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Research Article

Congenital portosystemic shunts: surgical treatment experience

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Abstract

Congenital porto-caval shunts are rare and may have a different morphological structure (intra- and extrahepatic shunts, with or without portal blood flow). The main method of treating patients with this pathology is endovascular shunt occlusion. However, in some cases, this method is ineffective.

The article contains a description of six clinical examples of surgical treatment of congenital porto-systemic shunts in children. In the diagnosis of congenital portosystemic shunts, the leading role belongs to Doppler ultrasound, multislice computed tomography, and angiography. The indication for surgical treatment was the anatomical features of the shunt, which makes endovascular occlusion technically impossible. In one observation a wide Arantian duct was diagnosed, its open ligation was performed. In another case, the portal vein emptied directly into an aneurysmal dilatation, performed reconstructive plastic surgery on the vessels of the portal vein. In the next observation, a pronounced retrograde blood flow was determined along the dilated inferior mesenteric vein, blood was discharged through the sacral plexus into the internal iliac vein. The left internal iliac vein was isolated and ligated, the dysplastic inferior mesenteric vein was ligated and partially removed. In 2 patients, the portal vein flowed directly into the inferior vena cava in the area of aneurysmal expansion; an operation was performed — open ligation of the shunt. In one observation, a deep hypoplasia of the intrahepatic branches of the portal vein was diagnosed, and therefore the restoration of portal blood flow after the closure of the shunt is impossible. The child was sent to decide on a liver transplant.

Each case of congenital porto-caval shunts is unique. The surgeon determines the tactics directly during the operation, depending on the morphological structure of the organs, since the preoperative examination does not always give an unambiguous idea.

Keywords: case report; congenital portosystemic shunt; Abernathy malformation; surgical treatment; angiography; children.

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Научная статья

Врожденные портосистемные шунты: опыт хирургического лечения

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Аннотация

Врожденные портокавальные шунты встречаются редко и могут иметь различную морфологическую структуру (внутри- и внепеченочные шунты, с наличием или отсутствием портального кровотока). Основной метод лечения пациентов с данной патологией — эндоваскулярная окклюзия шунта. Однако в некоторых случаях подобный метод оказывается неэффективным.

В статье содержится описание шести клинических примеров хирургического лечения врожденных порто-системных шунтов у детей. В диагностике врожденных порто-системных шунтов ведущая роль принадлежит ультразвуковому исследованию с допплерографией, мультиспиральной компьютерной томографии, ангиографии. Показанием к оперативному лечению стали анатомические особенности шунта, делающие эндоваскулярную окклюзию технически невозможной. В одном наблюдении диагностирован широкий аранциев проток, была выполнена его открытая перевязка. В другом случае воротная вена впадала непосредственно в аневризматическое расширение, выполнена реконструктивно-пластическая операция на сосудах воротной вены. В следующем наблюдении определялся выраженный ретроградный кровоток по расширенной нижней брыжеечной вене, сброс крови через сакральное сплетение во внутреннюю подвздошную вену. Произведено выделение левой внутренней подвздошной вены и ее перевязка, дисплазированная нижняя брыжеечная вена перевязана и частично удалена. У двух пациентов воротная вена впадала непосредственно в нижнюю полую вену в области аневризматического расширения, проведена операция — открытая перевязка шунта. В одном наблюдении диагностирована глубокая гипоплазия внутрипеченочных ветвей воротной вены, в связи с чем восстановление портального кровотока после закрытия шунта невозможно. Ребенок был направлен для решения вопроса о трансплантации печени.

Каждый случай врожденных портокавальных шунтов уникален. Хирург определяет тактику непосредственно в ходе операции в зависимости от морфологического строения органов, поскольку предоперационное обследование не всегда дает однозначное представление.

Ключевые слова: клинический случай; врожденный портосистемный шунт; мальформация Абернети; оперативное лечение; ангиография; дети.

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ЭКО•ВЕКТОР

BACKGROUND

The Abernethy malformation is a rare congenital abnormality associated with a congenital portosystemic shunt (CPSS). The disease prevalence is 1 per 30,000 newborns [1]. This malformation has two variants: type I, when hepatic portal perfusion is completely absent, and type II when perfusion is partially preserved. Type I has two subtypes: IA, when the superior mesenteric vein and splenic vein end separately into the inferior vena cava (IVC) or other veins of the systemic circulation (left renal vein and left gastric vein), and subtype IB, when the superior mesenteric, and splenic veins form a common trunk that ends into the IVC. Type II implies partial preservation of portal perfusion of the liver, and there is a trunk of the portal vein and a portosystemic shunt (Fig. 1).

Depending on the angioarchitecture and shunt localization, extrahepatic (portocaval "end-to-side", "side-to-side", and H-shunts) and intrahepatic portosystemic shunts, which include the persistent Arantius' duct [2]. The leading method of treatment is the endovascular occlusion of the vicious shunt. This method is less traumatic, can significantly reduce the hospital stay, and is highly effective. In most cases, endovascular occlusion enables us to achieve a complete cure [3, 4]. However, in some cases, this technique is ineffective or impossible, and open surgery is required.

CASE DESCRIPTION

Twenty-four pediatric patients with CPSS were examined and treated in the Department of Endovascular Surgery, Surgery Department No. 2, and Microvascular Surgery No. 2 of the Russian Children's Clinical Hospital (RCCH) from 2017 to 2021. Endovascular occlusion was successfully performed in 18 of them. In six children, endovascular occlusion was not possible, and five of them underwent surgical correction of the defect, whereas in one case, the blockage of blood flow through the CPSS critically increased the pressure in the portal vein, which was associated with severe hypoplasia of its lobar branches; therefore, further attempts to eliminate CPSS were no longer performed. All six cases are presented in detail below.

Case 1

The patient was a 12-year-old boy at the time of hospitalization. *The anamnesis revealed* that the child was from the fifth pregnancy and the third-term delivery. In the late pregnancy, toxicosis, nephropathy associated with vegetative-vascular dystonia of the hypertensive type, and polyhydramnios occurred. His birthweight and length were 4300 g and 55 cm, respectively. The Apgar score was 7 points. The condition was severe because of respiratory distress syndrome.

At the age of 2 years, the patient was hospitalized in the Surgical Department of the Children's Regional Hospital of Tula. Examination findings led to the diagnosis of portal hypertension due to a malformation of the portal vein. Given the low degree of varicosities in the esophagus and varicose vein dynamics did not increase, he was under a follow-up until the age of 10, when complaints of recurrent abdominal pain, nausea, and pruritus became more frequent. The patient was hospitalized and examined in the N.F. Filatov Children's City Clinical Hospital No. 13, and he was diagnosed with Abernethy malformation type II. Subsequently, he was hospitalized in the RCCH Department of Surgery No. 2.

Upon admission, he had complaints of pruritus, recurrent abdominal pain, decreased appetite, asthenia, fatigue, periodic nausea, heartburn, belching, and headaches.

