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EXPERIENCE IN TREATMENT OF EXTRAHEPATIC PORTAL HYPERTENSION IN CHILDREN

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Summary

Extrahepatic portal hypertension accounts for up to 80% of all causes of portal hypertension in children and is associated with obstruction of hepatopetal blood flow at the portal vein.

Objective. To analyze the results and effectiveness of surgical treatments for extrahepatic portal hypertension in children.

Materials and methods. From 2016 to 2024, 106 children with portal hypertension were treated at the Republican Scientific and Practical Center for Pediatric Surgery (Minsk, Belarus). The average age of patients was 6,5 years (1 year – 15 years).

Results. Meso-Rex bypass was performed in 15 patients with extrahepatic portal vein obstruction. The operation was possible in 15/42 patients, which was 35,7%. In two children, stenosis of the upper (portal) anastomosis developed in the postoperative period. Splenorenal shunt was performed in 25 patients. Modifications were used in the form of distal splenorenal shunt in 13 patients and side-to-side anastomosis in 12 children. Thrombosis of the distal splenorenal shunt occurred in 2 patients (15%). H-mesocaval bypass was performed in 15 patients with widespread thrombosis and cavernous transformation of the splenoportal trunk, when other surgical options were impossible. Shunt thrombosis in the early postoperative period occurred in one patient. Endoscopic ligation of esophageal varices was performed in 16 patients, 29 manipulations with ligation of 1 to 5 nodes were performed. Endoscopic sclerotherapy of esophageal veins was performed in 89 patients. As a first-line therapy, endoscopic sclerotherapy was used in 17 patients with intrahepatic portal hypertension.

Conclusion. Meso-Rex bypass is a radical treatment of extrahepatic portal hypertension and, according to our data, is possible in 35,7% of patients. If Meso-Rex bypass cannot be performed, the operation of choice is splenorenal shunt: distal or side-to-side when the diameter of the splenic vein is less than 5 mm. Indications for performing H-mesocaval shunting are widespread portal system thrombosis, as well as thrombosis of previously performed vascular anastomoses. Endoscopic methods of hemostasis and preventing gastroesophageal bleeding remain the treatment of choice for children with chronic liver disease and intrahepatic portal hypertension.

Keywords. *Extrahepatic portal hypertension, meso-Rex bypass, portosystemic shunts, distal splenorenal shunt, H-type mesocaval shunt.*

Extrahepatic portal hypertension (EPH) accounts for up to 80% of all causes of portal hypertension syndrome (PHS) in children and is associated with hepatopetal blood flow obstruction at the level of the portal vein (PV). More than 70% of patients with EPH first seek medical care with the clinical presentation of gastroesophageal reflux (GFR), which most often manifests before the age of 10 years [1, 2]. Advances in drug therapy and endoscopic treatment of esophageal varices (EVs) have reduced the need for surgical interventions in children with EPH. However, with the widespread use of endoscopy, the appearance of ectopic varices and the development of severe portal gastropathy have become a new challenge in the treatment of the disease [3].

According to the Baveno VI international consensus, mesoportal shunting (MPS) is the method of choice for the prevention of PZH in children with HSV and is a radical method of treating the disease [4]. However, this method of reconstructing portohepatic blood flow does not solve the problem of treating children with HSV, since the operation can be performed only in 34.6% of patients in whom the left branch of the PV is patent [5]. For the majority of patients, portosystemic shunting (PSS) operations remain the most effective treatment method. Portosystemic anastomoses effectively reduce portal pressure and lead to regression of symptoms and reliable prevention of PZH. Growing experience of pediatric surgeons in using PSS operations in young children confirms the reliable and long-term effect of these interventions. They have a significant advantage over repeated sessions of endoscopic interventions, and the neurophysiological consequences of these operations are minimal [6]. The choice of the PSS operation option is largely determined by the anatomy of the portal system and the clinical manifestations of the disease.

The aim of the study. To analyze the results and effectiveness of surgical treatments for HSV in children.

Material and methods. From 2016 to 2024, 106 children with HSV were treated at the Republican Scientific and Practical Center for Pediatric Surgery (Minsk, Belarus). The average age of patients was 6.5 years (range, 1–15 years). Most patients had HSV infection—85 (80%). Intrahepatic block due to parenchymal liver disease was the cause of HSV in 21 (20%) children.

