

原 著

Surgical Correction of the Inferior Vena Cava Obstruction with Budd-Chiari Syndrome

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The Budd-Chiari syndrome is an uncommon disorder caused by obstruction to hepatic venous outflow. Since the early description by BUDD¹⁾ in 1846, and by CHIARI⁵⁾ in 1899, its concept has progressively broadened to include among its etiologies all the complete or incomplete obstruction of the hepatic veins and/or the corresponding portion of the inferior vena cava, whether they are congenital or consequent to thrombosis, tumoral compressions, abscesses, or vascular inflammations. In recent years there have been an increasing number of reports^{2,6-9,11,13,14,18)} of the Budd-Chiari syndrome caused by a fibrous membrane or septum occluding the inferior vena cava in the hepatic portion. The purpose of this paper is to report 3 cases of obstructive lesions of that portion which were surgically treated, and to discuss pathogenesis, diagnosis, and several surgical procedures.

Clinical Materials

Three patients were surgically treated for the occlusion of the hepatic portion of the inferior vena cava during the period from 1975 to 1979 at the 1st Department of Surgery, Ehime University Hospital and Takayama Red Cross Hospital. Representative symptoms and signs, results of liver function tests, and other laboratory data are listed in table 1 and diagrammatic representation of cavogram, thickness of occluding septum, surgical procedures and their results are listed in table 2.

Report of Cases

Case 1. A 50-year-old woman was admitted to the Takayama Red Cross Hospital on February 25, 1975, with an internal supramalleolar ulcer and brown pigmentation of the right leg. This woman also had occasional swelling of the lower extremities, varicose of legs, abdominal

Key words: Budd-Chiari syndrome, Inferior vena cava obstruction, Hepatic vein occlusion, Portal hypertension, liver cirrhosis.

索引語: バッド・キアリ症候群, 下大静脈閉塞, 肝静脈閉塞, 門脈圧亢進症, 肝硬変.

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Table 1. Symptoms and signs, laboratory findings.

Case	1		2		3	
	50.	F.	53.	M.	15.	M.
Edema of lower extremities	+	for 6y.	+	for 6y.	+	at 14
Weakness of lower extremities	+	for 6y.	+	for 6y.	—	
Varices of legs	+	for 6y.	—		—	
Abdominal distention	+	for 6y.	+	for 3y.	—	
Superficial collaterals	+	for 6y.	+	for 3y.	+	for 11m.
Hyperpigmentation of legs	+	for 2.5y.	+	for 6y.	—	
Ulcer of legs	+	for 1.5y.	—		—	
Dyspnea and vomiting on exertion	—		—		+	for 1.5y.
Ascites	+		+		—	
Hepatomegaly	+		+		+	
Splenomegaly	—		—		—	
Jaundice	—		—		—	
Esophageal varices	+		+		+	
IVC pressure (mm H ₂ O)	325		265		396	
Pressure gradient across obstruction (mm H ₂ O)	235		215		270	
Serum protein (g/100 ml.)	7.7		6.6		7.2	
Serum A/G ratio	1.7		1.5		1.6	
Serum bilirubin (mg./100 ml.)	1.5		1.2		1.5	
Serum GOT	22		18		16	
Serum GPT	10		16		12	
Retention rate of ICG (% at 15 minutes)	31.6		46		32	

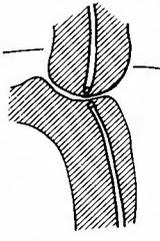
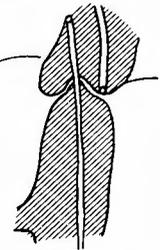
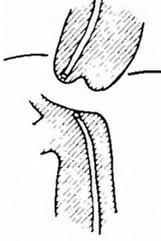
+ : present. — : absent. y.: years. m.: months.

distention and superficial collaterals on the thorax and the abdomen for the past six years.