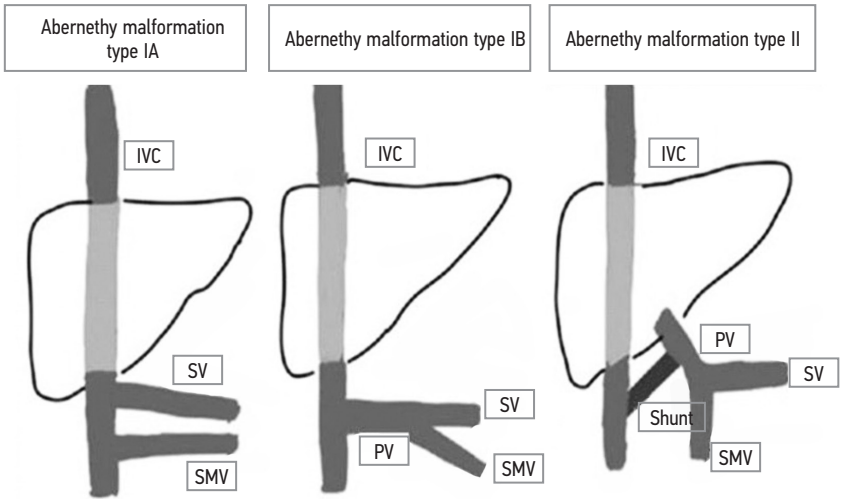


Fig. 1. Variants of Abernathy malformation. IVC — inferior vena cava; SV — splenic vein; PV — portal vein; SMV — superior mesenteric vein

Рис. 1. Варианты мальформации Абернети. НПВ — нижняя полая вена; СВ — селезеночная вена; ВВ — воротная вена; ВБВ — верхняя брыжеечная вена

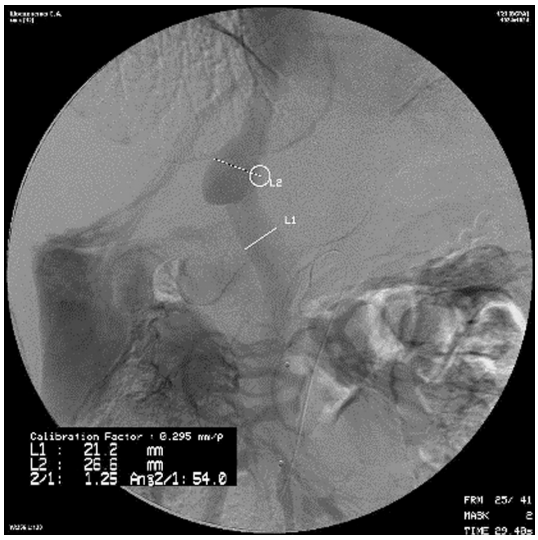


Fig. 2. Case No. 1. Mesentericoportography upper. Examination before surgery, measurement of the structures of the open Arantium duct
Рис. 2. Случай № 1. Мезентерикопортография верхняя. Исследование до оперативного вмешательства, измерение структур открытого аранциева протока

Examination. On ultrasonography (US), the liver was not enlarged. The anteroposterior sizes of the right and left lobes were 160 and 50 mm, respectively. The hepatic veins were obscurely differentiated. The main trunk of the portal vein was 15–16 mm, its right branch with an orifice into the IVC had a persistent Arantius' duct, and the left one had no abnormalities. Echo-signs of diffuse changes in the liver and malformation of blood vessels in the liver were noted.

The blood ammonia level was 342 μg/dL (norm maximum 110). Indirect liver elastometry showed that in 10 measurements of liver elasticity, the values ranged from 7.8 to 11.8 kPa, with a median of 9.6 kPa, indicating altered liver elasticity. The results indicated fibrosis F3 according to the METAVIR scale [5], with a reliability of 100%.

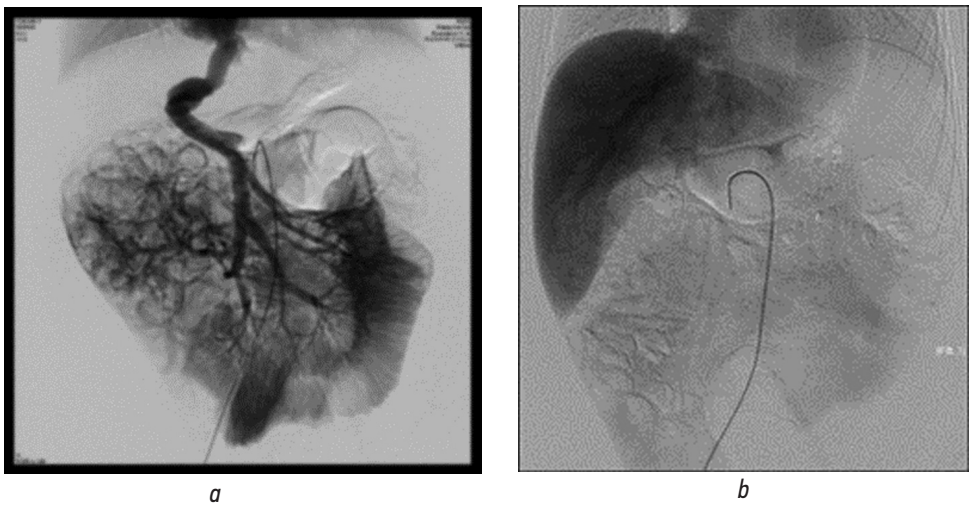
Angiography (Fig. 2) showed that a balloon catheter was placed in the IVC at the level of the portocaval anastomosis. The balloon was inflated, and the portocaval anastomosis was temporarily occluded. Direct portography was performed. The intrahepatic branches of the portal vein were not defined. No contrast agent was released into the IVC. As the portocaval anastomosis was occluded, the pressure in the portal vein was >30 cm WG.

A case conference was performed to discuss the examination results and determine further approaches. The diagnosis was deep hypoplasia of intrahepatic branches of the portal vein in a pediatric patient with CPSS; therefore, restoration of portal blood flow after CPSS closure was impossible. As the CPSS was occluded, the pressure in the portal vein increased to 30 mm WG (water gauge) or more (hypertension). Thus, the elimination of the CPSS was inexpedient. To address the issue of liver transplantation, the child was referred for a consultation to the B.V. Petrovsky Russian Scientific Center of Surgery.

Case 2

The patient was a boy who was 2 years 7 months old at the time of the initial hospitalization in the RCCH.

The anamnesis revealed prolonged (2 months) jaundice with hyperbilirubinemia during the neonatal period. Dispensary US at the age of 1 year revealed a mass lesion in the liver. Contrasted computed tomography revealed a vascular trunk (non-obliterated Arantius' duct) between the portal vein and the IVC, without pathological accumulation of the contrast agent. In addition, the child had retarded psychomotor development.



a *b*

Fig. 3. Case No. 2. Mesentericoportography upper: *a* — examination before surgery; *b* — examination after 2.5 years. Portal blood flow restored
Рис. 3. Случай № 2. Мезентерикопортография верхняя: *a* — до оперативного вмешательства; *b* — через 2,5 года. Портальный кровоток восстановлен

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Examination. US revealed an enlarged right liver lobe. The anteroposterior sizes of the right and left lobes were 111 and 32 mm, respectively. The main trunk of the portal vein was 8 mm, with single collaterals up to 1 mm parallel to it. A non-obliterated Arantius' duct with a diameter of 9 mm was identified. The spleen was enlarged (75 × 26 mm), with granular parenchyma, and moderately increased echogenicity. The diagnosis was non-obliterated Arantius' duct and echo-signs of extrahepatic portal hypertension.