The patient examination plan included standard general clinical examinations with an assessment of liver function and the hemostasis system, esophagogastroduodenoscopy (EGD), methods of visualizing the portal vein system: abdominal ultrasound with Doppler, multispiral computed tomography with angio-enhancement (MSCT-A), ultrasound of the neck vessels, and liver puncture biopsy.

Results and discussion. MPSh was performed in 15 patients with extrahepatic venous variceal obstruction. The average age of the patients was 5 years (minimum age – 2 years). According to EGDS, they had grade 3 varices with dilated submucosal veins of the subcardia; 9 patients had a history of bleeding. Severe course of the disease was also indicated by significant splenomegaly and severe hypersplenism (decrease in the white blood cell count below 3.0×109/L, platelet count – below 60×109/L). Preoperative examination of patients included: general clinical tests, assessment of coagulation system parameters and biochemical parameters of liver function, liver biopsy (if indicated), ultrasound of the portal system with Doppler ultrasonography and MSCT-A of the abdominal cavity to assess the patency of the venous variceal system and its intrahepatic branches. All patients, according to ultrasound data, had a normal structure of the liver parenchyma and reference values for laboratory parameters of liver function.

The operation consisted of bypassing the obstructed area of the MV and cavernoma by interposition of a graft from the patient's own left internal jugular vein between the superior mesenteric vein and the left branch of the MV. Since the liver structure and hepatic sinusoidal resistance are normal in these patients, direct bypass of the thrombosed area of the MV and subsequent restoration of physiological hepatopetal blood flow can radically alleviate the symptoms of portal hypertension [7, 8].

The final decision to perform MPS was made immediately during the procedure. The choice of this procedure was determined by the presence of a patent left branch of the cerebral vein with a diameter greater than 4 mm and intense retrograde blood flow during intraoperative exploration in the sinus Rex (Figure 1).

The spread of thrombosis to the left branch of the internal jugular vein is the main limiting factor for performing this procedure in children with HSV. In our experience, the procedure was feasible in

