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Effect of Spontaneous Portosystemic Shunts on Hemorrhage from Esophagogastric Varices

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Abstract. The role of a massive spontaneous portosystemic shunt (MSPSS) in cirrhotic patients with portal hypertension remains unclear. The aim of this study was to investigate clinical outcomes and portal hemodynamic changes following ligation of the MSPSS during devascularization surgery. Portography and gastroendoscopy were performed before and after surgery for hemodynamic and follow-up studies. Three types of MSPSS were demonstrated portographically: 22 portoumbilical shunts, 18 splenorenal shunts, and 2 inferior mesenteric-caval shunts. A total of 40 MSPSS patients with esophagogastric variceal (EGV) bleeding underwent surgery: 26 had ligation of the MSPSS, and the remaining 14 served as the nonligation group. Neither the preoperative mean portal pressure (MPP) nor the postoperative MPP were significantly different between the ligation and nonligation groups ($p > 0.1$), and there was no significant difference regarding surgical mortality, recurrent varices, or cumulative survival rate for the two groups in the follow-up study. However, postoperative portography demonstrated persistent drainage of portal flow and decreased intrahepatic portal perfusion in the nonligation patients. Clinical signs of hepatic encephalopathy subsided after ligation of the MSPSS in three patients. Therefore ligation of the MSPSS, which may be responsible for the development of encephalopathy, is recommended during devascularization surgery for EGV in cirrhotic patients.

Cirrhotic patients frequently develop a vast variety of portosystemic collaterals as a consequence of long-standing portal hypertension [1, 2]. With the use of direct portography, previously unknown spontaneous portosystemic shunts have been increasingly recognized. Massive spontaneous portosystemic shunts (MSPSSs) are occasionally identified in a small group of patients. They are usually of large diameter and drain a significant amount of splanchnic blood into the systemic circulation, bypassing the esophagogastric veins [1, 2]. The clinical importance of the shunts remains of great interest, as there is a discrepancy regarding the protective effect of MSPSSs from esophagogastric variceal (EGV) bleeding and the development of hepatic encephalopathy in cirrhotic patients [1–8]. Many cirrhotic patients continue to maintain an elevated portal pressure and suffer from variceal hemorrhage even in the presence of an MSPSS. We know of no literature concerning whether MSPSSs should be eradicated or preserved during EGV surgery. Thus we attempted to investigate the clinical outcomes and changes in the portal hemodynamics of cirrhotic patients following ligation of the MSPSS during devascularization surgery to determine if an MSPSS affects the risk of variceal hemorrhage and encephalopathy.

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Patients and Methods

Patients

A total of 42 patients (35 men, 7 women) with a median age of 52 years (range 37–68 years) were included in this study during 1991–1997. Liver cirrhosis was documented in all patients by histologic diagnosis, and they had had at least one previous or current episode of gastrointestinal bleeding. All of the patients were subjected to routine clinical, laboratory, endoscopic, and portographic examinations; and esophagogastric varices were well documented by endoscopic examination in all of them. Altogether, 40 patients underwent surgery and were then divided into ligation and nonligation groups depending on whether the MSPSS was ligated or preserved during surgery. The clinical data for these patients are shown in Table 1.

Percutaneous Transhepatic Portography

Percutaneous transhepatic portography (PTP), previously described, was used to evaluate the number, size, and flow pattern of the shunts and EGV preoperatively in all patients [9]. The portal pressure was measured when the tip of the catheter was in the main portal trunk. The puncture site in the mid-axillary line was the reference level. The presence of an MSPSS was considered when the contrast drained along the shunting channel, clearly delineating the inferior vena cava on a portogram (Fig. 1).

Types of EGV and MSPSS

All 42 patients with MSPSSs had EGV, as evidenced by preoperative endoscopy and portography. According to the endoscopic findings, 15 had esophageal varices, 16 had gastric varices, 9 had EGV,

Table 1. Clinical characteristics of MSPSS patients with surgery.

