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# Cruvelier-Baumgarten Syndrome Type II in Combination with Arteriovenous Splenic Artery Fistula: Literature Review and Clinical Case

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**ABSTRACT**

Cruvelier-Baumgarten syndrome is a rare disease that causes portal hypertension in association with unclogged umbilical or paraumbilical veins, hypoplasia of the intrahepatic portal system, and liver atrophy with minimal or no fibrosis. Patients with this condition experience the classic signs of portal hypertension such as gastrointestinal bleeding, ascites, and encephalopathy. Frequent clinical manifestations are the symptom of "jellyfish head" and venous murmur over the umbilical region due to the presence of large umbilical and paraumbilical veins.

In the presented clinical case of Cruvelier-Baumgarten syndrome type II in combination with splenic artery arteriovenous fistula, pathological changes were not pronounced. There was no "jellyfish head" symptom, but a venous murmur was heard in the umbilical region. The patient complained of moderate pain in the epigastrium, right hypochondrium. Examination of the patient, which revealed, in addition to the typical manifestations of this disease, also multiple SAVFs, as well as angiography data regarding pressure in the umbilical vein - 5 mm Hg, determined the tactics of treatment in the volume of endovascular occlusion of the splenic artery branches, which was successfully completed. As a result, there was a decrease in pressure in the portal system and a sharp decrease in discharge through the recanalized umbilical vein into the basin of the inferior vena cava, which made it possible to level out complaints. At the control examination two months after treatment, the patient had no complaints.

A review of the literature on the Cruveilhier-Baumgarten syndrome and disease is presented.

**KEYWORDS:** Cruveilhe-Baumgarten syndrome type II, recanalization of the umbilical vein, portal hypertension, splenic artery aneurysm, splenic arteriovenous fistula, diagnosis, treatment.

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## INTRODUCTION

N. Pegot in 1833, addressing the members of the Parisian Anatomical Society, described the case of an alcoholic soldier patient with auscultated subcutaneous venous murmur in the paraumbilical region without a visible substrate [1]. In 1835 Jean Cruveilhier described a similar case. At autopsy, it was found that the venous murmur was caused by collateral circulation through the widely passable umbilical vein, and the liver was hypoplastic [2]. In 1907, Paul Clemens von Baumgarten reported a similar case in a sixteen-year-old boy who died from gastric bleeding. He noted the nosological independence of the disease and believed that the widely patent umbilical vein, splenomegaly, and liver atrophy were associated with hypoplasia of the portal vein system [3]. In the future, cases of the liver cirrhosis with this anomaly became known. The name "cirrhosis of Cruveilhier—v. Baumgarten" was offered in 1922 by M. Hanganutz, who considered this syndrome to be a separate clinical and pathological unit. He collected six cases, each of which was a slight cirrhosis of the liver, portal hypertension, venous anastomosis in the falciform ligament and abdominal wall connecting the portal and epigastric veins, and a noise or hum auscultated over the epigastrium and the lower end of the sternum [Hanganutz M. Pr. Med. 1922; 30:732] (adapted from Macpherson A. and Morton E. [4]).

The first complete review in the English-language literature was published by E.L. Armstrong et al. in 1942 [5], who coined the terms Cruvelier-Baumgarten syndrome and Cruvelier-Baumgarten disease. E.L. Armstrong et al. analyzed the cases published at that time and divided them into two groups, later this division was somewhat modified by J.S. Steinburg and J.T. Galambos [6]:

• Cruveilier-Baumgarten syndrome (or Type I syndrome): Liver cirrhosis or portal hypertension causes paraumbilical vein dilatation (that is an acquired condition in which the veins reopen due to high portal pressure).

• Cruveiler-Baumgarten disease (or type II syndrome): paraumbilical vein enlargement occurs due to patency of the umbilical vein, with little or no evidence of umbilical vein disease on liver biopsy that is a congenital condition in which umbilical vein failure leads to portal hypertension).

**Cruvelier-Baumgarten syndrome (or type I syndrome).** This syndrome is a rare complication of liver cirrhosis. It is characterized by the presence of cirrhosis of the liver or portal hypertension, leading to extensive dilatation of the paraumbilical veins and recanalization of the umbilical vein, the development of portosystemic collaterals between the umbilical, paraumbilical veins of the anterior abdominal wall and the epigastric veins flowing into the external iliac veins. Thus, the primary cause of the syndrome is not the absence of obliteration of the umbilical vein, but an increase in pressure in the portal vein. Secondary is the expansion of the residual trunk of the umbilical vein and other paraumbilical veins as a result of portal hypertension. In this regard, the entire vascular site with its collaterals is used as a path for blood outflow through the portal vein [5–9]. Characteristic symptoms of Cruvelier-Baumgarten syndrome:

1. Increased pressure in the portal vein (with ascites, liver enlargement, edema), which is based on cirrhosis of the liver of various origins.
2. The development of a pronounced venous network on the anterior abdominal wall ("jellyfish head") with auscultatory detected vascular noises in the umbilical region.
3. With the progression of the disease, secondary symptoms develop, depending on liver cirrhosis.

**Cruvelier-Baumgarten disease (or type II syndrome).** Malformation of the umbilical vein, which is not obliterated. The clinical picture is the result of congenital umbilical vein patency with minor damage to the liver or portal tract [5–9]. Typical symptoms of Cruvelier-Baumgarten disease:

1. Congenital disease, the clinical symptoms of which gradually appear in adolescence.
2. Preserved umbilical vein, hypoplasia of the portal vein system. The non-obliterated umbilical vein anastomoses with the portal vein system. In this regard, visible and palpable venous ectasias develop on the skin of the abdomen ("jellyfish head") with a clearly detectable venous murmur in the navel.
3. Splenomegaly with splenogenic delay in bone marrow activity.
4. Cirrhosis of the liver may develop, but it does not belong to the typical clinical manifestations of the disease and in each case is a secondary late manifestation.