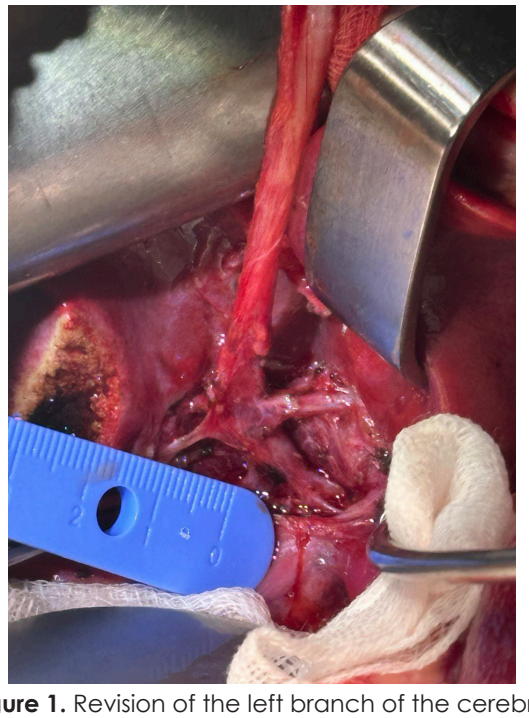

Figure 1. Revision of the left branch of the cerebral artery in the sinus Rex

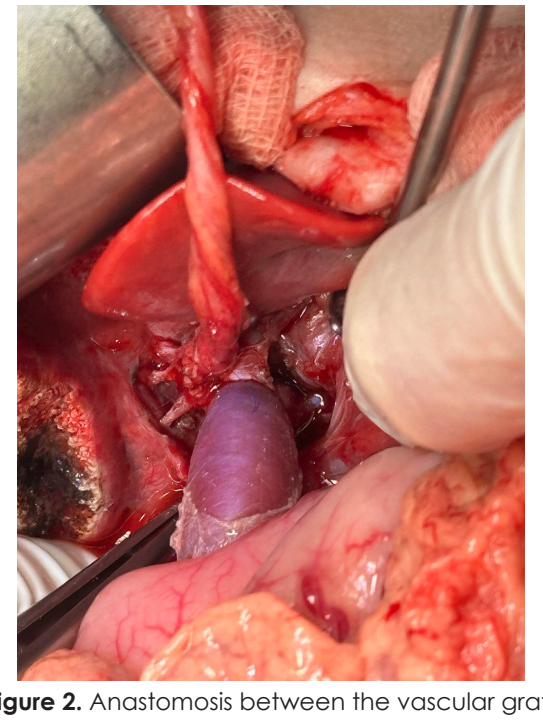

Figure 2. Anastomosis between the vascular graft and the left branch of the vascular vein

15 of 42 patients, representing a success rate of 35.7%. In all cases, the left internal jugular vein was used as a venous conduit between the superior mesenteric and left branches of the internal jugular vein (Figure 2).

The average surgical time was 320 ± 15 minutes, and intraoperative blood loss was 100 ± 30 ml. Low-molecular-weight heparins were used for the first 10 days postoperatively to prevent shunt thrombosis. The average length of stay in the intensive care unit was 2 days.

In the early postoperative period, ultrasound with Doppler imaging confirmed shunt patency in all cases. Postoperative complications, such as persistent chyloperitoneum, occurred in one child, requiring long-term conservative treatment. Follow-up examinations at 3 and 6 months after surgery demonstrated good shunt function: with an average diameter of 7.2 ± 2.1 mm, the average blood flow velocity was 43.6 ± 5.6 cm/s, and the volumetric blood flow was 840 ± 90 ml/min. Restored hepatopetal blood flow contributed to a gradual increase in the diameter of the intrahepatic branches of the hepatic vein and a decrease in the number of collaterals in the porta hepatis region.

Effective decompression of the portal system was confirmed by a decrease in spleen size. All patients showed a gradual resolution of hypersplenism: 6 months after surgery, the white blood cell count was 4.4×109/L, and the platelet count was 188×109/L ($p<0.005$).

There were no recurrences of bleeding from the varices during the observation period. Follow-up EGDSs at 3 and 6 months after MPS demonstrated positive results: a reduction in the severity of varices and regression of congestive portal gastropathy. Endoscopic treatment after MPS allowed for rapid and complete elimination of the varices, which, in our opinion, is essential for out-of-town patients living far from specialized medical care centers.

Two children developed stenosis of the upper (portal) anastomosis postoperatively. Clinically, this complication manifested as an enlarged spleen and the development of hypersplenism one year after surgery. MSCT-A revealed stenosis of the hepatic portion of the bypass graft of up to 3 mm with the development of splenomegaly (Figure 3).

Both patients underwent balloon angioplasty and stenting of the stenosis area with good angiographic (diameter increase up to 8 mm) and clinical results. In a systematic review of the literature conducted by S. Zielsdorf et al. [9] including 22 sources and 461 patients with HSV, a comparative analysis showed that the incidence of MPS thrombosis was 14.1% (48/340), and after PSS operations – 5.8% (7/121). At the same time, 11.8% (40/340) of patients after MPS required at least one invasive intervention due to stenosis or thrombosis of the bypass graft, and in 20/40 patients, MPS conversion to PSS was performed.