Clinical data	Ligation group (n = 26)	Nonligation group (n = 14)
Age (years), mean and range	52 (41–68)	53 (37–66)
M:F	21:5	12:2
Etiology of portal hypertension		
Postnecrotic cirrhosis	26	12
Alcoholic cirrhosis	0	2
Child-Pugh classification		
A	6	3
B	15	7
C	5	4
Type of MSPSS		
Portoumbilical	10	12
Splenorenal	16	2
Surgery		
ET	10 EV	5 EV + 2 EGV
PG	11 GV + 5 EGV	2 GV + 5 EGV

ET: esophageal transection; PG: proximal gastrectomy; EV: esophageal varices; EGV: esophagogastric varices; MSPSS: massive spontaneous portosystemic shunt; GV: gastric varices.

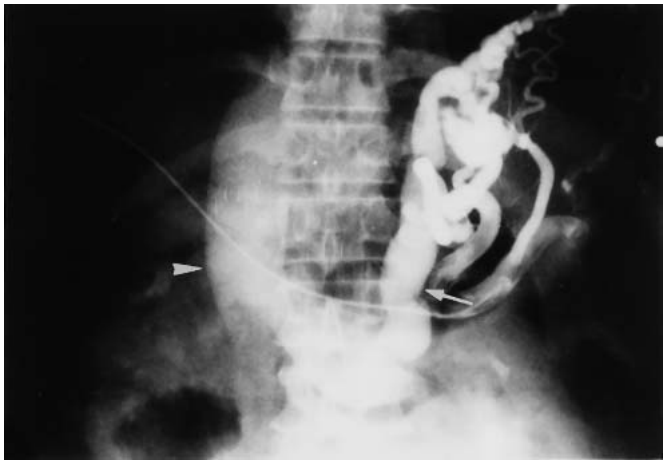

Fig. 1. Transhepatic portogram demonstrating gastric varices with a large splenorenal shunt (arrow) draining into the left renal vein and inferior vena cava (arrowhead).

and 2 had both esophageal and rectal varices. We also classified the 42 patients with MSPSSs into three types based on the portographic findings: type I, 22 patients with a portoumbilical shunt (PUS) (Fig. 2); type II, 18 patients with a splenorenal shunt (SRS) (Fig. 3); and type III, 2 patients with a inferior mesenteric-caval shunt (IMCS).

Surgery

To eradicate the EGV, nonshunting devascularization procedures including splenectomy, devascularization, and esophageal transection (or proximal gastrectomy) were undertaken. The surgical procedures have been described in detail in the literature [10–12]. The choice of esophageal transection or proximal gastrectomy depended on the predominance of the esophageal or gastric varices based on portographic findings.

One of the two patients with rectal varices received local injections of sclerosing agent to control the rectal bleeding; the other was treated conservatively with propranolol (Inderal). No additional surgical procedures were used in these two patients. One of the two died of hepatocellular carcinoma during the period of hospitalization; the other, who refused the operation, was lost to follow-up.

In the ligation group, ligation of the MSPSS was accompanied by a devascularization procedure in 16 patients with SRS and 10 with PUS. In contrast, in the nonligation group MSPSSs were preserved in 12 patients with PUS and in 2 with SRS. Of the ligation patients, 10 underwent esophageal transection (ET) and 16 proximal gastrectomy (PG), whereas in the nonligation group, 8 had ET and 4 PG. These patients were followed with endoscopy and portography for variceal recurrence and hemodynamic changes of the portal system for 3 to 6 years after surgery.

Statistics

All data were analyzed using the Statistical Package for the Social Sciences Version 8.0 software (SPSS, Chicago, IL, USA). Cumulative survival rates were calculated by the Kaplan-Meier method, and the difference in survival rates between the two groups was analyzed by the log-rank test. Results are expressed as means ± SD. Statistical analysis was done using Student's t-test or the χ^2 test. A probability of less than 0.05 was considered statistically significant.