Examination on admission disclosed the usual findings of cirrhotic portal hypertension. Blood pressure was 124/76 mmHg, pulse rate was 88 per min, and temperature was 36.6°C. The heart and lungs were normal to examination. The abdominal and chest wall presented numerous dilated, tortuous veins demonstrating reversed filling from below upward. A firm, but not tender liver edge was palpable 3 cm below the right costal margin in the midclavicular line. The abdomen was mildly distended, with a palpable fluid wave. There was no evidence of splenomegaly, jaundice or spider angiomas. The right leg showed a chronic stasis ulcer above the internal malleolus, with diffuse, brown pigmentation. A few tortuous varicose veins were present on the both legs. Moderate pitting edema of the lower extremities was present.

The hemoglobin was 14.1 g per cent; hematocrit, 48.5 per cent; red blood count, 478 million per mm³; white blood count, 3600 cells per mm³; platelets, 16.2 million per mm³; serum urea nitrogen, 15.4 mg per 100 ml; albumin, 4.8 g per 100 ml; globulin, 2.9 g per 100 ml; alkaline phosphatase, 8.7 K. A. units; prothrombin time, 14.5". The urinalysis was normal. The other laboratory datas are shown in table 1. An electrocardiogram showed a normal sinus rhythm. A chest X-ray film was normal. A clinical diagnosis of liver cirrhosis and portal hypertension

Table 2. Schematic representation of cavogram and operative procedures.

Case no.	1	2	3
Age and Sex	50. Female	53. Male	15. Male
Diagramatic representation of cavogram			
Thickness of occluding septum	1mm	1.5mm	7mm
Surgical procedure	Transatrial membranotomy	Membranotomy by a dilator	Transcaval resection and patch grafting
Result	Good	Good	GOOD

with a block in the inferior vena cava was made. To locate the site of the block a simultaneous double catheterization was performed on both vena cava. The superior catheter was introduced into the inferior vena cava via the right antecubital vein and the right atrium. The catheter could not be negotiated into the abdominal part of the inferior vena cava or the both hepatic veins. The inferior catheter passed up the right femoral vein via the right saphenous vein did not reach the thoracic inferior vena cava and the right atrium. An inferior caval venography was attempted by injecting a radiopaque dye through the inferior catheter. This revealed a block in the hepatic part of the inferior vena cava and the retrograde filling of the inferior vena cava, the right hepatic vein, the subdiaphragmatic veins, and extensive venous collaterals (Fig. 1-a). The caval venogram with two catheters revealed a membranous obstruction of the inferior vena cava in the hepatic portion (Fig. 1-b). Pressure in the thoracic inferior vena cava above the membranous obstruction was 90 mmH₂O; in the inferior vena cava below the obstruction, it was 325 mmH₂O; the hepatic

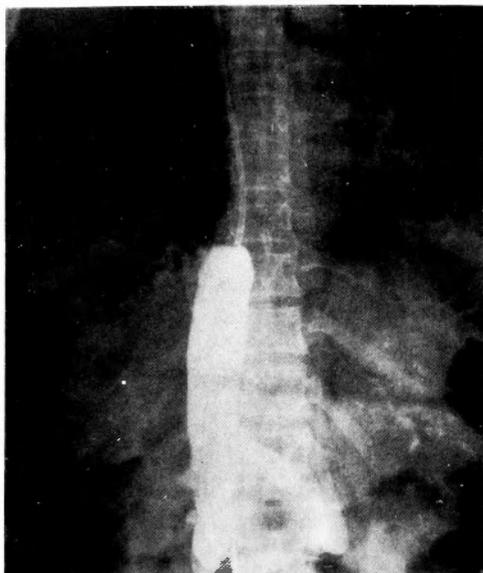


Fig. 1.-a

Fig. 1-a. Preoperative cavogram of case 1. with inferior catheter.
The inferior vena cava is occluded in the hepatic portion.