According to the literature, splenic artery aneurysms (SAA) account for 60–80% of aneurysms of the visceral arteries [10]. Approximately 80% of SAAs are asymptomatic and detected incidentally on imaging. SAA causing portal hypertension is rare. This is due to an aneurysmal mass effect compressing the splenic vein, causing congestion and thrombosis. Only a few cases have been described in the literature. There are reports of cases of extrahepatic portal hypertension caused by a single SAA [11, 12], slightly more reports of multiple SAA s causing extrahepatic portal hypertension [13-15]. In one of the cases of a large single aneurysm of the splenic artery, which caused portal hypertension, the presence of an aneurysm of the tortuous and dilated portal vein was also noted. The authors suggested that the development of portal vein aneurysm could be due to the progression of portal hypertension [12]. We encountered only one report of multiple aneurysms of the splenic artery in association with Cruveilhe-Baumgarten syndrome in a 24-year-old married woman who was admitted to the intensive care unit with a history of acute pain in the lower abdomen after eating [16].

Arteriovenous fistula of the splenic artery is a rare pathological condition, first described in 1886 by the German pathologist S. Weigert [17]. In 1999 C.F. Hung et al. reported 42 cases of this disease at that time [18]. In 2011, W. Woźniak et al. registered about 100 cases of splenic arteriovenous fistula (SAVF) [19].

The causes of arteriovenous fistulas in this localization are blunt abdominal trauma, rupture of a pre-existing aneurysm of the splenic artery, technical errors during splenectomy, and gunshot wounds [18, 20]. The rarity of SAVF is explained by the fact that injuries to these vessels lead to severe, often fatal bleeding [21].

With a small volume of arteriovenous shunt, SAVF are not accompanied by symptoms for a long time. With a larger volume of discharge and / or prolonged existence of the fistula, portal hypertension develops, which is clinically manifested by pain in the upper abdomen (70%), splenomegaly (55%), varicose veins of the esophagus or stomach (52%), ascites (35%) [18, 20].

The presence of SAVF, regardless of the severity of clinical symptoms, is an absolute indication for its elimination. The most common treatment is ligation of the fistula-bearing splenic vessels and splenectomy [22-24]. At the same time, surgical intervention, as a rule, is associated with technical difficulties due to a pronounced adhesive process and a large number of dilated veins, which significantly increases the likelihood of aneurysm rupture

during surgery. Endovascular intervention is an alternative and promising low-traumatic method of treatment that can be used both independently and as the first stage preceding splenectomy [24, 25].

We have not found a description of Cruvelier–Baumgarten syndrome and SAVFs combination in the literature.

We present a ***clinical case of Cruveilier-Baumgarten*** **type II syndrome in combination with SAVF.**

A 45-year-old woman was admitted to A.V. Vishnevsky National Medical Research Center of Surgery with complaints of moderate pain in the epigastrium, right hypochondrium

*Anamnesis.* Observed over the past eight years about portal hypertension and hypertension in the basin of the splenic vein of unclear etiology. During this time, she was repeatedly consulted by hepatologists. No changes in the liver were revealed according to radiology; histology of the liver tissue also showed no pathological changes. Examination at Hematology Research Institute revealed no pathological changes in the circulatory system. Gastroscopy revealed varicose veins of the stomach II degree. Severe portal hypertension, venous collaterals up to the saphenous veins with the inferior vena cava, hypertension in the basin of the inferior vena cava according to abdominal organs MDCT. No visible block data received. Signs of an arteriovenous fistula in the region of spleen hilum were revealed. The patient was hospitalized at A.V. Vishnevsky National Medical Research Center of Surgery for surgical treatment.

The general condition is satisfactory. Body temperature 36.3°C. There are no peripheral edema. Breathing is vesicular, carried out to all parts of the lungs, no wheezing is heard. Respiratory rate - 18 per min. Blood pressure - 130/80 mm Hg.

*Status localis:* Tongue moist, clean. The abdomen is not swollen, symmetrical, no visible venous collaterals are visualized (Fig. 1), the abdomen is involved in the act of breathing, soft on palpation, moderately painful in the upper sections, more on the right. On palpation of the abdomen, enlarged venous collaterals are palpated. A sharp venous murmur is auscult in the umbilical region. There are no peritoneal symptoms. Peristalsis is auscult. Tumor lesions are not palpable. Physiological functions are normal.