Results

Clinical Outcomes

In the ligation group, three patients died. One death was due to respiratory failure, and one was due to continuing massive bleeding within 1 week after surgery; the third patient developed progressive jaundice and died of hepatic failure within 1 month of surgery. All three patients were of Child's class C. One case was complicated by a left subphrenic abscess due to extravasation of the pancreatic juice. Regarding the other patients, during the follow-up period two died of progressive hepatic insufficiency within 1 year, and one died of hepatocellular carcinoma within 3 years. Of the remaining 20 patients, 3 had hepatic encephalopathy prior to surgery, but no recurrence was found postoperatively.

In the nonligation group, one developed pulmonary infection during the postoperative period and died 2 weeks later. The postoperative complications included anastomotic leakage in one and hepatic encephalopathy in one. These patients ultimately recovered with conservative treatment. Two other patients died of hepatic failure 2 years after surgery. All the surviving patients had been followed up with endoscopy and portography. The mean follow-up was 42 months (range 35–72 months).

The complication rate (p = 0.232), mortality rate (p = 0.658), and cumulative survival rate (p = 0.866) (Fig. 4) did not differ significantly between the ligation and nonligation patients.

Portal Pressure Change

Prior to surgery, pressure studies confirmed the presence of portal hypertension in all patients. In the ligation group, the mean portal pressures (MPPs) before and after surgery were 38.2 ± 5.2 and 36.8 ± 4.5 cm of saline, respectively. The difference was not statistically significant (p = 0.454). Postoperatively, the MPP decreased from 37.1 ± 4.8 to 36.1 ± 5.4 cm of saline in the nonligation group. There was also no significant change between the preoperative and post-