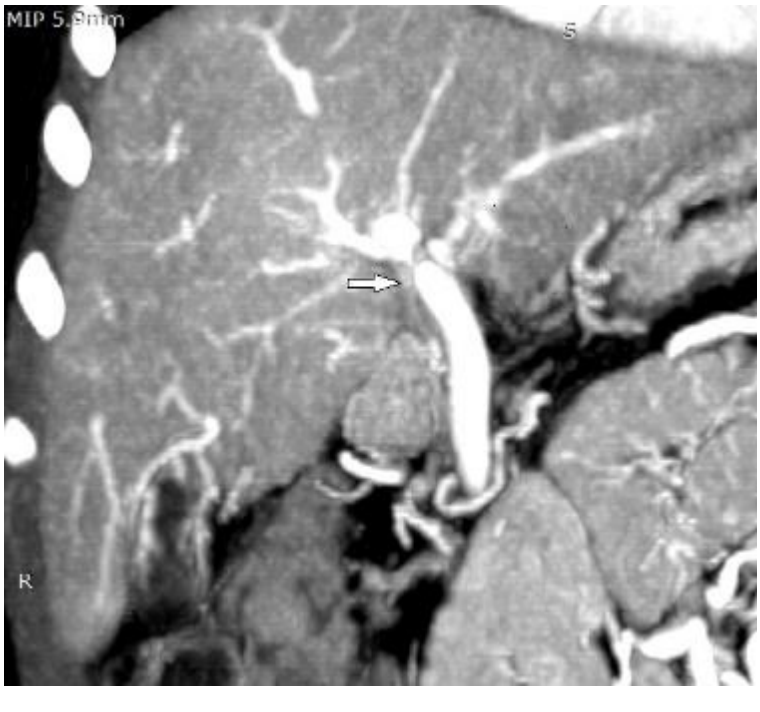

Figure 3. MSCT-A (venous phase). Stenosis of the portal anastomosis of the MPS (indicated by the arrow)

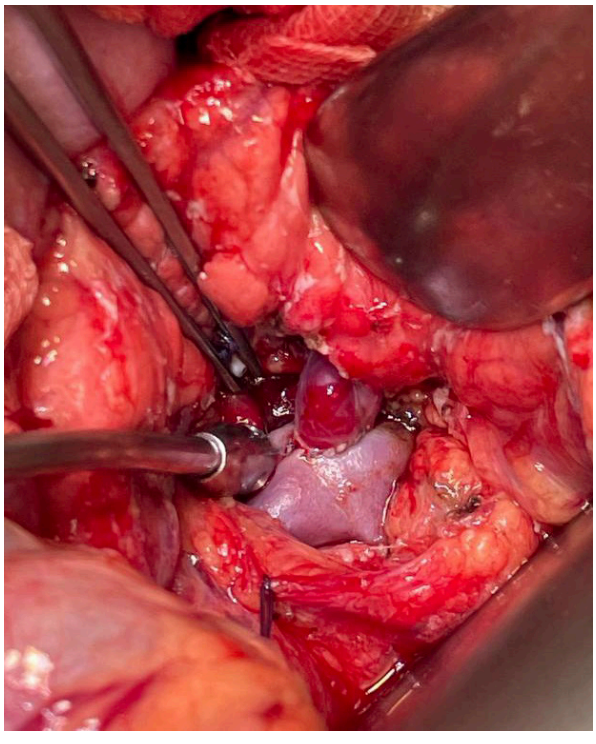

Figure 4. DSRA

Splenorenal bypass was performed in 25 patients with an average age of 6 years (minimum age 1.5 years). Modifications were used in the form of a distal splenorenal shunt (DSS) in 13 children and a side-to-side splenorenal anastomosis in 12 children. Indications for surgery were significant splenomegaly and severe hypersplenism (pancytopenia in a complete blood count), and in 13 patients, a history of subcutaneous fat hemorrhage and grade 3 varices. When performing emergency surgical interventions to stop recurrent subcutaneous fat hemorrhage, DSS was combined with gastrotomy and suturing of varices of the subcardia and esophagus in 5 cases (Figure 4). The average anastomotic diameter was 7 mm. Surgical hemostasis was achieved in all cases. There were no fatal outcomes.

As a selective portocaval anastomosis, DSRA has advantages over non-selective shunts, as it leads to decompression of the gastrosplenic basin, but at the same time preserves mesenteric blood flow and high pressure in the superior mesenteric vein [10]. Doppler ultrasound data show that in the immediate period after DSRA, it is possible to maintain slow hepatopetal blood flow in the portal vein basin – the average linear blood flow velocity was 10.1 ± 4.3 cm/s, indicating high pressure in the mesenteric venous system. Blood flow depletion in the portal vein was noted to a lesser extent than after side-to-side splenorenal anastomosis. At the same time, there are studies proving that DSRA, despite careful splenic-pancreatic disconnection, remains selective only in the early postoperative period and, on average, after 2–3 years, loses its selectivity due to the formation of new collaterals between the two basins with a pressure gradient [11].

