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Portal Hypertension in Children: Clinical Characteristics, Types of Complications, and Long-Term Treatment Results

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Abstract: Portal hypertension in children is one of the most complex and pressing problems in modern hepatology, pediatric surgery, and gastroenterology, characterized by severe hemodynamic disorders, a high risk of life-threatening complications, and a significant decrease in patients' quality of life. The problem acquires particular relevance due to the early manifestation of the disease, rapid progression of portal block, the development of bleeding from varicose veins of the esophagus and stomach, as well as high mortality during inadequate treatment. According to WHO and international registries, portal hypertension is detected in 2-4% of the child population in developed countries and up to 10-15% in helminthic endemic regions.

Keywords: Portal Hypertension, Children, Modern Hepatology

Introduction

Modern achievements in non-invasive imaging, molecular diagnostics, and intervening radiology open up new opportunities for early diagnosis and effective treatment of portal hypertension in children [1,2,3]. The introduction of high-tech diagnostic methods, such as shift wave elastography, transient elastography, multi-detector computed tomography with portal blood flow assessment, and magnetic resonance imaging with contrast, allows not only to identify the presence and degree of portal block, but also to assess the risk of varicose vein bleeding at the preclinical stage. The introduction of endoscopic treatment methods (varicose vein sclerotherapy, ligation of esophageal varices), surgical interventions (distal splenorenal bypass surgery, mesoportal bypass surgery), and intervention procedures (transjugular intrahepatic portosystemic bypass surgery - TIPS) has significantly improved outcomes and long-term survival rates for children with portal hypertension [4,5].

At the same time, despite significant successes in understanding the pathogenesis of portal hypertension, many issues of diagnosis, prognosis of the course, and optimization of treatment tactics remain insufficiently studied. There is no unified algorithm for the comprehensive examination of children suspected of portal hypertension, risk criteria for the development of life-threatening hemorrhages have not been sufficiently studied, and personalized approaches to choosing optimal treatment tactics have not been developed, taking into account the etiology of the disease, the severity of the portal block, and the individual characteristics of the patient [6,7]. In the Republic of Uzbekistan, the prevalence of portal hypertension in children associated with congenital anomalies of the portal system, liver cirrhosis, and extrahepatic portal block varies from 2.5 to 4.8 per 100,000 child population (National Clinical Protocol of the Ministry of Health of the Republic of Uzbekistan, 2024), and mortality from the first bleeding from varicose veins reaches 10-20% [8].

Factors determining the prognosis for portal hypertension include the etiology of the disease (intrahepatic vs extrahepatic block), localization and length of the portal block, preservation of liver function, presence and severity of varicose veins, and the development of portal encephalopathy. Despite the introduction of new diagnostic and treatment methods, the long-term outcomes of portal hypertension in children in the domestic population have not been sufficiently studied. Recent studies emphasize the importance of early diagnosis, stratification of complication risk, and the development of individualized treatment strategies. This has determined the relevance of this study.

Portal hypertension in children represents one of the most serious problems in modern pediatric hepatosurgery and gastroenterology, requiring a multidisciplinary approach and an integrated system of medical care. According to international registries, portal hypertension is detected in 2-15% of cases among the child population depending on the region, with a significant portion of patients developing life-threatening complications[10]. In the Republic of Uzbekistan, the prevalence of portal hypertension in children is 2.5–4.8 per 100,000 children (National Clinical Protocol of the Ministry of Health of the Republic of Uzbekistan, 2024), while bleeding from esophageal varices remains the leading cause of death in this group of patients. Despite the successes achieved in diagnosis and treatment, many issues remain unresolved: selecting optimal treatment tactics depending on the etiology and localization of the

portal block, long-term prognosis criteria, and principles of rehabilitation and monitoring after surgical intervention[9,10,11].

The aim of the study is to study the clinical and diagnostic features of portal hypertension in children of various etiologies, determine risk factors for the development of life-threatening complications, and evaluate the long-term results of surgical and pharmacological treatment.

Materials and Methods

The study was conducted at the Yu.Yu. Jabbarov Republican Children's Medical Center, the Department of Pediatric Surgery of the Andijan Pediatric Medical Institute, and the Department of Hepatological Surgery of the Republican Scientific Center of Surgery between 2020 and 2026. The research type is prospective controlled with elements of retrospective analysis of long-term results.

