**Extra Hepatic Portal Vein Obstruction**

**Definition**: Portal hypertension caused by EHPVO occurs when the site of block is in the portal vein with or without involvement of extra hepatic portal vein or splenic or superior mesenteric vein (SMV). Isolated occlusion or splenic vein / SMV does not constitute EHPVO. Portal vein obstruction associated with CLD or neoplasia does not constitute EHPVO.

**Etiology**: The etiology of EHPVO in children has not been well documented but is thought to result from infection (umbilical sepsis / umbilical vein cannulation, abdominal surgery, trauma etc) congenital anomaly, prothrombotic states (deficiency of protein C, protein S, antithrombin 3 deficiency etc.) or idiopathic (in >50% of patients)

**Clinical Presentation**: EHPVO is childhood is most often chronic and presents with features of variceal bleeding, feeling of lump in abdomen (spleenomegaly). It is usually associated with growth retardation in ~50% of children. Ascites develops in a small proportion of patients.

**Natural History**: The bleeding risk factor (no. of bleeds/ year) is about 1.3. The bleeding risk remains unchanged after puberty. Incidence of rebleeds from varices after obliteration is low in long term follow up studies. Repeated transfusion increase risk of acquired hepatitis B and hepatitis C infection. EHPVO in children has a relatively benign course.

**Diagnosis**: Imaging is mainstay for diagnosis. Ultrasound is a reliable noninvasive technique with a high degree of accuracy for the detection of portal cavernoma and is the investigation of choice. CT and MRI also have a high degree of sensitivity and specificity.

**Management**

1. **Endoscopic therapies**: Endoscopic sclerotherapy and band ligation have been found to be effective in the control of acute variceal bleeding in several studies. Band ligation has been found to be superior to sclerotherapy. For management of gastric variceal bleeding injection of glue is effective.

2. **Shunt surgery**: Both total and selective shunts such as lineorenal shunt have been used is reserved in patients who fail endoscopic therapy, have significant growth retardation, symptomatic portal biliopathy and symptomatic hypersplenism. It can also be offered to patients who demand a ‘one time’ treatment.