Splenorenal anastomosis "side to side" is an effective and reliable option for PSSh. Some authors [12] consider splenorenal anastomosis "side to side" (Mitra shunt) to be a selective operation, however, most likely, it occupies an intermediate position, since according to the results of ultrasound with Dopplerography, it leads to a decrease in the parameters of hepatopetal blood flow [13]. This operation option should be considered in patients with a splenic vein diameter of less than 5 mm. The ability to perform a wide anastomosis with additional blood flow from the central part of the splenic vein determines the high patency of this anastomosis in the postoperative period. In the presence of a wide left adrenal vein, splenorenal shunting was performed using its stump (n = 9), which simplifies the intervention and helps prevent vascular kinking.

For adequate functioning of the anastomosis, we emphasize the following principles that must be observed: the anastomosis must be performed with a diameter of 7-10 mm using monofilament threads PDS-7/0 and strict adherence to all the rules for applying a vascular suture, sufficient mobilization and correct topical application of the anastomosis are necessary to prevent the formation of an angle

or rotation of the vessels around the axis after the restoration of normal anatomical relationships, in the early postoperative period, hypocoagulation must be maintained using low molecular weight heparins, and after discharge from the hospital, aspirin is prescribed at a dose of 3-5 mg / kg / day for 3 months.

The average postoperative follow-up period was 2.5 years. Follow-up ultrasound examinations were performed at 1 and 6 months postoperatively. Patency of the splenic vein junction was confirmed in all patients. The average linear blood flow velocity in the splenic vein after surgery was 37.2 cm/s. High blood flow in the splenic vein after surgery facilitates adequate decompression of esophageal and gastric varices.

According to EGDS data, the severity and tension of esophageal varices decreased after surgery in all patients. After DSRA, the degree of varices was not as pronounced as with total bypasses; however, variceal decompression was sufficient to prevent recurrence of subcutaneous varices. We noted that performing esophageal vein endoskeletal sclerosis in these patients leads to rapid elimination of varices, which is likely due to effective decompression of the gastroesophageal basin. No recurrences of subcutaneous varices were observed during the observation period. DSRA thrombosis occurred in 2 patients (15%), an average of 2 years after surgery.

An N-mesocaval shunt was performed in 15 patients with HSV, with an average age of 5 years. Indications for the surgery included previous subcutaneous gastritis (SGI), grade 3-4 esophageal varices with "red marks" and a risk of bleeding. This surgical option was the procedure of choice in patients with extensive thrombosis and cavernous transformation of the splenoportal trunk, when other PSSh options were impossible (Figure 5). In 4 patients, the splenic vein diameter was less than 5 mm, which precluded splenorenal shunting with a reliable long-term outcome.

H-mesocaval bypass grafting (H-MCBG) may be the procedure of choice for repeat interventions following thrombosis of previously performed anastomoses. In our series, DSRA thrombosis was an indication for surgery in two patients. During the repeat intervention, splenectomy with H-mesocaval bypass grafting was performed. The left internal jugular vein was used as a vascular graft in all cases. The final decision on the surgical option was made after revision of the left branch of the internal jugular vein and the impossibility of performing MPBG. The caval anastomosis was performed first, as it is the most complex and lies deep in the wound—in the "well" of the retroperitoneal space. The final insertion length was determined after removal of retractors from the retroperitoneal space–the graft should be shortened to the optimal length, avoiding both excess and excessive tension (Figure 6).

Postoperatively, low-molecular-weight heparins were administered for 10 days to prevent shunt thrombosis. Prolonged chylous ascites (3 weeks postoperatively) was observed in one patient,