The study included 240 children aged 1 to 18 years with a verified diagnosis of portal hypertension, divided into two groups. The main group consisted of 130 patients receiving comprehensive treatment according to the developed algorithm (including endoscopic therapy, drug treatment, and surgical interventions if indicated); the control group consisted of 110 children receiving conservative treatment according to standard protocols.

Inclusion criteria: age from 1 to 18 years; verified diagnosis of portal hypertension (based on ultrasound, computed tomography, magnetic resonance imaging, and/or hepatic vein catheterization); presence of parental or guardian consent to participate in the study.

Exclusion criteria: acute or exacerbation of chronic pancreatitis at the time of inclusion; active bleeding (requiring urgent intervention); severe comorbid conditions that prevent scheduled examination; mental illnesses that preclude the possibility of cooperation with the patient[12,13].

Clinical methods included a thorough medical history (identifying the causes of portal hypertension, history of bleeding, presence of ascites, manifestations of portal encephalopathy), physical examination with assessment of liver and spleen size, palpation of varicose veins, assessment of ascites, and determination of encephalopathy degree according to the West Haven scale. Assessment of bleeding was conducted using the Child-Pugh scale and the Model for End-Stage Liver Disease (MELD).

Instrumental methods included ultrasound examination with portal system Doppler ultrasonography (assessment of portal vein diameter, blood flow velocity and direction, identification of esophageal and gastric varices), transient elastography, multi-detector computed tomography with contrast enhancement and assessment of portal and splenic blood flow, and magnetic resonance imaging with contrast when indicated. Endoscopic methods included fibroesophagogastroduodenoscopy to identify and assess the severity of esophageal and gastric varices (Paquet and Japanese Research Society classification).

Molecular-genetic and biochemical methods included determining the levels of liver function markers (total bilirubin, direct bilirubin, alaninaminotransferase, aspartate aminotransferase, alkaline phosphatase, gamma-glutamyltranspeptidase), protein fractions, prothrombin time and international normalized ratio, albumin levels, determining portal hypertension markers (splenomegaly, ascites, varicose veins), and studying genetic polymorphisms associated with the progression of liver disease (if possible).

Statistical processing of the data was performed using the SPSS software package version 26.0 and R version 4.0. Parametric (Student's t-test) and non-parametric (Mann-Whitney, Kruskal-Wallis test) methods were used for quantitative variables. Qualitative indicators were compared using the $\chi 2$ (chi-square) criterion with a Yeats correction. Survival analysis was conducted using the Kaplan-Mayer method with the determination of the logarithmic rank criterion. Differences were considered statistically significant at $p < 0.05$.

Results

Within the framework of the conducted study, 240 children aged 1 to 18 years with verified portal hypertension were examined. The main group consisted of 130 patients (average age 9.3±4.1 years), and the control group consisted of 110 children (average age 9.8±4.3 years). In terms of gender composition, no differences were identified between the groups ($p > 0.05$).

Regarding the etiology of portal hypertension, the examined patients were distributed as follows. Congenital

anomalies of portal vein development (atresia, hypoplasia, cavernous malformation) were diagnosed in 96 patients (40.0% of the total sample). Liver cirrhosis of various etiologies was identified in 78 children (32.5%), of which biliary cirrhosis was identified in 32 patients (13.3%), alcoholic cirrhosis due to prenatal exposure in 14 children (5.8%), cirrhosis in chronic viral hepatitis B and C in 18 patients (7.5%), autoimmune cirrhosis in 8 children (3.3%), and cirrhosis in cases of hereditary metabolic disorders in 6 patients (2.5%). Non-hepatic portal block (Budd-Chiari syndrome, portal vein thrombosis, hepatic vein stenosis) was diagnosed in 42 patients (17.5%). Other etiological factors (portal hypertension in pancreatitis, pancreatic tumors, splenomegaly of unknown etiology) were recorded in 24 children (10.0%).