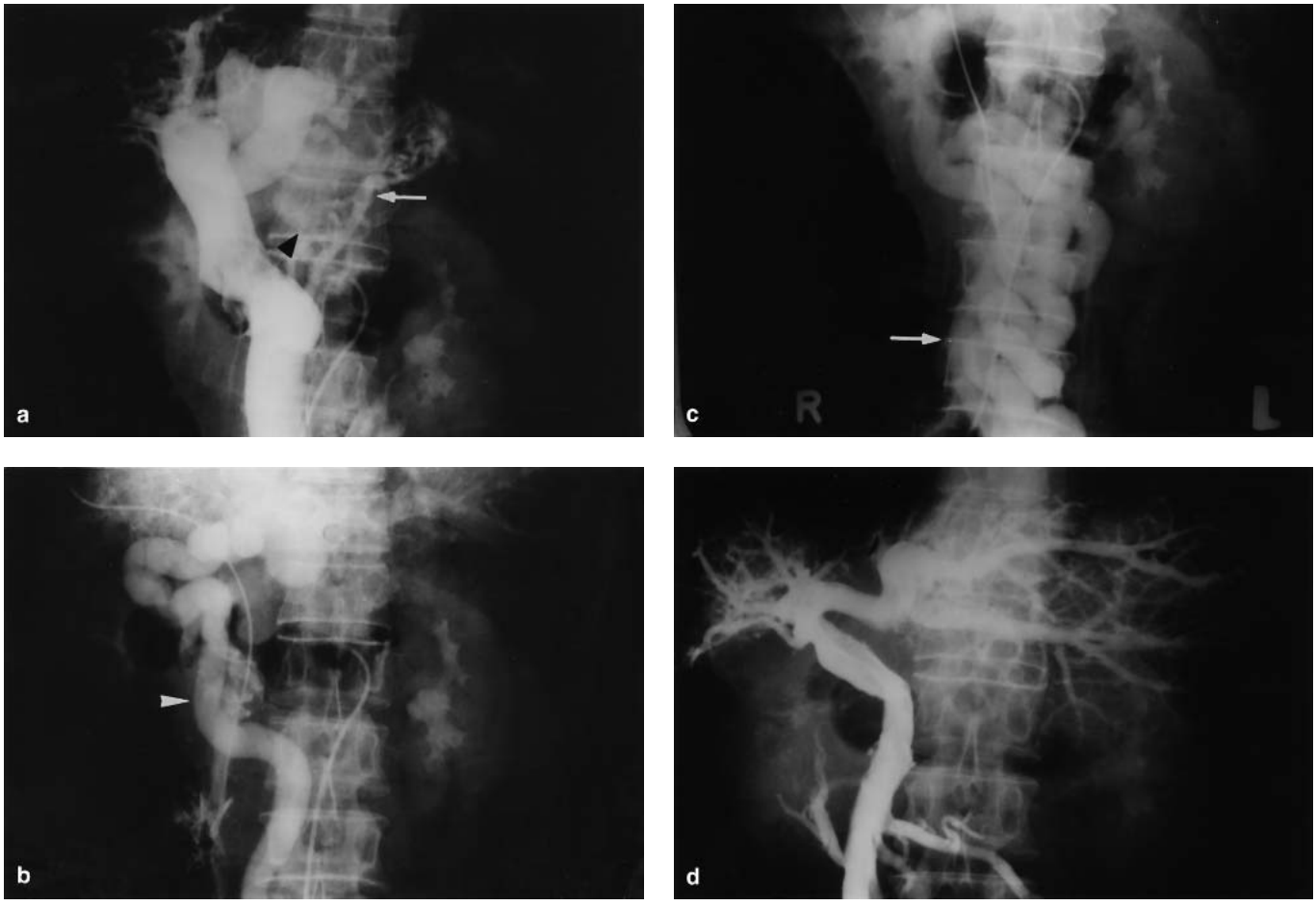

Fig. 2. a. Preoperative transhepatic portogram showing a coronary vein (arrow) arising from the bifurcation of the portal and superior mesenteric veins. Also shown is the origin of the paraumbilical vein (arrowhead). **b, c.** Hepatofugal flow continues to drain via a huge paraumbilical vein (**b,** arrowhead) into the superficial epigastric vein. **d.** Neither esophageal nor gastric varices are visible 5 years after surgery in the postoperative portogram, nor is the paraumbilical vein.

operative MPPs in the nonligation patients ($p = 0.465$). Neither the preoperative MPP nor the postoperative MPP were significantly different between the ligation and nonligation groups ($p > 0.05$) (Fig. 5).

Recurrence of Esophagogastric Varices

Postoperatively, 4 of the 20 patients in the ligation group had endoscopically demonstrable EGV, and recurrent varices developed in 2 of the 11 nonligation patients, although the recurrent varices were of mild severity. The difference in the recurrence of varices between these two groups was not statistically significant ($p = 0.926$). None of the patients presented with recurrent EGV bleeding during the follow-up period.

Postoperative Portography

Complete disappearance of the MSPSS was documented by follow-up portography in all 20 patients whose shunts were ligated. Neither the coronary vein nor other collaterals leading to the esophagogastric region were visualized in 17 patients (Figs. 2, 3). Three patients developed newly formed collaterals that drained into the esophagus. In the nonligation group, only one patient presented with EGV on postoperative portograms. Here, the features and flow pattern of the MSPSS remained the same as before, and persistent drainage of a large amount of portal blood via the MSPSS and decreased intrahepatic portal perfusion were demonstrated by postoperative portography (Fig. 6). Commonly, the recurrent varices supplied by several retroperitoneal veins originated from the portal or superior mesenteric veins.

