Abstract:
Background Congenital anomalies of the inferior vena cava are extremely rare with agenesis of retrohepatic segment of the inferior vena cava (RHIVC) with azygos continuation being a relatively well-known congenital anomaly. We present the rare case of absence of the RHIVC, with venous return entirely through the liver parenchyma by way of a cavo-portal shunt. This to our knowledge is only the second such case reported in English medical literature. Case report A eleven year old girl with normal milestones, presented with abdominal pain of four days duration. She gave no previous history of medical problems. She was evaluated and her abdominal examination and blood investigations were unremarkable. An Ultrasonogram revealed a cavo-portal shunt, with absent retrohepatic segment of IVC. CT angiogram was obtained which confirmed the findings. As she was asymptomatic, she was managed conservatively and continues to be asymptomatic on routine followup.

Conclusion We highlight an extremely rare case of an absent segment of IVC with congenital cavo-portal shunt, wherein the patient was well compensated and was managed by masterly inactivity.

Keyword: Cavo-portal shunt, Agenesis of retrohepatic IVC

Figure 1
Introduction:
Congenital anomalies of the inferior vena cava (IVC) occur in 0.3% to 0.5% of healthy individuals and 0.6% to 2% of those with other cardiovascular defects.\(^1\) Inferior vena cava agenesis is one of the rarest congenital anomalies, with an estimated prevalence ranging from 0.0005 to 1%.\(^1,2\) This phenomenon has been labeled in a variety of ways such as absence, agenesis, anomalous, and interruption of a particular segment (infra-hepatic, prerenal, renal, or infrarenal) of the IVC.\(^1,2,3\) Its etiology is controversial, but embryological dysgenesis is the most commonly proposed origin.\(^3\) The absence of the hepatic segment of the inferior vena cava with azygos continuation is a well-known congenital anomaly. An extremely variant is a missing retrohepatic segment of inferior vena cava (RHIVC) with venous return entirely through the liver parenchyma by way of a cavo-portal shunt. This to our knowledge is the only the second such case reported in English medical literature.\(^4\)

Case Report:
An otherwise healthy 11-year-old girl with normal developmental milestones presented with a history of upper abdominal pain of four days duration. She had no other abdominal symptoms, and denied having any symptoms suggestive of cardiovascular pathology. The patient had no history of previous hospitalization. On examination, her general and abdominal examinations were unremarkable. Her lower limbs were normal and she had no signs suggestive of IVC obstruction. All her blood investigations were unremarkable. An Ultrasound of the abdomen revealed a cavo-portal shunt, and an absent retrohepatic segment of IVC. The main portal vein was dilated at 9 mm with hepato-petal flow. Hepatic veins were normal and drained into a normal suprahepatic IVC. The infrahepatic segment of IVC was also normal. Liver, gallbladder and both kidneys were normal. There was no splenomegaly or free fluid in the abdomen. An Upper GI endoscopy was done which was also normal. A CT angiogram was done which confirmed the USG findings. (Figure 1, Figure 2, Figure 3) As the patient was well compensated, with no evidence of liver pathology, portal hypertension, cardiac decompensation or IVC obstruction; following a
Multidisciplinary team meeting it was decided to manage her conservatively with regular follow-up. Her abdominal pain settled with proton pump inhibitors and she remains asymptomatic on routine follow-up.

**Discussion:**
Embryologic abnormalities of the IVC and its tributaries are rare. The most common IVC malformations are hypoplasia of the pre-renal and renal segments, followed by hypoplasia of the postrenal segment and IVC duplicity. These anatomical variations occur between the sixth and eighth weeks of embryonic development. The embryogenesis of the IVC is a complex event that involves formation, regression and fusion of three pairs of embryonic veins. The infrahepatic segment of the IVC originates in three parts: suprarenal, renal and infrarenal. The right subcardinal vein becomes the suprarenal segment, while the supracardinal anastomoses with the subcardinal vein to give rise to the renal segment, and the infrarenal segment emerges from the right supracardinal vein. The proximal part of the inferior vena cava is formed by a union between the right subcardinal vein (venous return from the mesonephros) and the right hepatocardiac channel (paired vitelline veins entering into the primitive sinus venosus). The failure of union between these two vessels results in the so-called absence of the hepatic segment of the inferior vena cava. Connections between the subcardinal venous system, the vitelline venous system, and the foregut venous plexus are known to occur during an early stage of embryonic development. Development of hepatic vascularization and of the hepatic segment of the inferior vena cava are closely linked during this stage. The right hepatocardiac channel is destined to enlarge and form the hepatic segment of the inferior vena cava. The portal vein is formed by persisting parts of the right and left vitelline veins and anastomoses between these two systems. Hence a portocaval shunt develops from persisting embryonic vascular remnants of anastomoses between the different venous plexuses involved in the formation of both the hepatic segment of the inferior vena cava and the portal vein. Development of a cavoportal shunt may be explained by the fact that the portal system and the inferior vena cava have common precursors. In the absence of the inferior vena cava, the right subcardinal vein fails to make its connection within the liver and shunts its blood directly into the right supracardinal vein. Therefore, blood from the lower extremities and abdomen is forced to reach the right heart via the azygos system and superior vena cava. During early embryonic life, the blood flow follows the shortest way, involving the least resistance, to reach the right atrium. Because of this, and because of the lack of dilatation of the azygos system in this stage, the cavoportal shunt in our patient is likely to have formed before the hepatic segment of the inferior vena cava developed. The etiology of the IVC agenesis is controversial in literature. Theories for the anomalies include congenital dysgenesis, perinatal thrombosis of the IVC. IVC agenesis or hypoplasia may be accompanied by other congenital abnormalities such as splenic anomalies, disorders of intestinal rotation, pulmonary dysgenesis, renal agenesis, dextrocardia and other congenital heart diseases. These anomalies are identified in over 1% of the patients. Extrahepatic connections between the portal and caval systems have also been observed in neonates and in adult patients having no liver disease. The association of a congenital mesocaval shunt, azygos continuation
of the inferior vena cava, polysplenia, and dextrocardia, known as the Abernethy mal-
ofcation is an extreme variant of a porto-
caval shunt. With marked strides being made in the field of imaging coupled with their ubiquitous usage, there is an increased detection of asymptomatic IVC anomalies. The best imaging methods for the diagnosis of IVC anomalies are CT angiography and MRI. These anomalies may be associated with vague nonspecific symp-
toms and in most cases may even completely asymptomatic, as was noted in our patient. She had no evidence of portosystemicencephalopathy, Budd-Chiari syndrome or portal hypertension (varices, splenomegaly, ascites) as maybe observed in cases of portocaval anastomosis or membranous obstruction of inferior ve-
nacava. This patient was asymptomatic and the discovery of this condition was incident-
al, hence a conservative management without disruption of the existing congenital shunt and the status of homeostasis achieved by it, appears to be the best course of action. In our patient, the blood from the inferior vena cava is not only shunted into the portal vein but also reaches the heart through the liver. To our knowledge, such an anastomosis associated with absence of the inferior vena cava has only been reported once before in humans.

Conclusion:
We report only the second ever reported case of a congenital cavo-portal shunt associated with agenesis of the retrohepatic IVC. We also aim at highlighting the conservative line of management of this patient, which follows the basic tenets of medicine which is primum non nocere.

References
7 Konopka CL, Salame M, Padulla GA, Muradás RR, Batistella JC. Agenesis of inferior vena cava associated with deep venous thrombo-