Angiography (Fig. 3, *a*) showed that intrahepatic branches of the portal vein were hypoplastic and contrasted after the occlusion of the venous duct with a balloon. The duct ran (up to 14 mm in diameter) from the portal vein to the hepatic vein. The liver parenchyma was contrasted. The pressure in the portal vein system was measured after the occlusion of the venous duct with a balloon, measuring 22.5 cm WG.

The blood ammonia level was 367 μg/dL (norm maximum 110). Hyperammonemia was caused by the discharge of blood from the portal vein into the IVC, with the depletion of the hepatic blood flow. This resulted in encephalopathy and delayed psychomotor development. The level of bile acid was 177.6 μmol/L (norm <10).

According to the consultation with the involvement of the staff of the Department of Endovascular Surgery, due to the large width of Arantius' duct, it was technically impossible to perform an occlusion of the latter. Thus, an open ligation of the latter was performed.

Surgery. A laparotomy was performed with a right subcostal incision. The liver had normal size, color, and elastic consistency. Multiple inclusions of connective tissue and areas of induration were noted. The diameter of the portal vein was 9 mm; it was represented by one trunk to the hepatic hilum, and to the side, the IVC passes into Arantius' duct with a diameter of approximately 6 mm. The portocaval shunt was placed on the tourniquet, and its lumen was blocked. Moreover, no visible disorders in the venous outflow from the intestines or changes in the blood supply to the liver were noted. The portocaval shunt was ligated distal to the origin of the lobar branches of the portal vein (Fig. 4). The course of the postoperative period was uneventful.

On US with duplex examination of the vessels of the portal system (postoperative day 8), the size of the liver was 110 × 38 mm. The blood flow through the portal vein was antegrade, with a velocity of 10 cm/s. The lumen of the IVC was clear. The child was discharged for an outpatient follow-up.

After 1.5 years, the patient was hospitalized for a control examination. The condition was satisfactory. The child was growing and developing according to his age.

Examination. The US with duplex examination of the vessels of the portal system showed an enlarged liver, with anteroposterior sizes of the right and left lobes of 120 and 53 mm, respectively. The blood flow through the portal vein was antegrade, with a velocity of 12 cm/s. The outflow from the liver was not impaired. The blood flow through

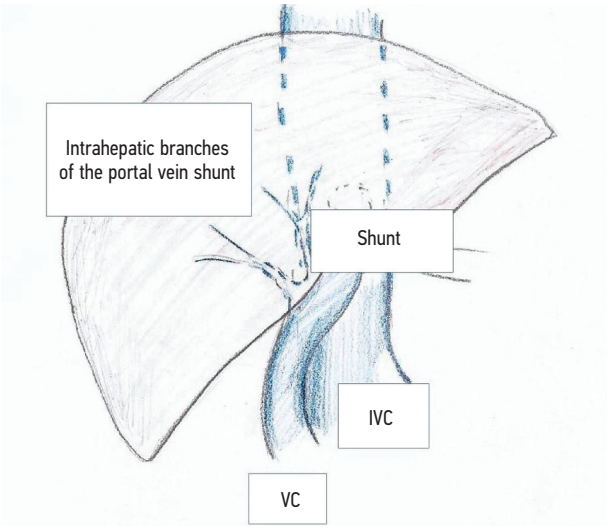


Fig. 4. Case No. 2. Scheme of the operation. VC — portal vein; IVC — inferior vena cava
Рис. 4. Случай № 2. Схема операции. ВВ — воротная вена; НПВ — нижняя полая вена

Arantius' duct was not recorded. The IVC had a clear lumen in the subhepatic segment, with unaltered blood flow. The blood ammonia level was 96 μg/dL (normal). The child was discharged for an outpatient follow-up.

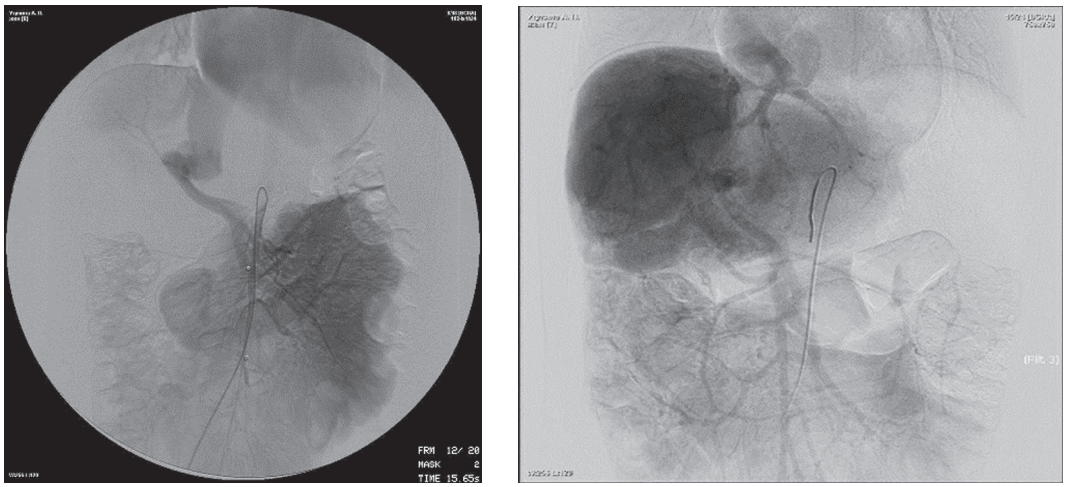
The patient was hospitalized a year later for a follow-up examination. TS with duplex examination of the vessels of the portal system showed antegrade blood flow through the portal vein, with a velocity of 20 cm/s. The spleen had normal size, the blood was flowing antegradely through the splenic vein, and its velocity was 18 cm/s. On angiography (Fig. 3, *b*), the portal vein was represented by a single trunk up to 10 mm in diameter. The intrahepatic branches of the portal vein were contrasted, and the blood flow through them was not impaired. A portosystemic shunt was not identified. Thus, the surgical treatment outcomes of a CPSS can be characterized as excellent.

Case 3

The patient was a 6-year-old girl hospitalized in the RCCH.

The anamnesis revealed that at the age of 1.5 months, the US at the primary healthcare facility revealed mass liver lesions (hemangiomas?). By the age of 1 year, these lesions have undergone involution. Skin hemangiomas (self-healing by 2 years) were also noted. At the age of 5, repeat US revealed diffuse focal changes in the liver. Throughout her life, the child experienced rapid fatigue and a delay in psychomotor development.

Examination in the RCCH. US showed slight enlargement due to the right lobe. The anteroposterior sizes of the right and left lobes were 112 and 35 mm, respectively. The contours were smooth. The parenchyma was fine and granular. In segments I, II, and IV, mass lesions of parenchymal density were determined, measuring 49 × 47, 30 × 25, and 30 × 20 mm, respectively, without capsules,



a *b*

Fig. 5. Case No. 3. Mesentericoportography upper: *a* — examination before surgery. An ungliterated arantium duct is visualized, flowing into an aneurysmal expansion located on the side of the inferior vena cava; *b* — examination after surgery. Complete restoration of portal blood flow

Рис. 5. Случай № 3. Мезентерикопортография верхняя: *a* — до оперативного вмешательства. Визуализируется необлитерированный аранциев проток, впадающий в аневризматическое расширение, находящееся со стороны нижней полой вены; *b* — после оперативного вмешательства. Полное восстановление портального кровотока

avascular, with unevenly increased echogenicity. The walls of the intrahepatic bile ducts were unevenly indurated. The gallbladder, pancreas, and spleen were normal. The conclusion was made on echo-signs of multiple lesions in the liver.