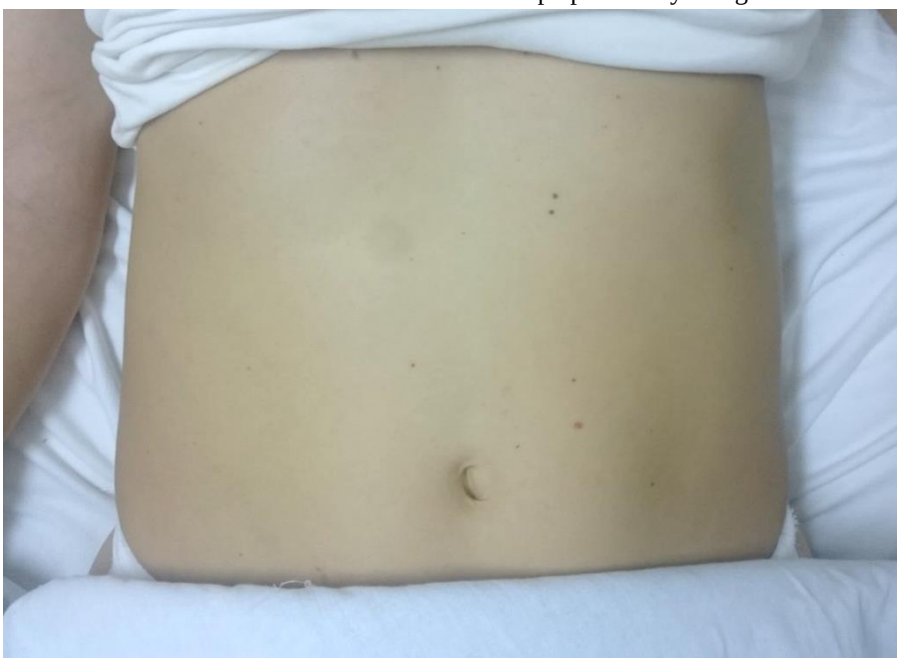


**Figure 1. Appearance of the patient**

*bdominal MDCT.* The size of the liver is not enlarged. Hepatic veins are defined with a diameter of up to 10 mm. The trunk of the portal vein was expanded to 21 mm, the splenic vein was expanded to 22 mm, and the superior mesenteric vein was 10 mm. A collateral vein with a diameter of up to 19 mm (recanalized umbilical vein) departs from the trunk of the portal vein, which passes along the dorsal surface of the anterior abdominal wall, forming many bends, and flows into the left iliac vein. The lumen of the left iliac vein above the confluence of the collateral was expanded to 32 mm. The inferior vena cava is dilated, the transverse dimensions of the lumen are up to 32x38 mm. The left renal vein was dilated up to 13 mm, the right one up to 10 mm (Fig. 2a, b).

The spleen is enlarged in size - 71x107x171 mm. The splenic artery is tortuous, expanded to 9 mm. In the region of spleen gate an aneurysmal expansion of the splenic artery up to 19 mm, for 22 mm is visualized. Small aneurysms, 5-7 mm in diameter, are visualized on the forks of small arterial branches in spleen parenchyma. Multiple dilated collateral veins are visualized in spleen hilum (Fig. 2c, d). An intensely contrasted zone 8x9 mm in size, 24 mm long, communicating

with the lumen of splenic vein collateral, is visualized in the lower pole of the spleen in arterial phase. Early contrasting of the collateral splenic vein is determined (up to 125 HU in the arterial phase).

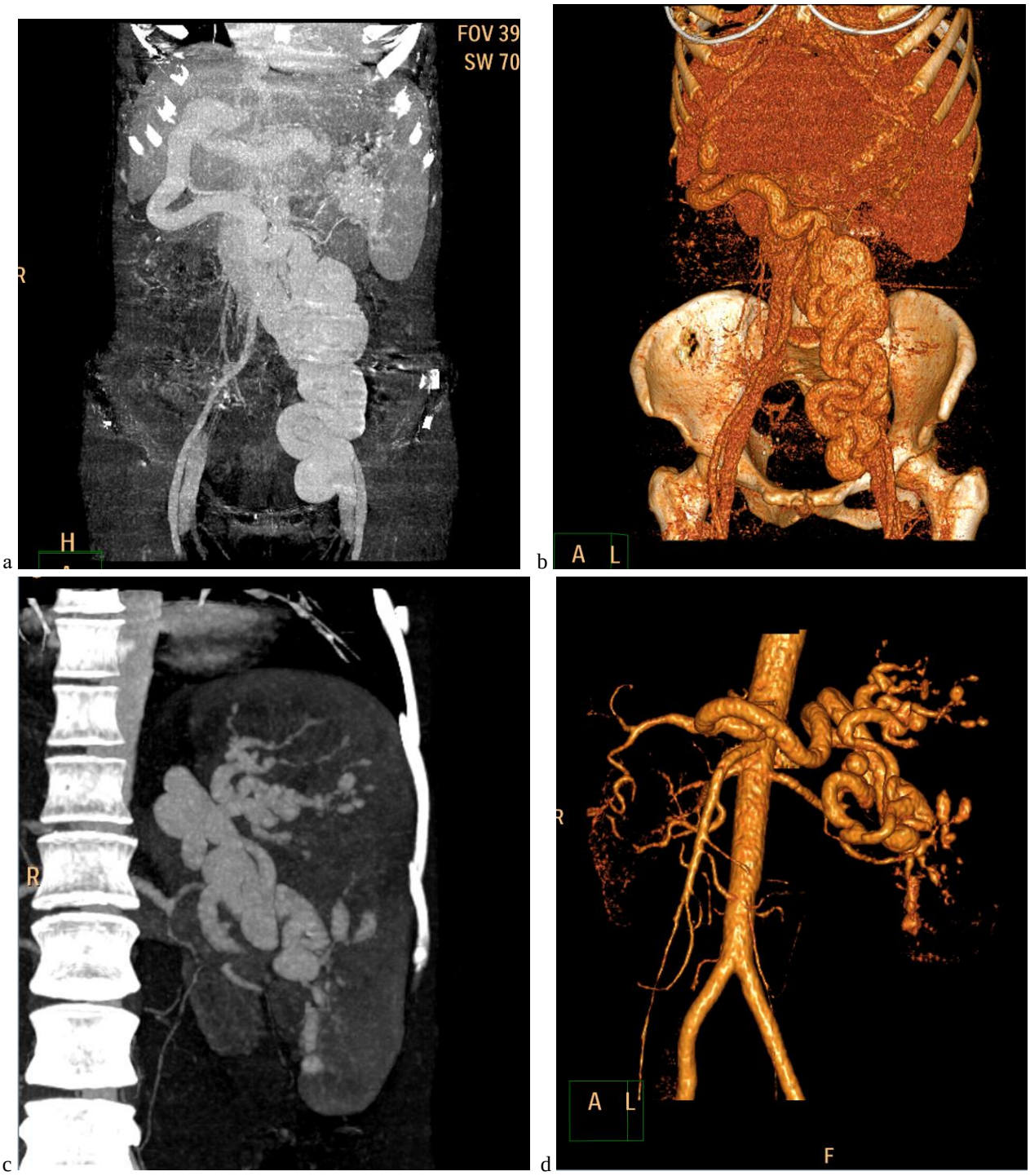



**Figure 2. Contrast enhanced MDCT: a - volume MIP; b – 3D; c - volume MIP; d - 3D of abdominal aorta and visceral brunches**

The diameter of truncus celiacus was expanded to 11 mm, common hepatic artery - to 7.5 mm.

