

Poster #: 10

Title of Congenital absence of portal vein.

Abstract:

Institution: UCLA

Authors: Shahnaz Ghahremani, MD

Modality: MR

Organ System: GI

Intro: N/A

Purpose: N/A

Methods Used: N/A

Results of N/A

Abstract:

Discussion: N/A

Scientific and/or Clinical Significance? Congenital absence of portal vein with a complete loss of liver portal perfusion leads to developmental and functional alterations that predispose liver to focal or diffuse hyperplastic or dysplastic changes, also there is increased incidence of hepatic tumors or tumor like conditions in these patients. This poster presents 2 cases of congenital absence of portal vein in very young ages and with some degrees of liver failure. Both patients are in liver transplant list . Due to complex vascular anatomy they need to have a whole graft.

Relationship to existing work Young age of the patients at the time of presentation.

This poster presents 2 cases of congenital absence of portal vein: Case # 1 is a 4 year old boy with history of regenerative nodular hyperplasia of the liver, and persistent low grade cholestasis. MRV of the abdomen showed absence of portal vein and drainage of superior mesenteric vein to the IVC, just inferior to the IVC/right atrium junction. case # 2 is a 2 year old girl with pulmonary hypertension, bilateral pulmonary arteriovenous malformation. She presented with feeding intolerance and emesis. MRV of the abdomen showed absence of portal vein and direct drainage of mesenteric veins to the left sided IVC with Azygus vein continuation. Congenital absence of portal vein was considered a rare event in the past. However with advances in imaging techniques the number of detected cases are increasing. This condition usually presents in older patients, our patients are presenting in very young ages.