US with duplex examination of the vessels of the portal system revealed reduced outflow from the liver, and the hepatic branches were not visible. The main trunk of the portal vein was 5–6 mm in diameter, the right branch had no abnormalities, and the left one was deformed and narrowed. Arantius' duct was preserved, with active antegrade blood flow into the IVC, which showed no visible pathology.

Angiography (Fig. 5, *a*) revealed an unaltered main trunk of the portal vein. A pronounced blood overflow from the portal system to the IVC through the non-obliterated Arantius' duct, which entered the aneurysmal expansion from the IVC, was determined. The pathological duct had two branches. One of them was short (practically the portal vein entered the aneurysmal expansion), and the other branch was up to 1.5 cm long, approximately 1 cm in diameter. The right branch of the portal vein was well contrasted. The conclusion was made on the non-obliterated Arantius' duct.

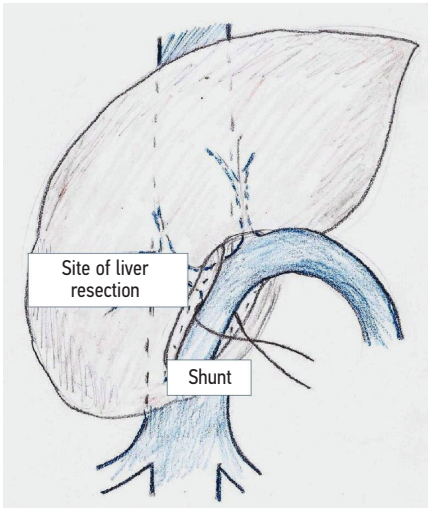


Fig. 6. Case No. 3. Scheme of surgical intervention
Рис. 6. Случай № 3. Схема оперативного вмешательства

The blood ammonia level was >400 μg/dL (norm maximum 110). Determining the exact indicator was not possible because of the diagnostic limit of the device. The level of bile acid was 155.2 (normal <10). Magnetic resonance imaging of the brain was performed to determine the degree of toxic encephalopathy, but no pathologies were noted.

To discuss the results of the examination and determine the further treatment approach, a case conference was conducted, with the participation of the employees of the Department of Endovascular Surgery. The conclusion was made on the non-closure of Arantius' duct. Given the anatomical aspects of the portal vein malformation, i.e., it entered directly the aneurysmal expansion, endovascular occlusion of pathological vessels was technically impossible. Taking into account the increased levels of ammonia in the blood and bile acids, low degree of encephalopathy, and ineffectiveness of conservative therapy, surgical treatment was indicated, which included mobilization, and ligation of Arantius' duct.

Reconstructive plastic surgery was performed on the vessels of the portal vein (Fig. 6).

Surgery. Laparotomy by a subcostal incision on the right was performed. The liver had normal size, color, and elastic

consistency, with multiple inclusions of the connective tissue and areas of induration. The portal vein, approximately 9 mm in diameter, was represented by one trunk up to the hilum. Directly in the parenchyma of the liver, a trunk up to 4 mm in diameter departed from it, toward the IVC, the portal vein passed into Arantius' duct with a diameter of approximately 8 mm, and was covered by the liver parenchyma approximately 2/3 of the circumference. An atypical resection of segment VII of the liver was performed, which enabled us to visualize most of Arantius' duct. With technical difficulties, mobilizing the duct along the circumference between the intraparenchymal trunk of the portal vein and the IVC was possible. Arantius' duct was placed on a tourniquet, and its lumen was occluded. The portal vein pressure was 28–30 cm WG. Visually, the venous outflow from the intestine was not impaired, and no changes in the blood supply to the liver were noted. The situation was discussed with the staff of the Department of Endovascular Surgery. Arantius' duct was ligated at the tourniquet site using silk 2/0. Liver biopsy was performed. The drainage was installed in the hepatic hilum and placed through a separate puncture in the anterior abdominal wall.

Pathological conclusions based on the liver biopsy revealed that the histological presentation corresponded to fibronodular hyperplasia.

In the early postoperative period, a low-velocity (<5 cm/s) blood flow was observed in the portal vein with the formation of parietal thrombi, which required heparin therapy. The latter contributed to the occurrence of bleeding into the abdominal cavity from the wound surface of the liver (biopsy site) in the postoperative period. Conservative therapy was ineffective; therefore, on postoperative day 3, relaparotomy was performed, parenchymal bleeding was stopped, and the intra-abdominal hematoma was evacuated. The further course of the postoperative period was uneventful. Owing to the use of heparin, the portal blood flow was completely restored (velocity 18 cm/s). The child was discharged for an outpatient follow-up.

The patient was repeatedly hospitalized in the RCCH in 1 year. Upon admission, the condition was satisfactory. US with duplex scanning of the portal system vessels revealed normal liver size, with smooth contours. The blood flowed antegradely through the portal vein, with a velocity of 18 cm/s. The second trunk of Arantius' duct with a diameter of 7–8 mm was identified.

Angiography (Fig. 5, *b*) showed an undeformed portal vein in a typical location. Intrahepatic branches of the portal vein were unaltered. The liver parenchyma was contrasted fully and evenly. Hepatic veins were patent and not deformed. The dilated, tortuous veins of the natural portoportal anastomosis into the distal branches of the portal vein were contrasted. Of the latter, single small fistulas in the IVC were noted. The conclusion was made on the non-obliterated Arantius' duct and the condition after its ligation.

Laboratory studies revealed the normalization of the blood levels of ammonia and bile acids. The child was discharged for outpatient management in satisfactory condition. The surgical treatment results can be regarded as excellent.

Case 4

The patient was an 11-year-old girl hospitalized in the RCCH.

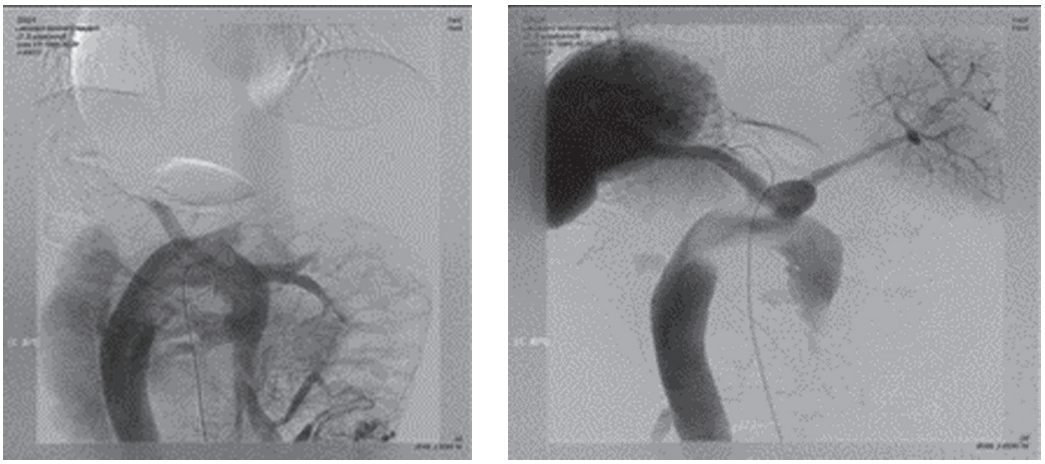
The anamnesis revealed that at the age of 3 years, she was examined for bleeding from varicose veins of the hemorrhoidal plexus at the primary healthcare facility, and surgical treatment was performed. On US, congenital portocaval shunt was suspected; therefore, an in-depth examination in a specialized clinic was recommended. Before the age of 11, the examination was not conducted. At this age, the patient was first hospitalized in the RCCH with a referring diagnosis of a suspected congenital portocaval shunt, with frequent nasal hemorrhage, and progressive mental retardation.