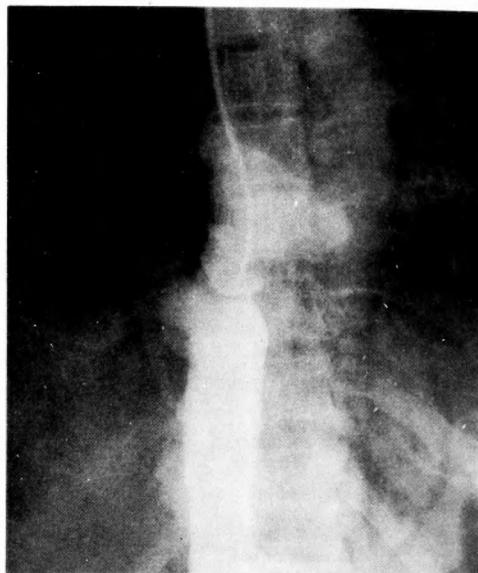


Fig. 1.-b

Fig. 1-b. Preoperative cavogram of case 1. with two catheters.
A membranous obstruction is revealed.

vein wedge pressure was 370 mmH₂O.

On March 6, 1975, median sternotomy was performed and the extracorporeal heart pump was readied. During incision of the subcutaneous tissue, numerous venous channels were divided, but achievement of precise hemostasis was not difficult. Intrapericardial exploration demonstrated a normal-appearing inferior vena cava communicating with the right atrium in a normal fashion. However, on intracardiac palpation through the atrial appendage, it was found that the inferior vena cava was completely occluded by a fibrous septum at the level of the diaphragm. The membrane was fractured by using the index finger tip which was inserted into the inferior vena cava through the right atrium (Fig. 2). Following rupture of the membrane, pressure within the inferior vena cava immediately upstream from the area of repair measured 140 mm H₂O. After operation hepatomegaly decreased, and ascites, edema of legs, and marked collateral veins disappeared. Cavography performed 4 months after the operation revealed that the inferior vena cava was wide open, and stasis of contrast material in the inferior vena cava below the diaphragm was not seen (Fig. 3-a). At 4 years follow-up, the patient was asymptomatic, without ascites, peripheral edema, or ulcer of the right leg, and a repeat inferior vena cavogram showed that the penetrated site was still open (Fig. 3-b).

Case 2. A 53-year-old male was admitted to the 1st Department of Surgery, Ehime University Hospital on January 26, 1977, presenting edema and hyperpigmentation of lower extremities. He had noted abdominal distention and superficial collaterals for the past three years.

On physical examination no cardiac murmurs were heard and the heart was not enlarged.

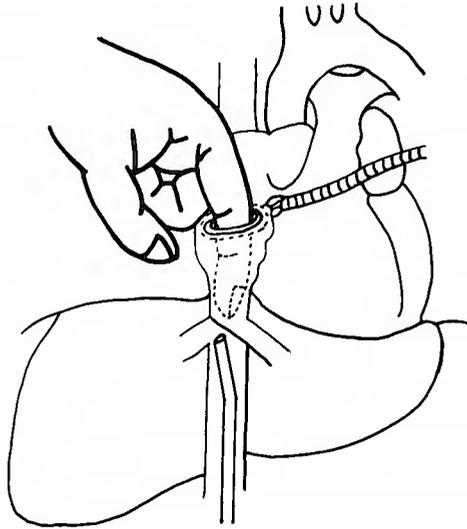


Fig. 2. A schema of transcatheter membranotomy.

Arterial pressure was 130/88 mmHg, pulse rate was 67 per min, and temperature was 36.6°C. The abdominal wall and the flank presented numerous dilated tortuous veins. The direction of blood flow was from below upward. The liver was moderately enlarged. It was smooth, and not tender. The abdomen was mildly distended, and the edema of the both legs were noted. There was no evidence of splenomegaly, jaundice, spider angiomas, varices and ulcer of the legs.

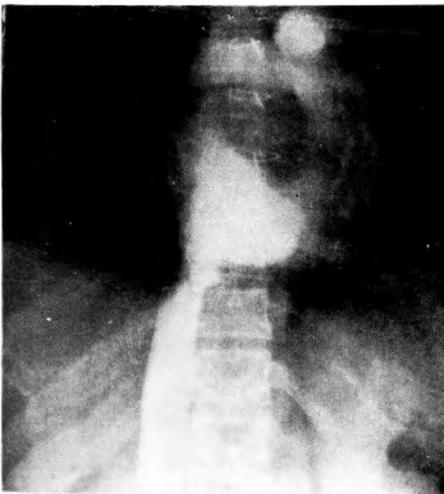


Fig. 3-a.