*Gastroscopy.* Varicose veins of the stomach of the 2nd degree.

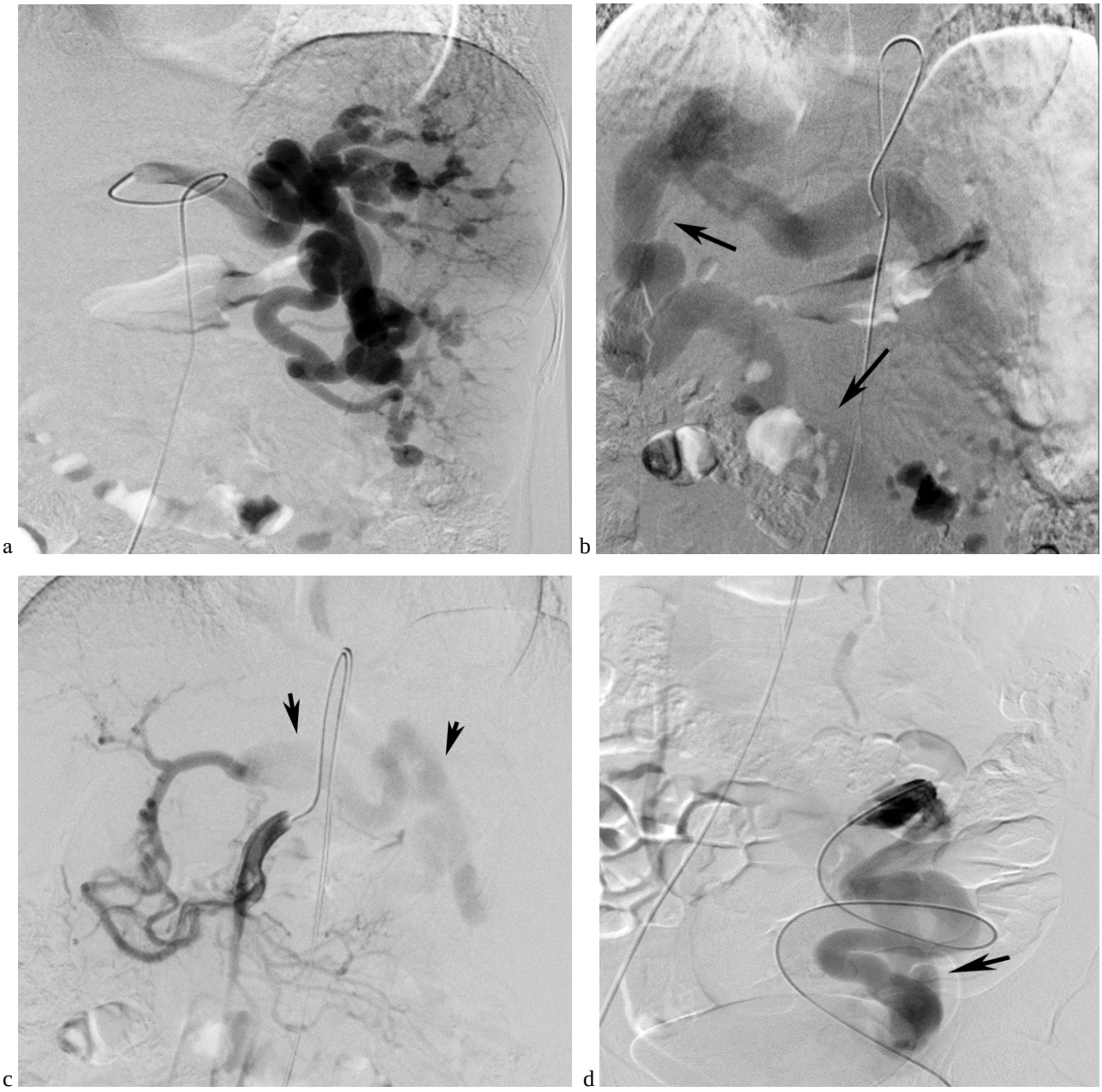
According to the preoperative examination, the diagnosis was made: Cruvelier-Baumgarten syndrome type II: recanalization of the umbilical vein, dilatation of the portal vein, splenomegaly. Multiple splenic artery aneurysms. Splenic artery. Splenic arteriovenous fistula. Varicose veins Of the stomach of the 2nd degree. Preoperative conclusion made a decision to perform endovascular treatment of splenic arteriovenous fistula.

*Angiography celiacography, mesentericography, return portography, endovascular occlusion of the lower splenic artery.*

Under combined anesthesia, the right common femoral artery was punctured and catheterized, a 5F introducer was placed, a Simmons-2 5F catheter was inserted through it, which was

alternately inserted into the truncus celiacus and the superior mesenteric artery, and angiography was performed.

Angiograms (Fig.3). With celiacography: the celiac trunk and its branches have even contours, the contrast is homogeneous. The common hepatic artery is not contrasted due to the pronounced blood flow in the splenic artery. The splenic artery has even contours, extremely tortuous, small aneurysms with a maximum diameter of 7 mm are visualized on the forks of small branches in the parenchyma of the spleen. The diameter of the splenic artery is up to 7 mm. There is an accelerated onset of the venous phase (at 7-8 sec), which is a sign of intraparenchymal microfistulas of the splenic artery. With return portography, contrasting of the dilated splenic vein and branches of the portal vein is noted. Further discharge of contrast from the right branch of the portal vein occurs into the umbilical vein, followed by evacuation of the contrast through the left inferior epigastric vein into the basin of the left femoral vein. With upper mesentericography, the pool of the common hepatic artery is contrasted and the contrast is partially discharged into the splenic artery. The diameter of the common hepatic artery is 4 mm.