Examination. The blood ammonia level was 346 (normal, up to 110) μg/dL.

US with duplex scanning of the vessels of the abdominal cavity and retroperitoneal space turned out to be uninformative. The disease presentation was clarified by angiography (Fig. 7, *a*), which revealed an unaltered main trunk and intrahepatic branches of the portal vein. Severe hypoplasia was not detected. Pronounced retrograde blood flow was noted along the inferior mesenteric vein, which was sharply dilated, with the discharge of blood through the sacral plexus into the internal iliac vein. The pressure in the portal system at the closing of the overflow (compression) was 17.5 cm WG, and when removing the compression, it was 15.5 cm WG. Endovascular occlusion was technically impossible.

Based on the examination results, surgical treatment was decided.

Surgery. A lower median laparotomy was performed. During the revision, a dilated (diameter up to 25 mm) inferior mesenteric vein was noted, and the walls were thickened and sclerotic throughout. The liver size was slightly reduced, of common consistency, and color. The spleen had no abnormalities. The superior mesenteric vein had an elastic wall, dilated to 16 mm at the site of confluence with the inferior mesenteric vein. Moderate dilation of the peripheral veins ending in the superior mesenteric vein was observed. Angiomatous nodes were found in the appendix mesentery. Superficial angiomatosis was noted in the cecum head. The rest of the intestine had no abnormalities. The inferior mesenteric vein was mobilized and placed on rubber tourniquets 3 cm away from the confluence with the superior mesenteric vein and in the region of the transitional fold of the peritoneum. Intraoperative US of the blood flow in the superior mesenteric and portal veins was performed before and after clamping the inferior mesenteric vein



a *b*

Fig. 7. Case No. 4. Mesentericoportography upper: *a* — examination before surgery; *b* — control angiography. Condition after ligation of the pathological aorto iliac shunt. Portal blood flow is fully restored
Рис. 7. Случай № 4. Мезентерикопортография верхняя: *a* — до оперативного вмешательства; *b* — контрольная ангиография. Состояние после перевязки патологического портоподвздошного шунта. Портальный кровоток полностью восстановлен

with a mesenteric-caval shunt. Before clamping, the blood flow through the portal vein was scarcely recorded, and the blood flow velocity in the superior mesenteric vein was 7 cm/s. After clamping, the blood flow velocity in the superior mesenteric vein decreased to 5 cm/s, and the blood flow in the portal vein increased sharply to 15 cm/s. Following the clamping of the inferior mesenteric vein, no impairment in the venous outflow from the intestine was noted. Ligation and removal of the inferior mesenteric vein with a mesenteric-caval shunt were decided. The left internal iliac vein was separated, and its ligation was performed near the confluence of the shunt and at a distance of 0.6 cm from the confluence with the external iliac vein. The dysplastic inferior mesenteric vein was ligated, and 12 cm of its length was removed (Fig. 8). The venous outflow from the left half of the colon through the arcade veins was unobstructed. No blood supply impairment to the intestinal wall was noted. An appendectomy was performed with stump immersion. A safety drain was installed in the pelvic cavity. The postoperative period was uneventful.

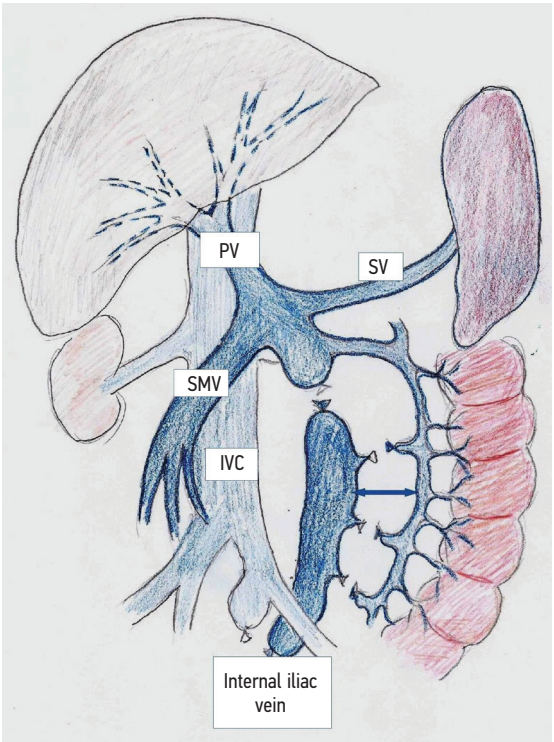


Fig. 8. Case No. 4. The scheme of surgical intervention. The arrow shows the resected part of the inferior mesenteric vein: SMV — superior mesenteric vein; PV — portal vein; IVC — inferior vena cava; SV — splenic vein
Рис. 8. Случай № 4. Схема операции. Стрелкой показана резецированная часть нижней брыжеечной вены: ВБВ — верхняя брыжеечная вена; ВВ — воротная вена; НПВ — нижняя полая вена; СВ — селезеночная вена

After 3 months, control angiography was performed (Fig. 7, *b*), revealing that the portal vein and its intrahepatic branches were unaltered. The superior mesenteric vein was not contrasted, with the outflow through the arcade collaterals. In the projection of the rectum, multiple phlebectases, and a part of the remaining portoiliac shunt were contrasted; however, the contrasting of the iliac veins was not pronounced. The stump of the inferior mesenteric vein was determined. The outflow from the rectum and sigmoid colon was through the arcade vein. The blood ammonia level was 98 mg/mL.

Thus, in this case, good results were achieved, and the pathological portoiliac shunt (i.e., flow from the inferior mesenteric vein into the internal iliac vein through the sacral plexus) was eliminated. Intestinal bleeding after surgery did not recur.