Fig. 3-a. Cavogram taken 4 months after transcatheter membranotomy in case 1. The inferior vena cava is wide open.

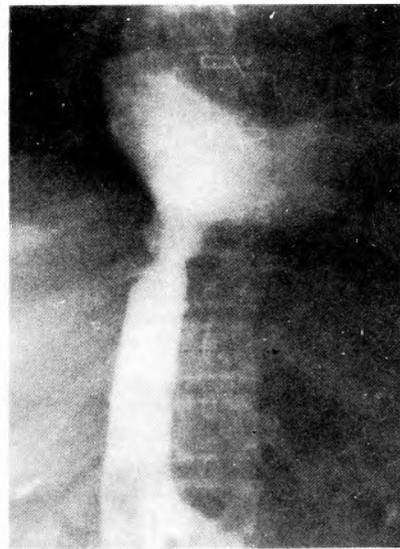


Fig. 3-b.

Fig. 3-b. Cavogram taken 4 years after transcatheter membranotomy in case 1. The penetrated site of the inferior vena cava is still open.

The hemoglobin, 11.9 g per cent; hematocrit, 33.4 per cent; red blood count, 379 million per mm^3 ; white blood count, 3500 cells per mm^3 ; platelets, 5.5 million per mm^3 ; serum urea nitrogen, 14 mg per 100 ml; albumin, 4.0 g per 100 ml; globulin, 2.6 g per 100 ml; alkaline phosphatase, 175 I.U. The urinalysis was normal. Other laboratory datas are listed in table 1. An electrocardiogram showed a normal sinus rhythm. A chest film was unremarkable. Hepatic vein catheterization through the right antecubital vein was performed, but the tip of the catheter could not be introduced into the hepatic vein or the inferior vena cava below the diaphragm. Another catheter was inserted through the right femoral vein by Seldinger technique into the inferior vena cava. It could be inserted into the right atrium. A cavogram showed incomplete membranous obstruction of the inferior vena cava at the level of the diaphragm (Fig. 4-a). The left and right hepatic veins were not visualized. Pressure in the thoracic inferior vena cava above the membrane was 50 mm H_2O ; in the inferior vena cava below the membrane, it was 265 mm H_2O ; the hepatic vein wedge pressure was 300 mm H_2O .

On March 15, 1977, under local anesthesia a Fogarty venous thrombectomy catheter (8-10 F) introduced via the right saphenous vein was passed into the right atrium through the hole in the membrane, and the balloon was inflated to approximately 1.5 cm in diameter and then pulled from the right atrium into the inferior vena cava against some resistance (Fig. 5-a). After the membrane was partially broken using a Fogarty catheter with a balloon, a dilator developed by Eguchi was inserted into the right femoral vein and advanced to the thoracic inferior vena cava

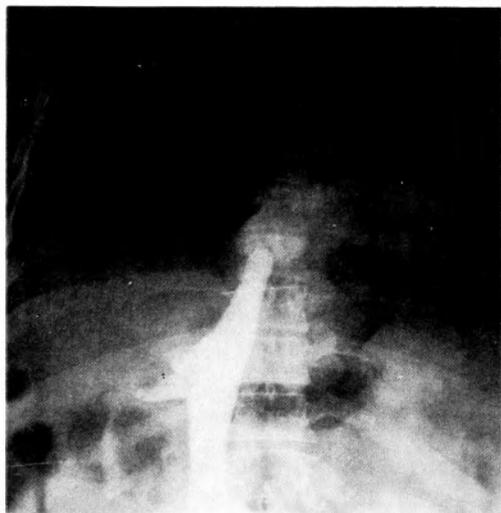


Fig. 4-a.

Fig. 4-a. Preoperative cavogram of case 2. with inferior catheter. Incomplete membranous obstruction of the inferior vena cava at the level of the diaphragm is shown.



Fig. 4-b.

