



# **PEDIATRIC SURGERY UPDATE ©**

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### **Bile-Plug Syndrome**

Define as a partial or complete intraluminal obstruction of the extrahepatic bile ducts caused by an abnormal, viscous, inspissated bile. Clinical presentation is that persistent (conjugated) jaundice. This condition is associated to: massive hemolysis from Rh or ABO incompatibility, cystic fibrosis, TPN, and ileal resections. Ultrasound findings are: dilated gallbladder with sludge or stone formation, dilated common bile duct with sludge (echogenic appearance), and no acoustic shadowing. DISIDA shows no intestinal excretion of the radioisotope. Most cases subside spontaneously. Those associated to worsening of direct hyperbilirubinemia and liver function tests should be managed surgically. Irrigation of bile ducts with saline or N-acetylcysteine through the gallbladder is curative.

### **Gallstones**

With the increase use of sonography in the work-up of abdominal pain, cholelithiasis is diagnosed more frequently in children. Gallstones occur as consequence of loss of solubility of bile constituents. Two types are recognize: cholesterol and bilirubin. Those of cholesterol are caused by supersaturation of bile (lithogenic) by cholesterol overproduction or bile salt deficiency. Bilirubin stones occur due to hemolysis (Sickle Cell, thalassemia) or bacterial infection of bile. Other etiologies include: *Ascaris Lumbricoides* infestation, drug-induced (ceftriaxone), ileal resection, TPN. etc. Gallbladder sludge is a clinical entity that when it persists can be a predisposing factor for cholelithiasis and cholecystitis. Management of symptomatic gallstones consists of laparoscopic cholecystectomy which has found to: reduce hospital stay, increase child return to normal activities, is well tolerated, low complication rate, but increases cost of procedure.

### **On HD Etiology**

Hirschsprung's disease (HD) is characterized by lack of enteric ganglion cells, hyperplasia of abnormal nerve fibers and a non-propulsive, non-relaxing segment of bowel. Classically the etiology is attributed to a failure of cranio-caudal migration of parasympathetic neural crest cells to the distal

bowel. A plausible explanation for the failure of relaxation of the bowel involved is a deficiency of enteric inhibitory nerves that mediates the relaxation phase of peristalsis. These nerves are intrinsic to the gut and are classified as non-adrenergic and non-cholinergic. Nitric oxide (NO) has recently been implicated as the neurotransmitter which mediates the relaxation of smooth muscle of the GI tract in HD. Its absence in aganglionic bowel might account for the failure of relaxation during peristalsis. Besides adhesion molecules (absent in aganglionic bowel) during early embryogenesis might restrict the neuro-ectodermal origin involved in the initial contact between nerves and muscle cell (synaptogenesis) suggesting that developmental anomaly of innervated muscle and absent NO causes the spasticity characteristic of HD.

### **CF Prenatal Diagnosis**

Cystic Fibrosis is an autosomal recessive lethal disease affecting 1 in 2000 live newborns. It is caused by different mutations in the cystic fibrosis transmembrane conductance (CFTC) regulator gene. Prenatal diagnosis is possible to couples of carriers with a one-fourth risk of having an affected newborn. Reliability of the test is 98%. The sample is taken as chorionic villus (10-14 week gestation) or amniocentesis. A DNA polymerase chain reaction analysis has shown a major mutation in the delta F508 position (is a three base pair deletion in exon 10 which results in a protein without the amino acid phenylalanine at position 508) of the long arm of chromosome 7.

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