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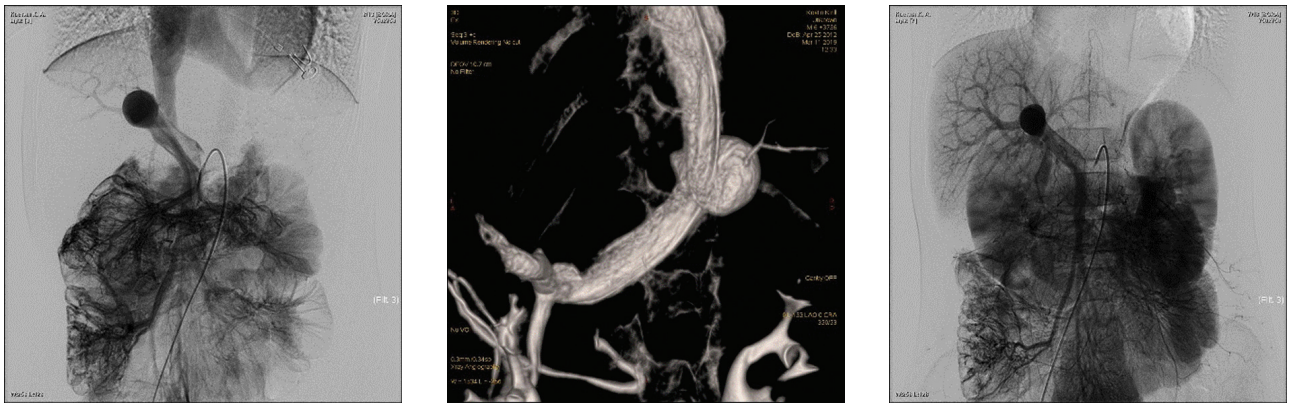


a *b* *c*

Fig. 9. Case No. 5. Results of angiographic studies of patient: *a* — upper mesentericoportography. Examination before surgery; *b* — rotational transjugular phlebography of the Arantian duct with 3D reconstruction; *c* — mesentericoportography upper. Examination after surgery. Portal blood flow is fully restored

Рис. 9. Случай № 5. Результаты ангиографических исследований пациента: *a* — мезентерикопортография верхняя. Исследование до оперативного вмешательства; *b* — ротационная трансюгулярная флебография аранциева протока с 3D-реконструкцией; *c* — мезентерикопортография верхняя. Исследование после оперативного вмешательства. Портальный кровоток полностью восстановлен

Case 5

The patient was a 7-year-old boy hospitalized in the RCCH.

The anamnesis revealed that from birth, the patient was monitored, and treated at the primary healthcare facility for pronounced hepato-pulmonary syndrome and pulmonary hypertension. He was referred to the Dmitry Rogachev National Medical Research Center for Pediatric Hematology, Oncology, and Immunology because a shunt was found between the portal vein and IVC. During fiber-optic esophagogastroduodenoscopy (FEGDS), Grade 0–I esophageal varicose veins were noted, and perfusion scintillation lung imaging revealed signs of functioning of the pulmonary shunts and discharge from the pulmonary circulation to the systemic circulatory system of 50%. The child was transferred to the RCCH.

On admission, pronounced signs of respiratory failure were noted, including severe fatigue, shortness of breath, cyanosis with minor physical exertion, and clubbed fingers. SpO_2 values without oxygen supply did not exceed 85%.

Examination. US with duplex scanning of the portal system vessels revealed that the liver size was normal, and its contours were smooth. The parenchyma was granular and somewhat cord-like. Echogenicity was moderately increased. The walls of the intrahepatic bile ducts were unevenly indurated. The lumen closer to the hilum was differentiated. The lobar branches of the portal vein were dilated (right 11 mm, left 17 mm). The gallbladder, pancreas, and spleen had no abnormalities. The diagnosis was echo-signs of diffuse changes in the liver parenchyma and abnormal development of the portal vein system. The blood ammonia level was 186 μg/dL.

Angiography (Fig. 9, *a, b*) revealed that the portal vein represented by one trunk passing directly into the IVC and having an aneurysmal expansion, from which intrahepatic branches of small diameter depart. The diagnosis was CPSS.

Because of severe respiratory failure, the child received hyperbaric oxygen therapy. A case conference was conducted, with the participation of the staff of the Department of Endovascular Surgery. They emphasized the technical impossibility of endovascular occlusion of a portosystemic shunt because the portal vein entered directly the IVC in the area of aneurysmal expansion. Surgical treatment with open ligation of the shunt was indicated.

Surgery. A median laparotomy was performed. The liver was slightly reduced, brick-colored, and flaccid when touched. The stomach and intestinal loops were bluish. The spleen was not enlarged. Aneurysmal dilatation of the portal vein was revealed in the liver hilum, extending into the parenchyma along the right hepatic duct. Isolation of the portocaval anastomosis from the hepatic hilum was not possible. The right liver lobe was separated from the diaphragm. The IVC was placed on a rubber tourniquet over the hepatic veins. With the further mobilization of segment VII along the IVC, a portocaval fistula was detected at the level of the right adrenal gland. The fistula with a diameter of approximately 14 mm in the narrowest part was surrounded by the liver parenchyma by 2/3 of the circumference. The bridge of the parenchyma above the anastomosis was transected. A dissector was used to form a tunnel around the anastomosis. The portocaval duct was ligated (Ethibond 0) directly above the IVC without deformation of the vessels (Fig. 10).

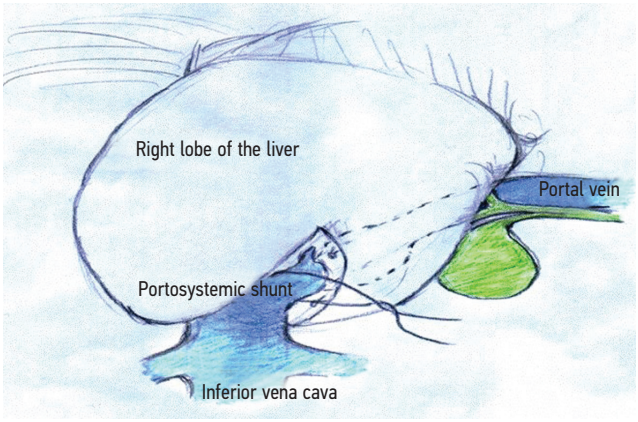


Fig. 10. Case No. 5. Scheme of surgical intervention
Рис. 10. Случай № 5. Схема оперативного вмешательства

The tourniquet was removed from the IVC. Moreover, the pressure in the portal vein did not change significantly, and the liver, stomach, and intestinal loops became pink. No changes in the spleen were noted.

On postoperative days 5–6, a certain improvement in the lungs was noted, i.e., the respiratory failure significantly improved. By day 8, the SpO_2 indicator was 96%–98% without oxygen supply. Thus, the relief of the hepatopulmonary syndrome occurred on days 5–6 after the ligation of the pathological shunt. The postoperative course was uneventful.

According to the US with duplex scanning of the portal system vessels on postoperative day 8, the liver had normal size. The blood flowed antegradely through the portal vein and its branches, at a velocity of 14 cm/s, and the lumen of the vessels was clear. The blood flow velocity in the splenic vein was 11 cm/s. The lumen of the IVC was clear, and the blood flow in the suprarenal section was unaltered. The child was discharged with a significant improvement in his condition for outpatient follow-up 11 days after the surgery.

After 8 months for a control examination, the patient was repeatedly hospitalized in the RCCH. No complaints on admission were noted, and the condition was satisfactory. No signs of respiratory failure were observed.