Fig. 4-b. Cavogram after breaking the membrane by balloon and dilator in case 2. A complete opening of the inferior vena cava is shown.

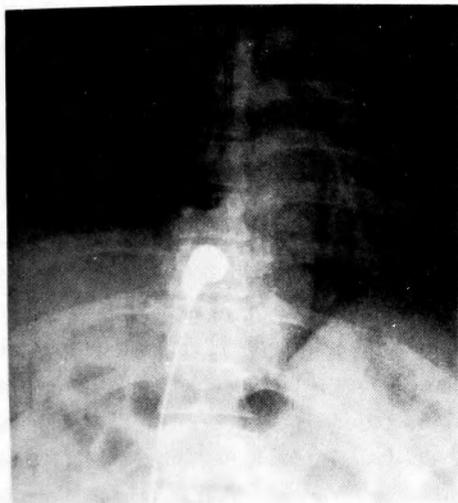


Fig. 5-a. Membranotomy by balloon in case 2.

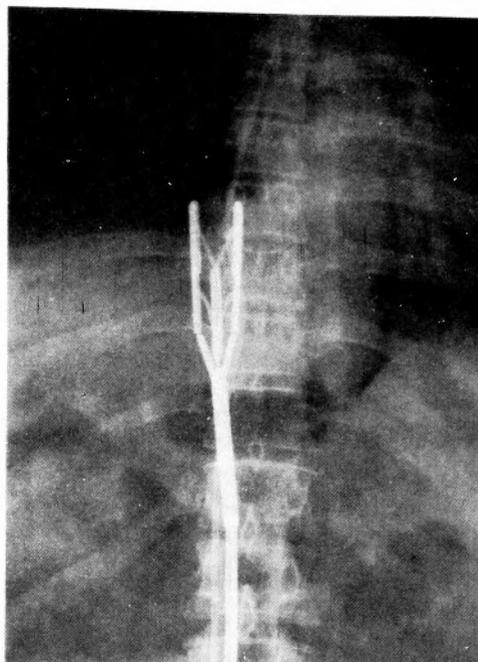


Fig. 5-b. Membranotomy by dilator in case 2.

through the opening in the membrane under fluoroscopic control. The dilator was then expanded to the previously set width to break the membrane (Fig. 5-b). Venography immediately after breaking the membrane revealed a complete opening of the inferior vena cava (Fig. 4-b). The postoperative course was satisfactory. The prominently dilated superficial veins on the abdominal wall and flank had markedly diminished. He was discharged on the 20th postoperative day. The patient is doing well and is free from ascites, edema, and varicose veins on the abdominal wall.

Case 3. A 15-year-old man was admitted to the third Department of Internal Medicine, Ehime University Hospital on March 19, 1979 for evaluation of possible liver disease due to obstruction of the inferior vena cava at the level of the diaphragm. He had dyspnea, palpitation, nausea, and vomiting on exertion for the past one and a half years. Nine months previously he had edema of the legs. Hepatomegaly and a few dilated vein over the abdomen and the thorax were noted by his physician. After making the diagnosis of Budd-Chiari syndrome due to an obstruction of the inferior vena cava at the level of the diaphragm, he was referred to the 1st Surgical Department of Ehime University.

At the time of the admission in our department, he appeared to be in excellent health. Blood pressure was 124/70 mmHg, pulse rate was 80 per min, and temperature was 37.4°C. There was no evidence of demonstrable physical abnormalities except for moderate hepatomegaly, and slightly dilated veins over the abdomen and the chest. The urinalysis was normal.