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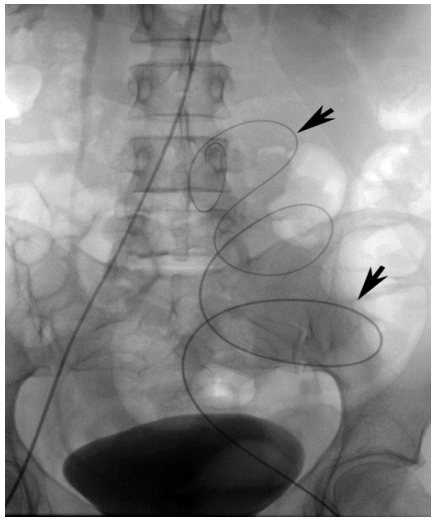


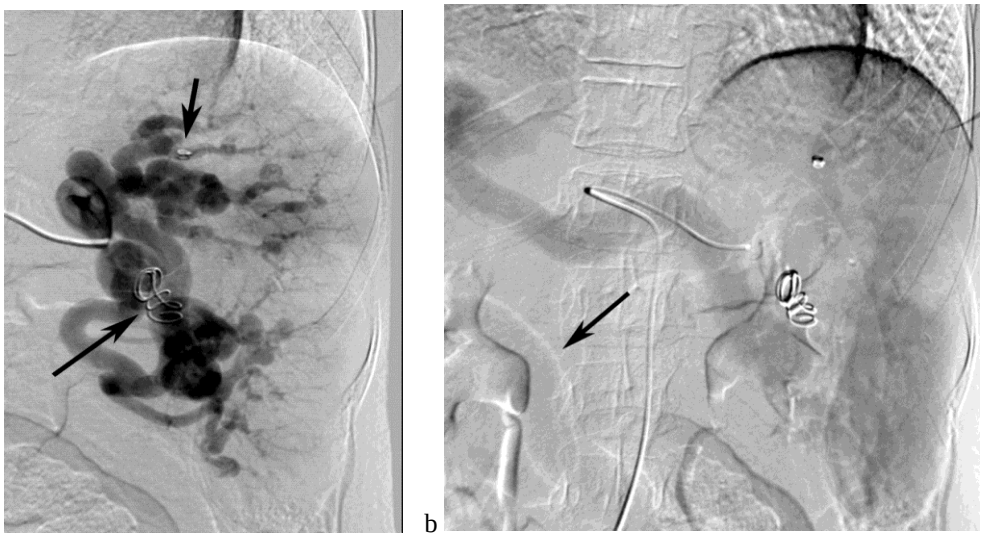
e

**Figire 3. Angiographic images**

a - celiacography. There is no contrasting of the common hepatic artery, multiple intraparenchymal aneurysms of the spleen. b - return portography. The umbilical vein is marked with an arrow. c - upper mesentericography. The basin of the common hepatic artery and the splenic artery are contrasted (marked by arrows). d - phlebography of the left epigastric vein. The catheter along one of its branches was brought to confluence with the umbilical vein. The vein is dilated and tortuous, with a massive discharge of contrast into the left femoral vein (marked with an arrow) and further into the left external iliac vein. e - passage of the conductor and catheter (marked by arrows) into the umbilical vein.

Puncture and catheterization of the left femoral vein was performed. A multi-purpose 4F catheter was passed through the left inferior epigastric vein with a 0.035-inch hydrophilic guidewire passed into the umbilical vein, umbilical vein pressure ¬ 5 mm Hg. Given the low pressure in the umbilical vein, it was decided to refrain from its endovascular occlusion. In order to reduce the discharge of blood through the spleen into the portal vein pool and reduce portal hypertension, it was decided to perform endovascular occlusion of a part of the intraorgan branches of the splenic artery to reduce blood flow through the splenic artery. A Simmons-2 5F catheter was inserted into the branch of the splenic artery leading to the superior pole of the spleen using a hydrophilic guidewire. Endovascular occlusion was performed with cylindrical emboli 0.75 mm - 1 vial (30 pcs.). Then, alternately, 3 COOK IMWCE coils with a coil diameter of 3 mm and a length of 3 cm IMWCE-35-3-3 were implanted, and 2 coils with a coil diameter of 10 mm and a length of 10 cm IMWCE-35-10-10. The pressure in the umbilical vein was re-measured - 4 mm Hg. Control angiography of the splenic artery, celiacography, with return portography was performed. The tools have been removed. Hemostasis. Aseptic pressure bandage.

Control angiograms (fig. 4). There is a slowdown in contrast enhancement along the embolized branches of the splenic artery, a decrease in blood flow through the splenic artery. Contrasting of the parenchyma in the upper parts of the spleen is preserved. The venous phase occurs at 18 sec, the contrast of the umbilical vein is practically absent.



a b

**Figure 4. Angiographic images**

a - angiography of the splenic artery after ebolic. Spirals are marked with arrows. b - return portography after embolization. A significant decrease in the contrast of the umbilical vein (marked with an arrow) compared with the original portography.

Conclusion: Cravelle-Baumgartner syndrome.
Splenomegaly. Condition after endovascular occlusion of the branches of the splenic artery.

A control *ultrasound with duplex scanning* was performed one day after the intervention.

Free fluid in the abdominal cavity and small pelvis not determined.