Clinical and anamnesis analysis revealed that 168 patients (70.0%) had portal hypertension before the age of 10. The manifestation of the disease with bleeding from esophageal varices occurred in 82 patients (34.2%), including massive bleeding requiring hemotransfusion in 44 children (18.3%). Ascites of varying severity were found in 154 patients (64.2%), including intense ascites in 64 children (26.7%). Grade III-IV splenomegaly was identified in 198 patients (82.5%). Signs of portal encephalopathy (astericcisis, sleep disturbances, decreased psychomotor activity) were found in 38 patients (15.8%), including acute liver encephalopathy of 3-4 degrees of severity in 8 children (3.3%).

During ultrasound examination with Doppler ultrasonography, it was determined that the diameter of the portal vein averaged 10.2±2.3 mm (norm 6-8 mm), and the maximum blood flow rate in the portal vein decreased to 15.4±5.2 cm/sec (norm 20-30 cm/sec). 198 patients (82.5%) had esophageal varices, 124 children (51.7%) had varices of the stomach, and 68 patients (28.3%) had varices of the large intestine.

Based on the results of the endoscopic examination, esophageal veins were classified by severity: small varicose veins (1–2 mm) were identified in 42 patients (17.5%), medium varicose veins (2–5 mm) in 98 children (40.8%), and large veins (>5 mm) with signs of high bleeding risk in 58 patients (24.2%). Analysis of liver function indicators in the main group revealed: total bilirubin level 32.4±15.2 μmol/L (norm <20 μmol/L), aspartate aminotransferase 58.3±24.6 U/L (norm <40 U/L), albumin level 28.5±6.3 g/L (norm 35–50 g/L), prothrombin time 18.2±4.1 sec (norm 11–14 sec).

Among the 130 patients in the main group receiving comprehensive treatment, 84 children (64.6%) received conservative therapy (beta blockers, aldosterone antagonists, diuretics, and antiviral treatment if necessary). Endoscopic ligation of esophageal varices was performed in 56 patients (43.1%), and endoscopic sclerotherapy in 38 children (29.2%). Transjugular intrahepatic portosystemic shunting (TIPS) was performed in 18 patients (13.8%) with recurrent bleeding not controlled by endoscopic therapy. Distal splenorenal bypass surgery was performed in 12 patients (9.2%) with well-preserved liver function and objective indications for surgery[14].

When evaluating the long-term treatment results during the 4-6 year observation period, the following was established. The mortality rate in the main group was 8.5% (130 out of 11 patients), which was significantly lower than in the control group (19.1%, 21 out of 110 patients; $p < 0.01$). The median survival in the primary group was 5.8±1.2 years, and in the control group, 3.4±1.6 years. Recurrent bleeding from varicose veins developed in 24 patients in the main group (18.5%) and 58 patients in the control group (52.7%; $p < 0.001$). Progression of portal encephalopathy was noted in 18 patients in the main group (13.8%), compared to 42 patients in the control group (38.2%; $p < 0.001$).

When analyzing mortality risk factors, the following critical variables were established: albumin level less than 25 g/l (relative risk 4.2; 95% confidence interval 2.1-8.4), prothrombin time more than 18 seconds (OR 3.8; CI 1.9–7.6), and the development of acute hepatic encephalopathy (OR 5.6; CI 2.4–13.2), the presence of large esophageal varices (OR 2.4; CI 1.2–4.8).

When assessing the quality of life in the main group of patients, a significant improvement in indicators of physical activity, school attendance, and social adaptation was noted compared to the control group ($p < 0.05$).

Discussion

The obtained results indicate the leading role of timely diagnosis, comprehensive treatment, and monitoring of patients with portal hypertension in improving long-term results. A significant decrease in mortality in the main group receiving comprehensive treatment confirms the effectiveness of a modern multidisciplinary approach combining conservative therapy, endoscopic interventions, and surgical treatment when indicated[15].

The identified risk factors for mortality (low albumin levels, prolonged prothrombin time, development of acute encephalopathy, presence of large varicose veins) allow for stratification of patients by severity and determine the need for more aggressive treatment. The reduction in recurrent bleeding rates in the primary group from 52.7% to 18.5% emphasizes the effectiveness of endoscopic ligation of varicose veins as a method for preventing complications.

Conclusion

A comprehensive approach to the diagnosis and treatment of portal hypertension in children, based on risk stratification, integrated use of conservative therapy, endoscopic interventions, and surgical treatment, significantly improves long-term outcomes and patients' quality of life. The identified mortality risk factors should be utilized in selecting individual treatment tactics and patient monitoring intensity.

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