The hemoglobin was 10.2 g per cent; hematocrit, 33.1 per cent; red blood count, 448 million per mm^3 ; white blood count, 6700 cells per mm^3 ; platelets, 22.6 million per mm^3 ; serum urea

nitrogen, 16 mg per 100 ml; albumin, 4.4 g per 100 ml; globulin, 2.8 g per 100 ml; alkaline phosphatase, 246 I.U.; prothrombin activity, 73 per cent. An electrocardiogram showed a normal sinus rhythm. A chest x-ray film was normal. Caval catheterization and venography was attempted with two catheters, one introduced through the right antecubital vein and the other through the right femoral vein by Seldinger technique. The superior catheter could not be passed into abdominal part of the inferior vena cava. The inferior catheter could not be inserted into the thoracic inferior vena cava and the right atrium. A x-ray film after injection of contrast material into the abdominal inferior vena cava showed a complete block at the level of the diaphragm. Caval venogram with two catheters revealed a complete obstruction 7 mm in thickness in the vena cava just above the orifice of the right hepatic vein (Fig. 6-a). The right hepatic vein was widely patent, but the left was not visualized. Pressure in the thoracic inferior vena cava above the obstruction was 126 mm H₂O; in the inferior vena cava below the obstruction, it was 396 mm H₂O; the hepatic vein wedge pressure was 380 mm H₂O.

On July 12, 1979, under hypothermic general anesthesia, abdominal exploration was carried out through an upper midline incision. The incision was lengthened laterally and extended through a right sixth intercostal space. Right thoracotomy was performed. The diaphragm was incised radially toward the vena cava. The liver was completely mobilized except for major vasculature and the common bile duct. The round ligament was divided and the falciform liga-

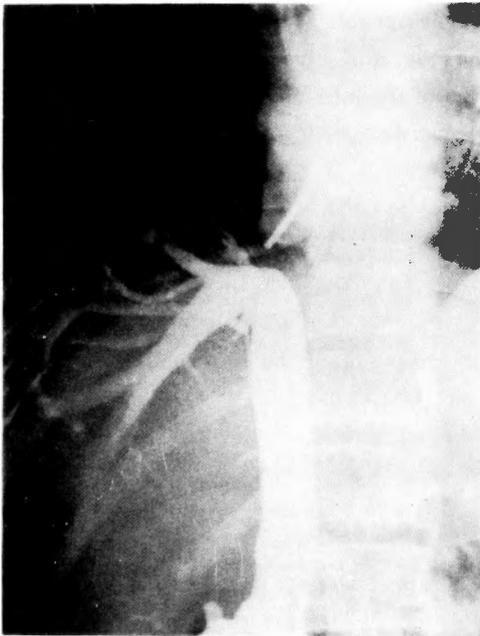


Fig. 6-a.

Fig. 6-a. Preoperative cavogram in case 3, with two catheters. A complete obstruction 7 mm in thickness in the vena cava is demonstrated.

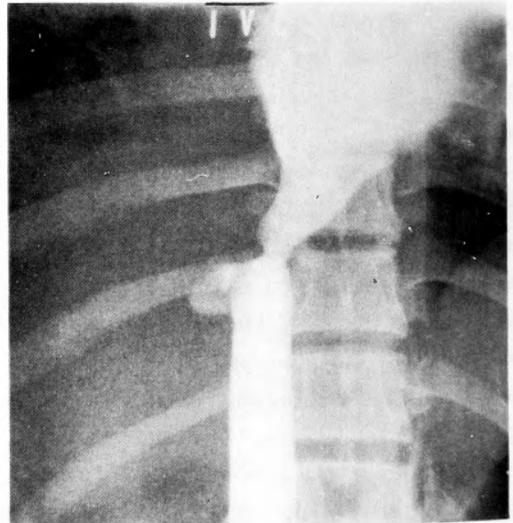


Fig. 6-b.

Fig. 6-b. Cavogram taken 2 months after operation in case 3. Patency of the inferior vena cava is demonstrated.