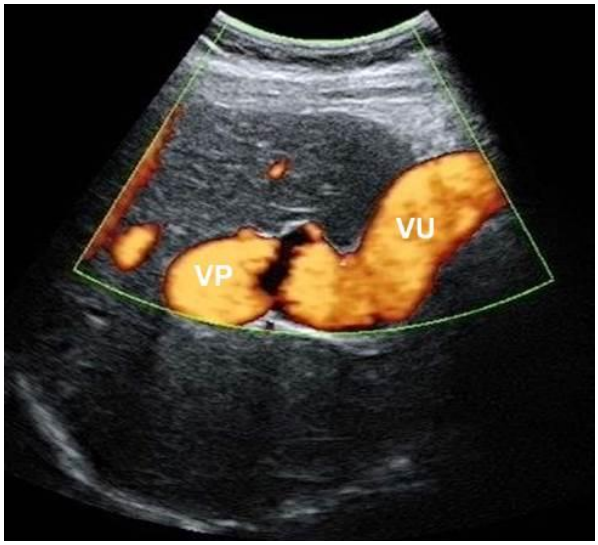
The liver is not enlarged in size (the size of the right lobe - 108x107 mm, the left lobe - 66x86 mm), the contours are even, clear, the parenchyma is of increased echogenicity, of a homogeneous structure. The vascular pattern is preserved. Intra- and extrahepatic bile ducts are not dilated, the lumen is free. The gallbladder is not enlarged (62 x 21 mm), the walls are 2.8 mm, curved in the body area, the content is homogeneous.

The hepatic veins are not dilated, the inferior vena cava on the hepatic segment is determined with a diameter of 17 mm, and a three-phase type of blood flow is maintained along them.

**Portal system veins:**

| Vein | Diameter | Linear blood flow velocity |
|---|---|---|
| Superior mesenteric vein | 7 mm | 19 m/s |
| Splenic vein | 6-18 mm | 0,26 m/s |
| Portal vein | 18 mm | 0,24 m/s |
| Right branch portal vein | 21 mm | 0,18 m/s |
| Left branch portal vein | 23 mm | 0,26 m/s |
| Umbilical vein | 20 mm | 0,16 m/s |

A collateral vein (recanalized umbilical vein) with a diameter of up to 20 mm (Fig. 5a) departs from the trunk of the portal vein; it can be traced along the dorsal surface of the anterior abdominal wall, pronouncedly tortuous, flows into the left external iliac vein (Fig. 5b).

a 

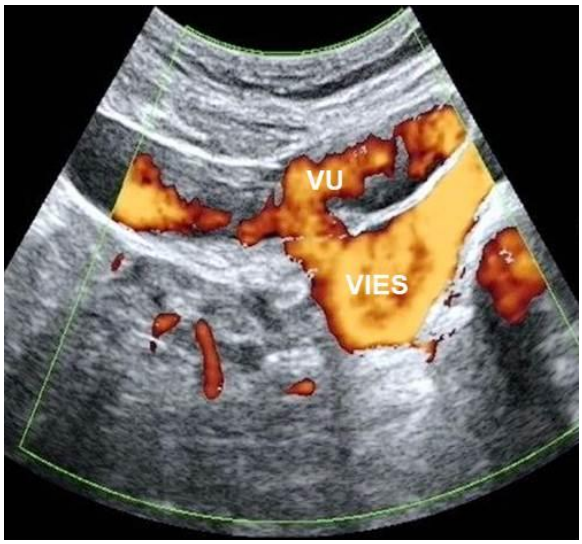
b 


**Figure 5. Ultrasound images, power doppler mapping**

a – portal (VP) and recanalized umbilical (VU) veins; b - a pronouncedly tortuous umbilical vein (VU) flows into the left external iliac vein (VIEs)

**Visceral arteries:**

| Artery | Diameter | Linear blood flow velocity | Resistance index |
|---|---|---|---|
| Superior mesenteric artery | 7 mm | 2,4 m/s | 0,82 |
| Truncus celiacus | 10 mm | 2,75 m/s | 0,69 |
| Common hepatic artery | 7 mm | 1,0 m/s | 0,62 |
| Splenic artery | 12 mm | 1,62 m/s | 0,66 |

In one of the splenic artery branches at the spleen hilum multiple hyperechoic emboli are visualized (Fig. 6a), the blood flow at the site of embolization is reduced, but preserved (Fig. 6).