US with duplex scanning of the portal system vessels revealed an enlarged liver due to the left lobe. The anteroposterior sizes of the right and left lobes were 107 and 41 mm, respectively. The contours were smooth. The parenchyma had indurated walls of vessels and ducts. No abnormal echogenicities were recorded. Intrahepatic bile ducts were not dilated. The portal vein was not clearly differentiated; in the hilum view, there was a conglomerate of vessels 3.5 × 2.5 cm with multidirectional blood flows. The splenic vein was 8 mm in diameter, and the blood flow velocity was 21 cm/s.

Angiography (Fig. 9, *c*) revealed the absence of a portocaval fistula. The contrasting of the portal vein branches was satisfactory. The child was discharged for outpatient management in satisfactory condition.

There were no complaints at the control examination after 1 year.

US with duplex scanning of the portal system vessels showed an enlarged liver due to the left lobe. The anteroposterior sizes of the right and left lobes were 117 and 52 mm, respectively. The contours were smooth. The parenchyma was fine and granular. The walls of the intrahepatic bile ducts were fragmentarily indurated, and the lumen of the ducts was not expanded. The blood flowed antegradely through the portal vein and its branches at a velocity of 10–13 cm/s. The diameter of the IVC was 6–8 mm.

Angiography revealed the absence of a portocaval fistula. The contrasting of the portal vein branches was satisfactory. FEGDS showed no varicose veins of the esophagus and stomach. A polyp of the cardia was detected, which was removed endoscopically. The blood ammonia level was 56.5 μmol/L (normal). The child was discharged in a satisfactory condition. Thus, the result of the surgical treatment of a CPSS can be regarded as excellent.

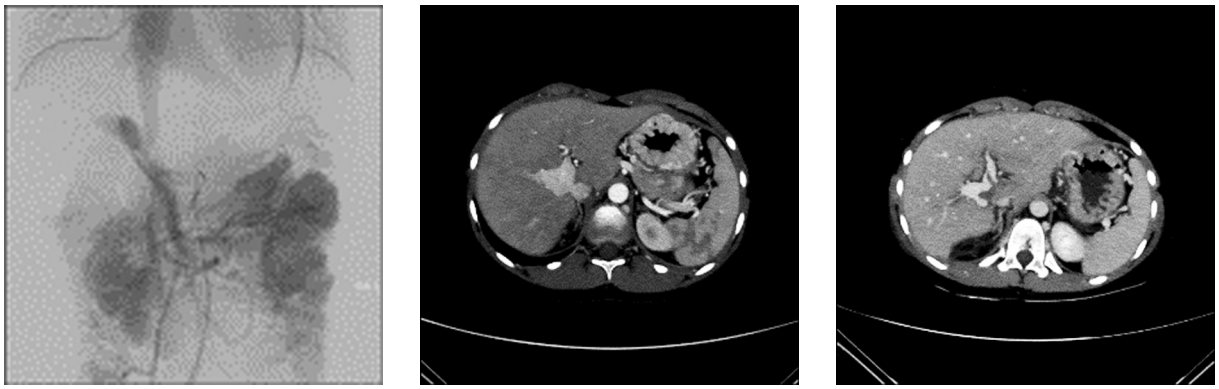
Case 6

The patient was a 12-year-old boy at the time of hospitalization in the RCCH.

The anamnesis revealed that 3 years before hospitalization in the RCCH, the patient received inpatient treatment at the primary healthcare facility (Surgut) for duodenal ulcers. Moreover, pains in the joints appeared, and a diagnosis of juvenile rheumatoid arthritis was made; therefore, he received methotrexate. The joint pain diminished; however, the child complained constantly of asthenia and increased fatigue (as it turned out later, due to hepatopulmonary syndrome). He was hospitalized in the Department of Clinical Immunology of the RCCH, where a portosystemic shunt was detected during the examination; therefore, the child was transferred to the Department of Microvascular Surgery No. 2.

Examination. US with duplex scanning of the vessels of the abdominal cavity and retroperitoneal space revealed that the liver was not enlarged and had smooth contours. The parenchyma was homogeneous, and the walls of the vessels were indurated. Echogenicity was unaltered. The main trunk of the portal vein was dilated, measuring 15 mm. Arantius' duct between the portal vein and the IVC had a diameter of 7 mm. The venous outflow was not impaired. The diameter of the splenic vein was 6 mm, with antegrade blood flow and velocity of 23 cm/s. The blood ammonia level was 214.3 (norm maximum 110) μg/dL.

Computed tomography revealed asymmetric liver at the level of the right lobe, hypertrophy of II, III, IVa, and IVb segments, non-dilated bile ducts, and thickened walls of the common bile duct, with ectasia up to 3.5–4 mm. The parenchymal density was not reduced (60 HU), the parenchyma was granular, and periportal fibrosis was noted. An ectasia (diameter 21 mm) of the portal vein was revealed in the area of the outflow; below the ectasic area,



a *b* *c*

Fig. 11. Case No. 6. The results of X-ray examination of patient: *a* — angiography. Examination before surgery. A congenital portosystemic shunt is visualized. There is marked hypoplasia of the intrahepatic branches of the portal vein; *b* — computed tomography. Examination before surgery. The portocaval junction is visualized; *c* — computed tomography after surgery. Significant improvement in hepatic blood flow. There are no signs of a functioning congenital portosystemic shunt
Рис. 11. Случай № 6. Результаты рентгенологических исследований пациента: *a* — ангиография. Исследование до оперативного вмешательства. Визуализируется врожденный портосистемный шунт. Отмечается выраженная гипоплазия внутрипеченочных ветвей воротной вены; *b* — компьютерная томография. Исследование до оперативного вмешательства. Визуализируется портокавальное соустье; *c* — компьютерная томография после оперативного вмешательства. Значительное улучшение печеночного кровотока. Признаки функционирования врожденного порто-системного шунта отсутствуют

there was an isthmus (anatomical variant? transformation?), and the diameters of the portal vein was 8 and 12.7 mm at the level of the confluence of the splenic vein. The IVC at the renal level had a diameter of 21.6 mm. The mesenteric veins were dilated. Arantius' duct was functioning. The azygos and hemiazygos veins were dilated. In the arterial phase, the contrasting of the liver was uneven, there were zones of hyper- and hypoperfusion with clearly defined nodule-like areas or formations up to 21 mm in diameter, with multiple ectatic collaterals of the portal vein and arterial vascular systems inside them, obviously with arteriovenous discharge against the pressure gradient. In the parenchymal phase, the densities were aligned. This phenomenon may indicate (a) changes associated with portal hypertension with a mixed type of blood flow and (b) morphological and regenerative changes in the tissue of the liver parenchyma, including their outcome. A neoplastic process was less probable.

Angiography (Fig. 11) showed that the main trunk of the portal vein was of normal size and typically located. The superior mesenteric and splenic veins had no abnormalities. The portal vein confluence with an aneurysmally altered bifurcation area, from which the IVC was immediately contrasted. There was marked hypoplasia of the intrahepatic branches of the portal vein, extending from the aneurysmal expansion.

The conclusion was CPSS; however. endovascular occlusion was technically impossible because of the direct confluence of the portal vein into the IVC in the area of aneurysmal expansion.

The pathological portocaval shunt was ligated, almost similar to that performed in case 5 (Fig. 12).