Discussion

New collateral channels almost invariably develop in response to portal hypertension to shunt portal blood into the systemic circulation, bypassing the liver. In most cirrhotic patients, the coronary-azygos system serves as the most common pathway for decompressing portal hypertension through the esophageal and periesophageal veins [13, 14]. However, portal decompression is poorly accomplished by EGV alone because of the insufficiency of these varices to relieve portal hypertension. Therefore in some patients recanalized embryonic venous channels reopen and provide an alternate route to divert more portal flow into the systemic circulation [1–3]. In our study, with the use of direct portography, sev-

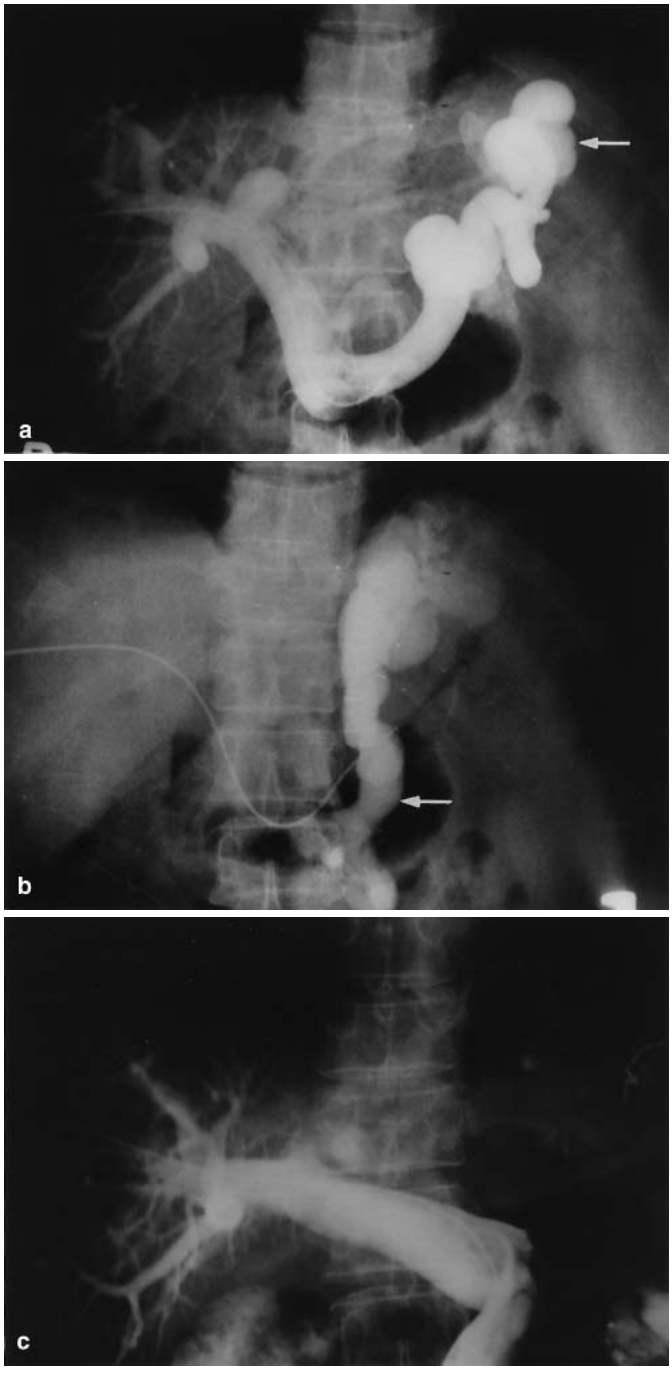

Fig. 3. a. Preoperative portogram shows gastric varices (arrow) without esophageal varices. **b.** Hepatofugal flow continues to drain into the left renal vein through the splenorenal shunt (arrow). **c.** Postoperative transhepatic portogram demonstrates the superior mesenteric vein and the portal vein. No varices are visible 5 years after surgery.

eral such channels were clearly demonstrated, including the PUS, SRS, and IMCS. These collaterals are usually of large caliber and thus shunt substantial amounts of portal blood into the systemic circulation. According to Poiseuille's law, small changes in vessel diameter have profound effects on vascular resistance, assuming the flow remains constant. Theoretically, one might expect that these channels would protect cirrhotic subjects from variceal hemorrhage by shunting portal blood, thereby lowering the portal pressure.