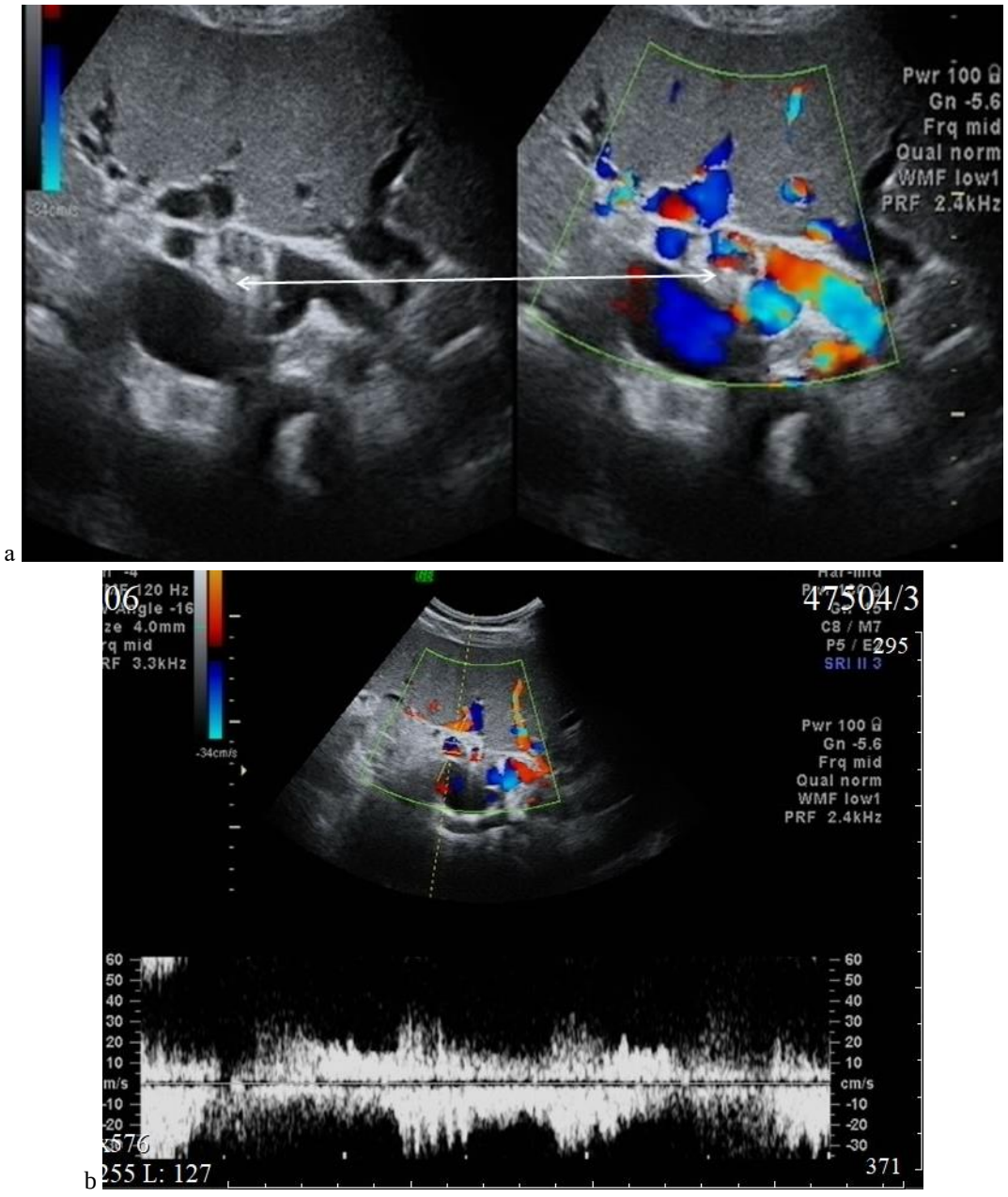



**Figure 6. Ultrasound images**

a – splenic artery in spleen hilum (arrow indicates emboli in the lumen of artery branch), combination of B-mode and color doppler imaging on one screen; b - registration of reduced turbulent blood flow in splenic artery branch with emboli

The pancreas is not enlarged in size: the head is 23 mm, the body is 13 mm, the tail is 19 mm, the contours are even and clear, the parenchyma is highly echogenic, the structure is homogeneous. Main pancreatic duct is not dilated.

The spleen is enlarged (200x69 mm, S=120 $cm^2$), the contours are even, clear, the structure of the parenchyma is homogeneous. There is an additional slice at the spleen gate, 32x23 mm in size. The vascular pattern of the spleen is Changed, single arteriovenous fistulas are determined intraparenchymatously.

Conclusion. Ultrasound signs of splenic arteriovenous fistulas, dilatation of the splenic and portal veins, and recanalization of the umbilical vein (Cruvelier-Baumgartener syndrome type II). Condition after endovascular occlusion of splenic artery branches.

*Control MDCT* two days after the intervention.

The trunk of the portal vein was expanded to 21 mm. A collateral vein (recanalized umbilical vein) up to 20 mm in diameter departs from the trunk of the portal vein, passes along the dorsal surface of the anterior abdominal wall, forming many bends, and flows into the left iliac vein. The

lumen of the left iliac vein above the confluence of the collateral was expanded to 32 mm (Fig. 7a).

Spleen with smooth, clear contours, enlarged (103x56x190 mm). In the upper spleen pole, there are areas that are weakly contrasted in the arterial and venous phases - ischemia zones (Fig. 7b). The rest parenchyma is not changed, the density in the native, arterial, venous phases is 46 HU, 120 HU and 100 HU respectively. In one of the splenic artery branches in the spleen hilum multiple hyperechoic emboli are visualized (Fig. 7b). Early contrast enhancement of the collateral splenic vein is not determined.

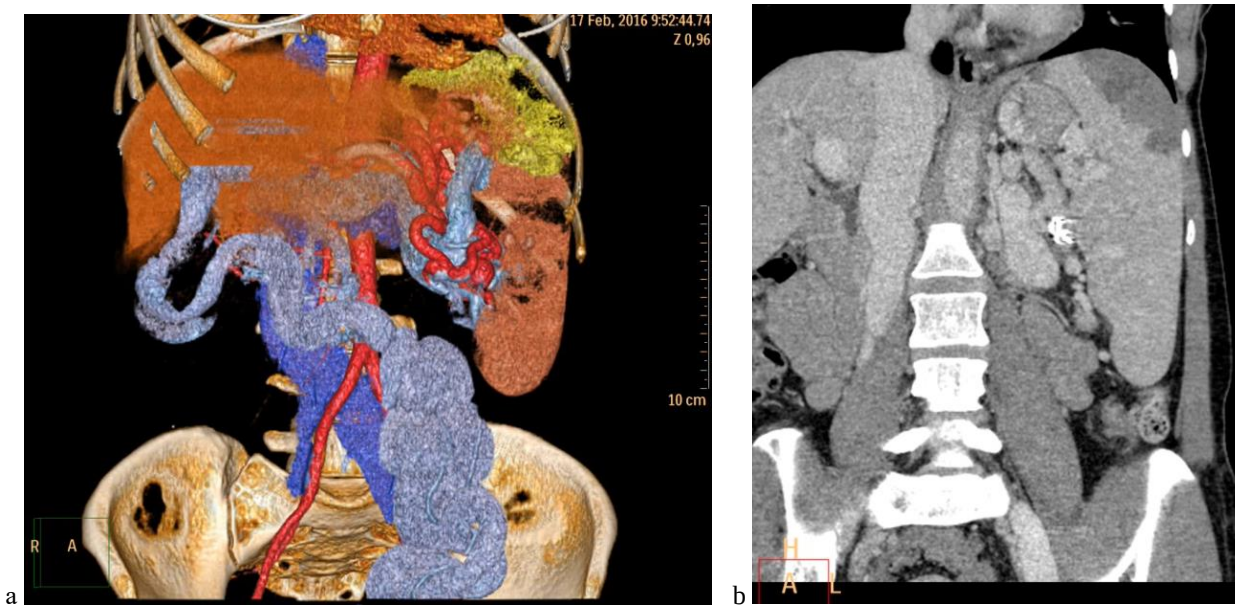



**Figure 7. Contrast enhanced MDCT: a - fused arterial and venous 3D; b – frontal view, venous phase**

Conclusion.

