

Etiopathogenesis of Portal Hypertension in Children: A Review Article

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Abstract

Portal hypertension (PH) is usually encountered as a complication arising from chronic liver disease and cirrhosis. It has been reported that PH is the 12th leading cause of death in the United States and a total of 40545 deaths were observed in 2016 [1]. Common presentation of PH in children include catastrophic variceal hemorrhage usually from esophagus. Other common clinical features of PH include splenomegaly, hypersplenism and ascites. Less commonly encountered complications of PH in children include encephalopathy, hepatopulmonary syndrome and portopulmonary hypertension. In this review article it is aimed to review the clinical and radiographic features of PH in children under the light of relevant literature.

Keywords: Portal Hypertension (PH); Children; Encephalopathy; Hepatopulmonary Syndrome

Etiology and pathogenesis

PH is a multiorgan disease with significant neurohormonal, cardiovascular, pulmonary, renal, immune, coagulation and metabolic consequences for children [2]. It occurs as a result of increased vascular resistance and blood volume through the portal venous system. Normal portal venous pressure is 7 - 10 mmHg and PH is defined as portal pressure greater than 10 mmHg or hepatic venous pressure gradient greater than 4 mmHg [3].

The causes of PH can be categorized into 3 categories with regard to the anatomical level of vascular resistance and histology of liver parenchyma: prehepatic, hepatic and posthepatic (Table 1). Arteriportal hypertension has a different mechanism and is usually regarded as a separate entity.

Prehepatic causes
Portal vein thrombosis
Congenital or acquired stenosis of portal vein
Splenic vein thrombosis
Hepatic causes
Presinusoidal

Congenital hepatic fibrosis
Polycystic liver disease
Nodular regenerative hyperplasia
Myeloproliferative disease (lymphoma, leukaemia)
Granulomatous disease (schistosomiasis, sarcoidosis, tuberculosis)
Non-cirrhotic portal fibrosis
Idiopathic portal hypertension
Sinusoidal
Liver cirrhosis (independent of cause)
Post sinusoidal
Veno-occlusive disease
Posthepatic causes
Budd-Chiari syndrome
Inferior vena cava obstruction
Constrictive pericarditis
Right heart failure

Table 1: Classification and causes of PH in children.

Arterioportal hypertension is caused by excessive arterial inflow into the portal venous system [4,5]. Derived from a vascular malformation in the hepatic parenchyme, there is an arterioportal fistula leading to hyperdynamic state in arterioportal hypertension. Initially the liver is usually normal and as the effect of barotrauma increases capillaries in the liver parenchyme become fibrotic with thickening of the intima. At that point PH is regarded as mixed with a combination of both increased flow and resistance. Management of these patients can be challenging and these patients may ultimately need liver transplantation.

Prehepatic causes

Portal vein trombosis (PVT) is the most common cause of extrahepatic portal vein obstruction (EHPVO) in children. Common contributing events leading to PVT in children include neonatal events such as umbilical vein catheterization, omphalitis and sepsis. There are various disorders in children that lead to PVT and include prothrombotic disorders such as protein C, protein S and antithrombin III deficiencies and factor V Leiden mutations which account for up to 35% of children with PVT while the cause of PVT remains unidentified in about 50% of cases [3].

Intrahepatic causes

There are various intrahepatic causes that give rise to increased portal bed resistance within the liver and these can be presinusoidal, sinusoidal and postsinusoidal. Congenital hepatic fibrosis and nodular regenerative hyperplasia are commonly seen as presinusoidal causes and often do not result in impairment of liver function. Whatever the underlying primary liver disease, liver cirrhosis ends up with sinusoidal obstruction. Dual effects are mechanical and dynamic factor that lead to sinusoidal obstruction syndrome. Architectural

derangement caused by fibrosis and nodule formation is the mechanical cause of increased intrahepatic vascular resistance in cirrhosis. Dynamic factor is related to vasoactive effect of endogenous factors leading to vasoconstriction. Veno-occlusive disease (VOD) is the best example of postsinusoidal obstruction. The characteristic features of VOD are microthrombosis and sclerosis of hepatic venules. Typical presentation of this entity includes hyperbilirubinemia, hepatomegaly and ascites developing within 3 weeks from haematopoietic stem cell transplantation.

Posthepatic causes

Budd Chiari syndrome which is uncommon in children can cause posthepatic portal hypertension. Increased right atrial pressure in certain cardiac diseases is another cause of posthepatic PH. In this type of PH chronic venous congestion results in hepatomegaly and the end result is liver dysfunction and cirrhosis.

Conclusion

In conclusion, the spectrum of causes leading to PH in children has a wide range. The management of these children focuses on the prevention and treatment of catastrophic variceal hemorrhage usually from esophagus. As the management strategies in these cases have improved during the past decade, provided that care is given by experienced clinicians and institutions, the long-term outcome of children with PH will be better than those previous cases.

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