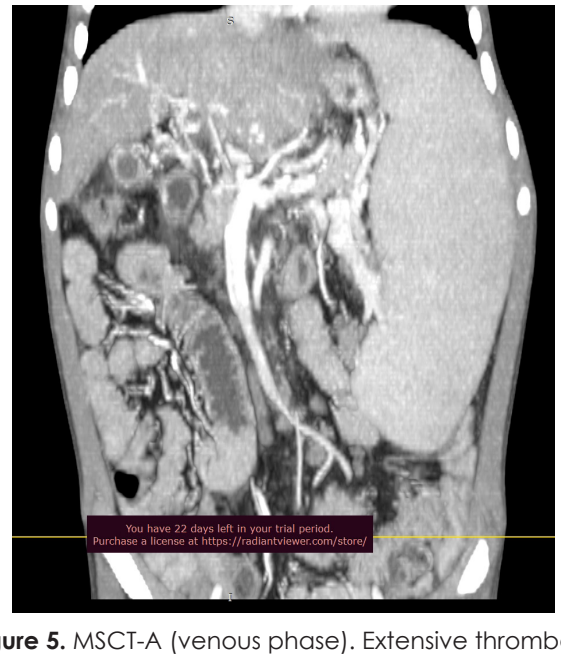

Figure 5. MSCT-A (venous phase). Extensive thrombosis and cavernoma of the splenial-portal trunk, preserved patency of the superior mesenteric vein

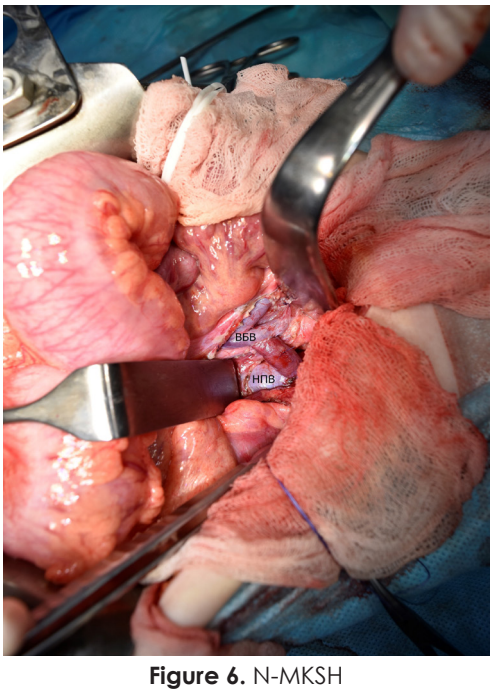

Figure 6. N-MKSH

requiring a low-fat diet, octreotide, and diuretics. Shunt thrombosis occurred in one patient in the early postoperative period. We attribute the failure to COVID-19 infection and severe virus-associated hypercoagulability that developed on day 5 postoperatively.

The first postoperative follow-up was performed 3 months after surgery. All patients with a patent shunt demonstrated a significant reduction in spleen size and resolution of hypersplenism. Post-shunt patients had normal liver function tests. Parents noted a reduction in abdominal volume, as well as accelerated physiological growth and improved physical parameters. Several studies have demonstrated that PSSh, like MPSh, improve growth and weight, which is associated with decompression of the spleen and mesenteric system [14, 15].

The average follow-up period after surgery was 18 months. There were no recurrent bleeding episodes. According to endoscopy data 3-6 months after surgery, complete reduction of esophageal and gastric varices was observed in 10 children, and a significant reduction in the severity of varices and the disappearance of "red marks" were observed in 5 children. Numerous studies confirm that N-MCSH is effective in preventing variceal bleeding, primarily due to effective decompression of the portal system [16, 17].

Most researchers note a very high long-term patency of the N-MCS – 95%, which in the long term exceeds the patency of the MPS (86-90% with strict patient selection) and the DSRA (90–92%) [9]. This is due to the high pressure gradient between the portal vein and the IVC and the use of sufficiently wide grafts (8–10 mm) to reduce resistance to blood flow.

No symptoms of portosystemic encephalopathy were observed in the postoperative period. To assess the degree of intellectual disability (latent encephalopathy), standardized psychometric tests (lines test and numbers test) were used in patients over 10 years of age. In the study of A. Yu. Razumovsky et al. [5], summarizing the experience of treating 718 children with HSV who underwent 657 PSSh operations (including 167 N-MKSh), it was proven that portosystemic encephalopathy in the postoperative period develops extremely rarely and is manifested by minimal behavioral disorders. At the same time, minimal portosystemic encephalopathy is observed in 32–45% of children with HSV and without PSSh operations [18]. In the work of A. Srivastava et al. [19] showed that minimal PE was recorded in 41% of patients with HSV after central splenorenal anastomosis compared to 32% of cases in non-operated children (the difference is not significant). Moreover, patients after CSRA had higher blood ammonia levels and glutamine/creatine ratio on H-MRI brain scans than non-operated patients.