Cruvelier-Baumgartener syndrome type II. Splenomegaly. Condition after endovascular occlusion of splenic artery branches. Spleen infarction.

The postoperative period was uneventful, without features. The department carried out conservative therapy, dynamic observation.

At the time of discharge, the patient's condition was satisfactory.

The control MSCT study two months after the intervention showed no additional information. The patient had no complaints at the time of examination.

## DISCUSSION

In classic Cruveilhe-Baumgarten syndrome, the umbilical portion of the left portal vein drains into the paraumbilical vein, which emerges from the liver and then traces to the umbilicus. The paraumbilical vein looks like a tubular structure that arises in the falciform ligament between the liver left lobe, leading from the left portal vein to the veins of the anterior abdominal wall. Recanalization and development of large paraumbilical veins prevents bleeding from esophageal varices and also prevents predisposition to hepatic encephalopathy. This situation is considered an acceptable means of decompressing the portal venous system without concomitant gastrointestinal bleeding [16, 26-28].

The clinical significance of Cruvelier–Baumgarten syndrome is that these subcutaneous collaterals may undergo spontaneous bleeding or unintentional significant bleeding during abdominal surgery or paracentesis. Since portosystemic varicose veins develop by stretching and lengthening the pre-existing small veins, the walls of varicose veins are thin. These vessels break easily and are difficult to repair. There are many reports of cases of intraoperative mortality due to accidental rupture of varicose veins [29, 30].

However, patients usually have a non-specific history, such as a sensation of a lump in the left side of the abdomen [28]. In our case, the patient complained of recurrent moderate pain in the epigastrium and right hypochondrium. In addition to the classic finding (severe portal hypertension, venous collaterals with the inferior vena cava, recanalized umbilical vein), splenic artery arteriovenous fistulas were identified, which most likely explained the clinical manifestations. Our patient did not have a symptom of "jellyfish head", there were no external manifestations of varicose veins at all.

Diagnosis of the Cruveilier–Baumgarten syndrome, as well as the diagnosis of SAVF, is possible only with duplex ultrasound or MDCT, which are usually performed to determine the cause of portal hypertension [31-33]. MDCT is the imaging modality of choice, which identifies this post-systemic collateral pathway with exceptional accuracy [28, 34].

There are conservative and surgical treatment of Cruvelier-Baumgarten syndrome, as well as general recommendations [7, 8, 35].

General recommendations include diet therapy (reducing the amount of salt consumed to reduce fluid

retention in the body; reducing the amount of protein consumed to reduce the risk of hepatic encephalopathy).

Conservative treatment is aimed at eliminating the symptoms of the disease (pituitary hormones; nitrates; beta-blockers; synthetic analogues of somatostatin; diuretics; albumins; lactulose preparations; leukopoiesis stimulants; synthetic analogues of adrenal hormones; hemostatic drugs).

Surgical treatment includes:

• Portosystemic shunting;

• splenorenal shunting;

• devascularization of the lower esophagus and upper stomach (Sugiura operation) – ligation of some arteries and veins of the esophagus and stomach. The operation is performed to reduce the risk of bleeding from the veins of the esophagus and stomach. Usually operation is supplemented with a splenectomy;

• Transplantation, which is performed when it is impossible to restore the normal activity of the patient's own liver. The most common transplant is a part of the liver of a close relative.

With severe splenomegaly, the following is used: splenectomy; embolization of the splenic artery.

In the presented clinical case the leading manifestation requiring surgical correction was SAVF. Treatment options for SAVF are conventional open surgery, endovascular therapy, or a combination of both [36]. As a result of the performed endovascular occlusion of the branches of the splenic artery, there was a decrease in pressure in the portal system, as a result of which the discharge along the recanalized umbilical vein into the basin of the inferior vena cava was sharply reduced, which made it possible to level out complaints.

## CONCLUSION

Cruvelier-Baumgarten syndrome is a rare disease that causes portal hypertension in association with unclogged umbilical or paraumbilical veins, hypoplasia of the intrahepatic portal system, and liver atrophy with minimal or no fibrosis. Patients with this condition experience the classic signs of portal hypertension such as gastrointestinal bleeding, ascites, and encephalopathy. Frequent clinical manifestations are the symptom of "jellyfish head" and venous murmur over the umbilical region due to the presence of large umbilical and paraumbilical veins.

In the presented clinical case of Cruvelier-Baumgarten syndrome type II in combination with splenic artery arteriovenous fistula, pathological changes were not pronounced. There was no "jellyfish head" symptom, but a venous murmur was heard in the umbilical region. The patient complained of moderate pain in the epigastrium, right hypochondrium. Examination of the patient, which revealed, in addition to the typical manifestations of this disease, also multiple SAVFs, as well as angiography data regarding pressure in the umbilical vein - 5 mm Hg., determined the tactics of treatment in the volume of endovascular occlusion of the splenic artery branches, which was successfully completed. As a result, there was a decrease in pressure in the portal system and a sharp decrease in discharge through the recanalized umbilical vein into the basin of the inferior vena cava, which made it possible to level out complaints. At the control examination two months after treatment, the patient had no complaints.

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