originate from the neural crest cells (neuroectodermal) of the embryo reinforced by the enunciation of the amine precursor uptake decarboxylase (APUD) theory.

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