Endoscopic ligation of varices was performed in 16 patients, with an average age of 9.5 years (the youngest was 3 years old). A total of 29 procedures were performed, ligating from 1 to 5 knots. Boston Scientific Speedband Superview Super 7 multi-shot ligators with seven preloaded rings were used. Urgently indicated, ligation at the peak of bleeding was performed in 3 patients (19%), with reliable hemostasis achieved in 2 cases, and emergency surgery required in one child. The intervals between sessions were 1-2 months. A complication of the procedure, active bleeding, developed in 3 patients. In all these cases, a control endoscopic examination revealed leakage and disruption of the latex rings in the area of active bleeding (Figure 7).

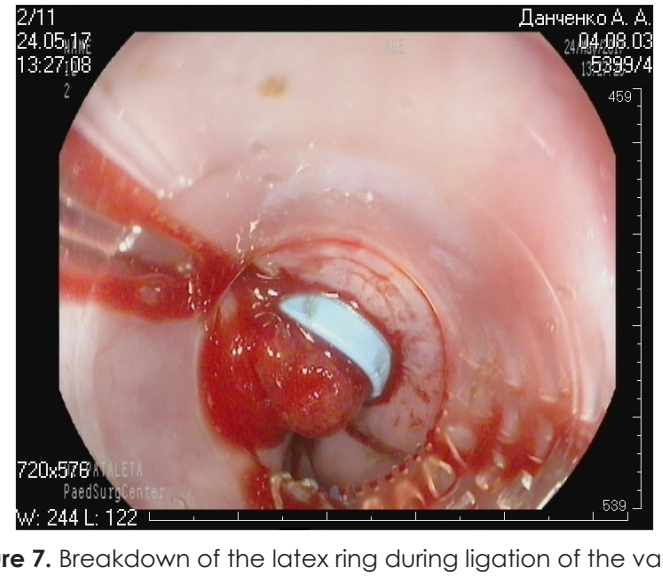

Figure 7. Breakdown of the latex ring during ligation of the varices

Endoscopic sclerotherapy of esophageal varices was performed in 89 patients, with 295 procedures performed. The average patient age was 6 years (the youngest was 1.5 years). Endosclerotherapy was used as first-line therapy in 17 patients with intrahepatic portal hypertension, as well as in 33 children with HSV who had grade 1-2 esophageal varices without a history of bleeding. In 5 cases, endoscopic sclerotherapy was performed as an emergency measure to arrest active subcutaneous fat. The technique was also used post-surgically to eradicate residual varices. In 36 patients, sclerotherapy was performed using ethoxysclerol foam. The foam sclerosant was prepared according to L. Tessari's method, mixing room air with a 1-2% ethoxysclerol solution in a 4:1 ratio. After injection, the foam immediately spreads through the varicose veins and vascular tracts, displacing blood from them, which facilitates prolonged contact between the sclerosant and the endothelium. There were no complications. A positive effect of foam sclerotherapy was noted in all patients. Endoscopic monitoring three months after the procedure revealed rapid obliteration of the venous trunks and the absence of "red marks": complete eradication of varicose veins was observed in 20 patients (55%), and significant reduction was observed in 16 (45%).

Conclusions. The anatomy of the portal system and the clinical manifestations of the disease are determining factors when choosing the surgical treatment option for children with HSV.

Revision of the left branch of the splenic vein should be a mandatory step in surgical intervention for HSV. MPS is a radical treatment for HSV. If MPS is not possible, the surgery of choice is splenorenal bypass grafting (SRS) or side-to-side splenic vein bypass (SRS) if the splenic vein diameter is less than 5 mm.

N-MCSH in children with HSV is an effective PSSH procedure. Indications for its performance include widespread portal vein thrombosis, as well as thrombosis of previously performed vascular anastomoses.

Endoscopic treatments for varicose veins remain the leading treatment for children with chronic liver disease and intrahepatic portal hypertension. In patients with HSV, endoscopic interventions can be used for stage 1-2 varicose veins, as well as in the postoperative period for complete eradication of varicose veins.

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