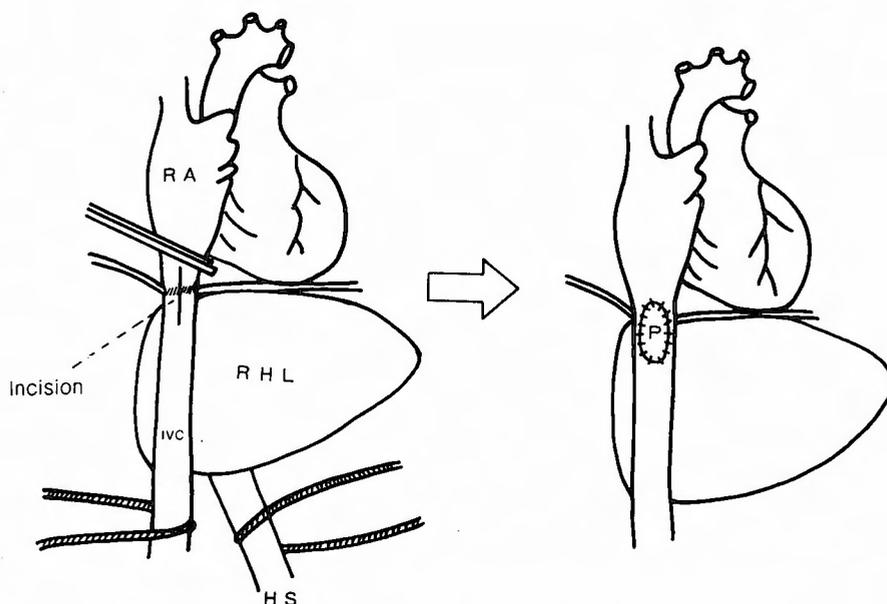


Fig. 7. Transcaval resection and patch-graft reconstruction under direct vision surgery (case 3).
 RA: right atrium, RHL: right hepatic lobe, IVC: inferior vena cava,
 HS: hepatoduodenal structures, P: patch-graft.

ment incised to the vena cava. Retraction of the liver was upward and to the left so that peritoneal reflections to the right lobe of the liver could be incised. Isolation of the right lobe of the liver was continued until the vena cava was completely free posteriorly. The hepatoduodenal structures, the infrahepatic and suprahepatic vena cava were separately occluded by cotton tapes and vascular clamp. A vertical incision was made in the vena cava over the obstructed site. The inferior vena cava was completely occluded by a fibrous septum at the level of the diaphragm. The occluding septum, measuring 7 mm in thickness, was excised. An autologous pericardial patch graft with teflon cloth was sutured into the caval wall defect to widen the vessel (Fig. 7). Patency of the anastomosis was demonstrated by angiography two months postoperatively (Fig. 6-b).

Discussion

Obstruction of the inferior vena cava by membrane or septum with secondary Budd-Chiari syndrome is characterized by signs and symptoms of inferior vena caval hypertension and portal hypertension and associated clinically with edema, brownish pigmentation, and varices of legs, superficial collateral veins on the abdomen and chest, abdominal distention, ascites, hepatomegaly, esophageal varices, and occasionally jaundice. If surgical decompression is not performed, the symptoms of portal hypertension become more prominent.

In 1963, KIMURA *et al*¹⁴⁾ collected 119 cases with complete or incomplete obstruction of the hepatic veins and/or the corresponding portion of the inferior vena cava. In 89 of these cases the

nature and extent of the obstruction were described according to venographic or postmortem examination. In 77 of these 89 cases, inferior vena cava were involved. Of 150 cases of Budd-Chiari syndrome collected by HIROOKA⁹⁾, 104 cases were accompanied with inferior caval obstruction or stenosis. In all of our 3 patients, all of Kimura's patients and all of YAMAMOTO's patients²⁰⁾, it was demonstrated by cavography or autopsy that the obstruction occurred at the same level in the inferior vena cava, i.e., at the level of the diaphragm and directly above the entrance of the main right hepatic vein. This vein was widely patent, but the left hepatic vein was not visualized by cavography. These findings have been confirmed at autopsy¹⁴⁾.

The cause of these cases is still uncertain, although the fact that the membranous structure or occluding septum is always found at the same site may favour the congenital origin. There are several theories which try to explain the embryological mechanism of this pathological condition. HIROOKA⁹⁾ explains it as an alteration in the development of the hepatic sinusoidal labyrinths that gives rise to the hepatic portion of the inferior vena cava. KIMURA¹⁴⁾ et al suggested a congenital origin related to the closure of the ductus venosus, which joins the left hepatic vein near its termination, comparing this venous anomaly with coarctation of the aorta, in which the aortic arch is constricted near its junction with the ductus arteriosus Botalli.

