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Review Article

Particularities of Portal Hypertension in Pediatric Patients

Oana Belei*

First Pediatric Clinic, University of Medicine and Pharmacy Victor Babes Timisoara, Romania

***Corresponding Author**: Oana Belei, First Pediatric Clinic, University of Medicine and Pharmacy Victor Babes Timisoara, Romania.

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Abstract

Portal hypertension is well described in a wide range of pediatric disorders, many of which are fundamentally distinct from the diseases that affect adults. Those differences have profound implications for diagnosis and management. The leader causes of portal hypertension in children are biliary atresia, cystic fibrosis and extrahepatic portal vein obstruction, also known as portal vein thrombosis. In the latest years, there was an increased number of young children unaware of any conditions, admitted with bleeding from esophageal varices as the initial presenting problem. Most of them were under-diagnosed children with portal hypertension due to portal cavernoma, former preterm babies with very low birth weight that survived due to advance medical services and high performance support treatment provided in maternity. Approaches to the management of portal hypertension and variceal hemorrhage in children remain controversial, due to lack of rigorous pediatric clinical studies. In this short review, the authors will display the particularities of portal hypertension in children.

Keywords: *Portal Hypertension; Esophageal Varices; Children*

Introduction

Portal hypertension is well described in a wide range of pediatric disorders, many of which are fundamentally distinct from the diseases that affect adults. Those differences have profound implications for diagnosis and management. The leader causes of portal hypertension in children are biliary atresia, cystic fibrosis and extrahepatic portal vein obstruction (EHPVO), also known as portal vein thrombosis. These conditions present critical differences from the common hepato-cellular based disorders that lead to portal hypertension in adult patients. Most important is the fact that portal hypertension in children is an early manifestation at a time when hepatic function is relatively intact. Approaches to the management of complications of portal hypertension in children are frequently driven by expert opinion and not fully evidence based [1].

Discussion

EHPVO is defined by the obstruction of the extrahepatic portal vein, with or without involvement of the intrahepatic portal veins or other segments of the splanchnic venous axis. Chronic EHPVO is characterized by features of thrombosis with portal cavernoma as a sequel of portal vein obstruction. Diagnosis is made by Doppler ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI) angiography, which demonstrate portal vein obstruction, presence of solid intraluminal material or portal vein cavernoma. Doppler ultrasound should be considered first line investigation and CT- or MRI-angiography should be performed subsequently for assessment of thrombosis extension [1]. EHPVO represents one of the commonest cause of portal hypertension and variceal bleeding in children. It is estimated that approximately 50% of pediatric patients with chronic liver disease and 90% of those with EHPVO will experience gastrointestinal bleeding [2]. Also, morbidity due to splenomegaly with hypersplenism, growth failure and portal biliopathy is significant.

The spectrum of complicated diseases that lead to portal hypertension in children continues to expand. The most frequent causes of intrahepatic portal hypertension in children are biliary malformation and metabolic conditions such as biliary atresia, Alagille syndrome,

cystic fibrosis, glycogen storage disease or Wilson disease. A variety of other advanced liver disease in children may be accompanied by portal hypertension. Chronic viral B, C or autoimmune hepatitis reach the stage of cirrhosis in childhood less frequent compared to adult population. Apart from primarily hepatocellular or sinusoidal conditions in children, there are certain extrahepatic disorders that can lead to portal hypertension such as Budd-Chiari syndrome or untreated choledochal cyst. Interesting recent case series of Fontan-related liver disease and obliterative portal venopathy have been published recently [1].

Approaches to the management of portal hypertension and variceal hemorrhage in children remain controversial, due to lack of rigorous pediatric clinical studies. Underpinning the practice of endoscopic screening, variceal ligation/banding and surgical intervention is an accurate understanding of the natural history of portal hypertension in children, which is distinct from adults. Therefore, caution should be exercised in the direct extrapolation of the screening and treatment practices for cirrhosis in adults to children. Primary prophylaxis of variceal hemorrhage in adults with cirrhosis is widely accepted, it is not clear if this should be extended to children. Available data in pediatrics is quite limited and mostly retrospective [3].

In EHPVO, portal hypertension develops early in the clinical course of disease leading to variceal hemorrhage in children who have well compensated disease and less comorbidity, who therefore can tolerate the bleeding episodes better than an adult with decompensated cirrhosis [4].

There is a remarkable paucity of high-quality reported literature on the event of acute variceal hemorrhage in children. Endoscopic information has been presented, but details of clinical course and related morbidity are almost nonexistent in the pediatric literature. Mortality after variceal hemorrhage can be extracted from a number of published experiences, although strict application of Baveno definitions related to timing is not generally employed. This information is absolutely critical for informed decision-making related to primary prophylaxis [1]. Despite significant progress since Baveno V, clinicians caring for children with portal hypertension face difficult clinical decision-making. Strict evidence-based decisions are difficult to derive. Numerous summaries have been written and concerted efforts to provide expert pediatric-oriented opinion on Baveno IV and V have been published [4-6].

The use of nonselective ß-blockers in the management of portal hypertension in children remains quite controversial and poorly informed by solid evidence of optimal approaches and efficacy [1]. Propranolol is the most widely used agent in pediatrics. Variable basal heart rate during normal development and difficulties in accurate measurement of heart rate in younger children have hampered the use of a standard reduction in heart rate as a guide to pediatric nonselective ß-blockers dosing. Hepatic venous pressure gradient has been measured in a limited number of children with some technical issue [1]. When bleeding is the initial presenting problem, endoscopic therapy should be used as secondary prophylaxis. Endoscopic variceal ligation should be the first approach in cases with variceal bleeding. The major limitation of this technique is the lack of pediatric specific equipment and hence it is not always possible to pass a loaded bander through the pharynx in small children. Thus, unfortunately size limitations may require injection sclerotherapy in children who are less than 10 or 15 kg.

A variety of very important clinical sequelae arise from the consequences of portal hypertension: variceal hemorrhage, hepato-pulmonary syndrome, porto-pulmonary hypertension, ascites, hypersplenism etc. Recent reports highlight the prevalence of some of these issues in children [7]. Variceal hemorrhage is clearly an issue for children and can be a dramatic mode of presentation.

The abundance of underlying liver disease etiologies along with extra-hepatic causes of portal hypertension in children have raised interest in developing rigorous models for risk stratification. Risk stratification is defined as a statistical process to determine detectable characteristics associated with an increased chance of experiencing unwanted clinical outcomes. By identifying factors before the occurrence of an event, it may be possible to develop targeted interventions to mitigate their impact. In children with portal hypertension, it would be of great interest to stratify the risk of variceal development and to predict the onset of haemorrhage by using reproducible, non-invasive methods such as liver and spleen stiffness measurements.

Conclusion

Quality of life in children with portal hypertension is reduced and related to the degree of hypersplenism and failure to thrive according to the underlying condition. Different components of quality of life are affected, including physical, emotional, social and school function. Variceal eradication and porto-systemic shunts does not always resolve these quality of life issues.

Conflict of Interest

I have no conflict of interest.

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