Surgery. Upper transverse laparotomy was performed. The size, texture, and color of the liver were normal. The hepatoduodenal ligament was isolated. At the liver hilum, the hepatic artery was isolated and retracted to the right. Above the pancreatic head, an ectatic portal vein that was traced to the division into lobar branches was found. Moreover, a pathological anastomosis between the portal vein and the IVC was not detected. The right liver lobe was mobilized. The IVC was mobilized from the renal veins to a branch originating from the right liver lobe. This branch was

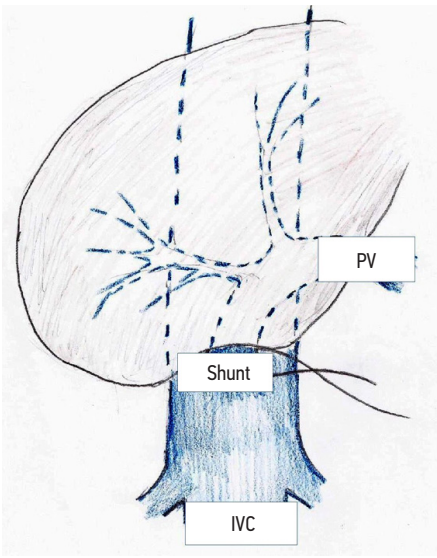


Fig. 12. Case No. 6. Scheme of surgical intervention. PV — portal vein; IVC — inferior vena cava
Рис. 12. Случай № 6. Схема оперативного вмешательства. ВВ — воротная вена; НПВ — нижняя полая вена

considerably dilated and entered the IVC at an obtuse angle. Above it, the branches of the right hepatic vein flow into the IVC. Upon further isolation of this vessel, it was found to be a pathological portocaval shunt, and the presentation turned out to be almost similar to that in case 5. The pathological vessel was ligated above the place of its confluence with the IVC.

During the first 2 days of the postoperative period, moderate respiratory failure (consequences of the hepatopulmonary syndrome) was noted, which was treated on day 3. There were no other complications.

Computed tomography was performed on postoperative day 2. In the area of confluence of the portal vein, a significant contrasting defect was detected, which had an irregular saddle embolism shape extending into the left branch of the portal vein (fragment 10 × 5 mm) and into the locally ectasic right branch of the portal vein in a fragment 16 × 9 mm (obviously, thrombus). The IVC lumen at this level was narrowed; a convoluted vessel of small caliber (up to 2 mm) departed from the right side wall, connecting with the right branch of the portal vein, with its trace contrast (ligated shunt). In the upper parts of the right liver lobe, a subcapsularly located focus of increased perfusion was noted. The hepatic artery was passable, with a diameter of up to 7.5 mm. In the arterial phase, contrast enhancement of the liver was heterogeneous. In the portal phase, good perfusion was noted. The pronounced ectasia of the lower posterior pulmonary veins, dilated to the peripheral sections (obviously, manifestations of the hepatopulmonary syndrome), was notable.

On postoperative day 9, US with duplex scanning was performed. The liver measured 121 × 43 mm. The blood flowed antegradely along the main trunk of the portal vein. No hemodynamic signs of the relationship between the portal vein and the IVC were noted. The lumen of the IVC was clean. The spleen was not enlarged. The blood ammonia level was 106.2 μg/dL. The child was discharged in a satisfactory condition.

DISCUSSION

CPSS is a rare developmental anomaly when the blood outflow from the abdominal organs bypasses the portal system of the liver immediately into the venous circulation of the systemic circulation. As a result, unpurified blood coming from the intestines causes the resulting clinical manifestations. CPSS occurs in utero due to an impairment of the embryological development of the portal vein system. For the first time, a portosystemic shunt was described in the medical literature by the London surgeon John Abernethy in 1793. During the autopsy of a 10-month-old child with multiple malformations, he found a direct flow of the portal vein into the IVC at the level of the renal veins [6]. In more recent literature, some authors have referred to CPSS with no portal blood flow as the “Abernethy malformation.”

Publications in the 1950–1980s reported isolated cases and described the treatment of patients with portosystemic shunts. Since the 1970s, the number of publications describing surgical ligation of portosystemic shunts in adults and children and endovascular occlusion and laparoscopic ligation since the 1990s has increased. In the last two decades, articles describing 15–20 patients have been published, and the maximum number of patients with CPSS in the largest single-center study was 40 [7]. By 2022, information about approximately 700 patients with CPSS has been found in the world literature. The literature describes many options for the use of portosystemic shunts. In 1994, Morgan and Superina were the first to propose a classification based on the complete absence (type I) or presence (type II) of the intrahepatic portal system [8]. In 2008, Stringer proposed dividing all portosystemic shunts into intrahepatic and extrahepatic [9].

In 2011, Lautz et al. [10] proposed their classification: in type I CPSS, there is no intrahepatic portal blood flow. Type II CPSS is divided into three subtypes, where IIa has a shunt corresponding to the venous duct, IIb has a shunt arising from the main trunk of the portal vein at the splenomesenteric fusion with a portal bifurcation, and IIc has a shunt arising from the mesenteric, gastric, or splenic veins. Patients with Lautz type 1 CPSS are considered candidates for liver transplantation [10]. New classifications continue to emerge. Blanc et al. [11] presented a surgical classification based on the variants of fusion of the portal system and the vena cava system with the aspects of the surgical approach for these shunts. Later, Kanazawa et al. [7] proposed supplementing this classification with an angiographic one according to the severity of the hypoplasia of the intrahepatic portal veins and conducting a balloon-occlusion test to identify patients at high risk in the postoperative period. To cure such patients, separating the portal, and systemic circulations is quite enough.

Despite equal efficiency between X-ray endovascular and open surgical treatment, the X-ray endovascular method is much more sparing, causes no complications associated with laparotomy, and offers access to the shunt, with a more uneventful postoperative period and significantly reduced treatment period, which is especially important in younger patients [12]. Unfortunately, endovascular occlusion was not possible in some anatomical variants of CPSS. In this case, open surgical ligation of the CPSS is required [13, 14].

In this series of cases, the use of endovascular occlusion was impossible, and there were indications for surgical intervention. Similar indications have already been previously described in the literature: (a) ineffectiveness of endovascular closure (relapse) and (b) anatomical aspects of the shunt [15]. In addition, The Abernethy malformation

has variants, i.e., when both endovascular occlusion, and ligation of the vicious shunt are not possible, due to a high probability of a completely impaired outflow from the portal vein system due to thrombosis. The critical indicator of pressure in the portal vein system during an occlusive test is 30 cm WG [16].

In the early postoperative period, constant monitoring (US) of the portal blood flow is necessary because, with a decrease in the rate of the latter, the probability of thrombogenesis is high, which requires anticoagulant therapy. Given this, careful hemostasis is necessary (primarily in the area of wound surfaces on the liver after taking a biopsy sample) during the surgery.

CONCLUSION

Congenital portocaval shunts are rare and may have different morphological structures (intra- and extrahepatic shunts, with, or without portal blood flow). Treatment is possible only in cases of preserved portal perfusion of the liver. It is usually possible to achieve good results after endovascular occlusion of a pathological shunt. Cases in which endovascular occlusion is technically impossible require surgical treatment. Each such case is unique, and each surgery is non-standard. The surgeon determines and changes the approach directly during the intervention, depending on the characteristics of the morphological structure of the organs, because even a careful preoperative examination does not always provide an unambiguous idea of this.

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