The low position of the obstructing diaphragm about 3 cm inferior to the attenuated eustachian valve indicates that the obstruction of the inferior vena cava is a different entity from the reported obstruction of the inferior vena cava by a persistent eustachian valve^{12,16)}.

The inferior vena cava was completely occluded in 2 of our 3 patients, and all of Kimura's patients¹⁴⁾ (6 cases), however, most patients did not show any signs and symptoms before adult life. YAMAMOTO²⁰⁾ reported a patient, aged 32, who was accidentally found to have the obstruction by cavography without any sign of circulatory disturbance. If the lesions are congenital in origin, the delayed appearance of clinical manifestation would probably indicate a deterioration in the compensating collateral circulation or thrombosis of the collateral vessels.

Surgical procedures for this condition are divided into conservative, bypass, and radical methods.

Conservative treatments—Supradiaphragmatic transposition of the spleen, fixation of the intestine with the abdominal wall, fixation of the omentum to the sternal bone marrow were reported to be effective in diverting portal blood flow into the superior vena cava.

Bypass operation—splenoazygos¹⁷⁾, mesoatrial³⁾, iliac-mesenteric-atrial⁴⁾, cavoatrial¹⁵⁾, azygocaval²⁰⁾, and others have been used with variable results.

Radical methods—Several techniques have been used for the direct surgical correction of the inferior vena cava obstruction. The closed transauricular fracture of the membrane^{8,11,13,14,18,20)} has been done with a finger. Recently, EGUCHI reported a new noninvasive technique using a dilator to fracture the membrane after breaking it partially by Fogarty balloon catheter.

According to our surgical experience, the selected surgical procedure for the inferior caval obstruction associated with the Budd-Chiari syndrome should be depended on the type of occlusion. When the obstruction is membranous or short and not complete in the cavography with a good diameter in the proximal and distal veins without thrombus, membranotomy by balloon and

dilator may be the procedure of choice. In cases with complete membranous obstruction, it is worthwhile to try a closed noninvasive procedure such as fracturing by balloon and dilator after penetrating the membrane using Brockenbrough catheter. If this procedure is unsatisfactory, Kimura's transcardiac membranotomy is very effective to evacuate the occlusion. If the cavography points to a long obstruction, teflon mesh or autologous pericardium patching after an open resection with direct view is the recommended operation^{19,20}. When the obstruction is so long and excision of the occluded segment is impossible, a bypass operation is recommended.

Summary

- 1) Three patients of inferior vena cava obstruction with Budd-Chiari syndrome were reported. Two patients were treated by direct surgical approach to the inferior vena cava block, i.e., transcardiac membranotomy, and patching with autologous pericardium after an open resection. One patient was treated using a dilator to fracture the membrane.
- 2) Some considerations of pathogenesis, diagnosis, and several surgical procedures are discussed. According to our experience, the selected surgical procedure for the inferior caval obstruction should be depended on the type of occlusion.

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和文抄録

Budd-Chiari 症候群を伴った下大静脈閉塞症の外科治療

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岩 橋 寛 治

Budd-Chiari 症候群を伴った肝部下大静脈閉塞症の報告例は次第に増加しつつあり、手術方法も種々考案されてきているが、閉塞の型式によって最も適当な手術方法を選ぶべきであると考え、自験例を参考に検討するとともに、その病因についても言及した。1) 下大静脈閉塞部が膜様であり、しかも不完全の場合は dilator による拡張が侵襲も少なく適当である。2) 閉塞が膜様でしかも完全閉塞であれば Brockenbrough's

catheter による穿孔後 balloon および dilator による膜裂開、または経心的膜破碎術が適当である。3) 閉塞部が広汎困の場合は直達手術の適応となる。本症の3例に対し手術を施行した、即ち完全膜様閉塞には経心的膜破碎術を、不完全膜様閉塞には balloon, dilator による膜裂開を、7mm の厚さの完全閉塞には直視下に閉塞部切除、パッチ縫着を行った。3例とも術後開存が認められた。