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Mini Review

Complications of Portal Hypertension in Chilhood: A Review Article

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Abstract

Portal hypertension (PH) is a hazardous entity which is usually encountered as a multiorgan disease. Clinical presentations of PH in children are variable including gastrointestinal haemorrhage, splenomegaly and hypersplenism, ascites, encephalopathy, hepatopulmonary and/or portopulmonary hypertension. If untreated, catastrophic end results and even death may occur. In this review article it is aimed to review the complications of PH in children under the light of relevant literature.

Keywords: *Portal Hypertension; Chidren; Complications*

Introduction

Portal hypertension (PH) is usually defined as either a hepatice venous pressure gradient greater than 5 mm Hg, or hepatic venous wedge pressure greater than 10 mm Hg [1]. It is usually encountered as a multiorgan disease predisposing these children to significant neurohormonal, cardiovascuar, pulmonary, renal, immune, coagulation, and metabolic consequences [2].

Clinical presentations of PH in children are related to the complications of this clinical entity and include gastrointestinal haemorrhage in the form of catastrophic variceal haemorrhage usually from esophagus, splenomegaly and hypersplenism, ascites, encephalopathy, hepatopulmonary and/or portopulmonary hypertension. In this review article it is aimed to review the complications of PH in children under the light of relevant literature.

Gastrointestinal bleeding

Variceal bleeing is the most common clinical presentation of extrahepatic portal vein obstruction (EHPVO) in noncirrhotic pediatric patients [3-7]. These varices are usually seen in the esophagus but may also be observed in the stomach as gastropathy or antral vascular ectasia, and duodenum, peri-stomal sites and rectum [8]. It has been reported that more than half of children with EHPVO develop variceal bleeding by 16 - 18 years of age while 17%-29% of children with biliary atresia present with gastrointestinal bleeding [8]. There are various reasons to contribute for variceal rupture and these include increased abdominal pressure, increased cardiac output which is commonly seen in these patients and use of nonsteroidal anti-inflammatory medication.

Splenomegaly

Splenomegaly is a common clinical finding in these children. The haematological end result of hypersplenism includes pancytopenia including thrombocytopenia, leucopenia and anemia. If the disease is not managed appropriately, increasing platelet sequestration in the spleen may aggravates the thrombocytopenia. After appropriate management of PH as in the form of liver transplantiation or shunt surgery it has been suggested that splenomegaly and hypersplenism are expected to improve over time.

Ascites

Ascites is usually seen in children with PH due to cirrhosis. Common postulated mechanism of ascites is the increased splanchnic inflow producing high filtration pressure and increased lymph formation which overcomes the drainage capacity of the lymphatic system. The consequence of these pathophysiological mechanisms is the accumulation of abdominal ascites which also decreases the effective circulating volume. The usual management of ascites includes salt and fluid restriction together with diuretic therapy. Spironolactone and furosemide are commonly used in these cases with albumin infusions. In patients with refractory ascites, paracentesis may be reserved to avoid respiratory compromise or for diagnostic evaluation.

Pulmonary complications

Hepatopulmonary syndrome and rarer portopulmonary syndrome are rarely seen in children with PH. Although the exact pathogenesis of these entites remain unclear, the hyperdynamic circulatory changes predispose these patients to develop pulmonary edema with increased pulmonary vascular resistance and impaired gas exchange. Clinical manifestations of hepatopulmonary syndrome include dyspnoea, cyanosis and digital clubbing. On the other hand portopulmonary syndrome may be seen in these cases in the clinical forms of exertional dyspnea, fatigue, palpitations, syncope and chest pain. It has ben reported that of the patients with PH, hepatopulmonary syndrome occurs in 20% of cases with cirrhosis and portopulmonary syndrome is seen in 5% of these patients [9].

Abnormal venous patterning

This finding also called as caput medusa may also provide an important clue to underlying PH. Abnormal venous patterning develops due to spontaneous porto-collateral shunting usually due to anastomoses between the paraumbilical veins and small epigatric veins of the anterior abdominal wall producing spiderlike venous images. It has been reported that in children with short bowel syndrome and intestinal failure-associated liver disease peristomal variceal bleeding often present and peristomal regions are common sites for bleeding in these children [8].

Other complications

Hepatic encephalopathy may be seen in these children due to decompansated liver disease and PH together with increased porto-systemic shunting. In young children manifestations may be quite subtle and a high index of suspicion is recommended. Renal dysfunction is also a frequent complication of chronic liver disease and these children usually come up with increased serum creatinine levels. Growth retardation is also a recognised complication in children with PH which may be due to hypertensive enteropathy, underlying liver dysfunction and growth hormone resistance [8]. Nutritional supplementation may improve outcomes in these patients.

Conclusion

In conclusion, the spectrum of complications in children with PH has a wide range. Variceal bleeding is the most serious complication which may produce hazardous consequences for these cases. All of the complications in childhood PH should be managed accordingly. Shunt surgery including liver transplantation prevents gastrointestinal bleeding, corrects hypersplenism, ascites and can alleviate growth retardation.

Conflicts of Interest

None.

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Author Contribution to the Manuscript

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