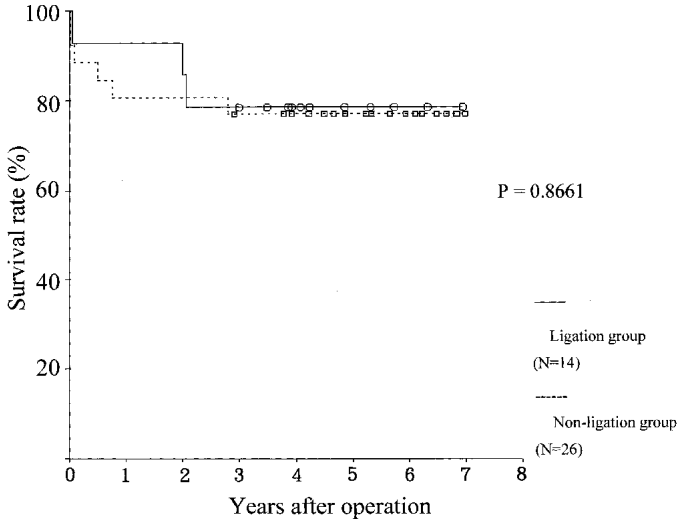

Fig. 4. Cumulative survival rates of the ligation and nonligation groups. Solid line: ligation group (n = 14); broken line: nonligation group (n = 26).

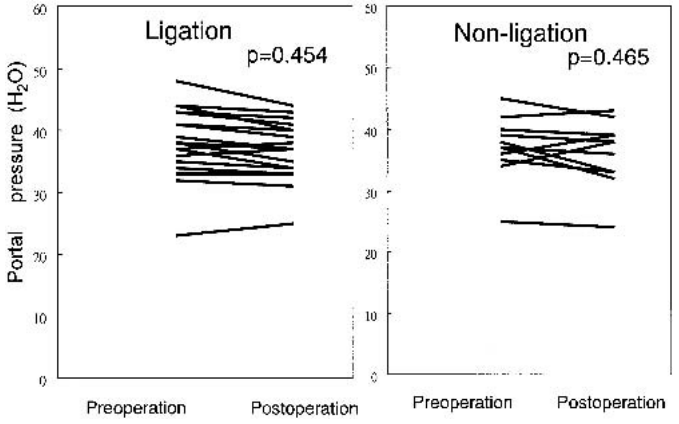

Fig. 5. Changes in portal pressure before and after surgery in the ligation and nonligation groups.

Although the role of MSPSSs in portal hemodynamics has been widely investigated, conflicting results have been reported in the literature regarding the potential efficacy of an MSPSS on portal decompression in terms of the risk of variceal hemorrhage. Some authors have found that a functioning MSPSS relieves portal hypertension, with an absence of EGV and the usual clinical appearance of liver disease [1–3]. In contrast, others have reported that neither the incidence nor the severity of variceal hemorrhage decreased even in the presence of large-bore collaterals [1–4, 7]. Consistent with recent findings [1–4, 7], EGV of varying degree may exist regardless of whether paraumbilical, gastrorenal, or mesenteric-caval communications are present, as shown by endoscopy and portography in this series. The actual incidence of MSPSSs in patients with liver disease may be much more common than we have recognized, as they are able to develop spontaneously.Most manometric studies have shown that cirrhotic patients with large shunts usually have a high portal pressure [1, 4, 15]. Several studies have reported that the mean splenic pulp pressure of patients with MSPSSs compared with that of patients without an MSPSS did not reach statistical significance [1, 4, 14, 16]. They also found, however, that among the patients with spontaneous shunts, however, the splenic pulp pressure was similar in patients with or without

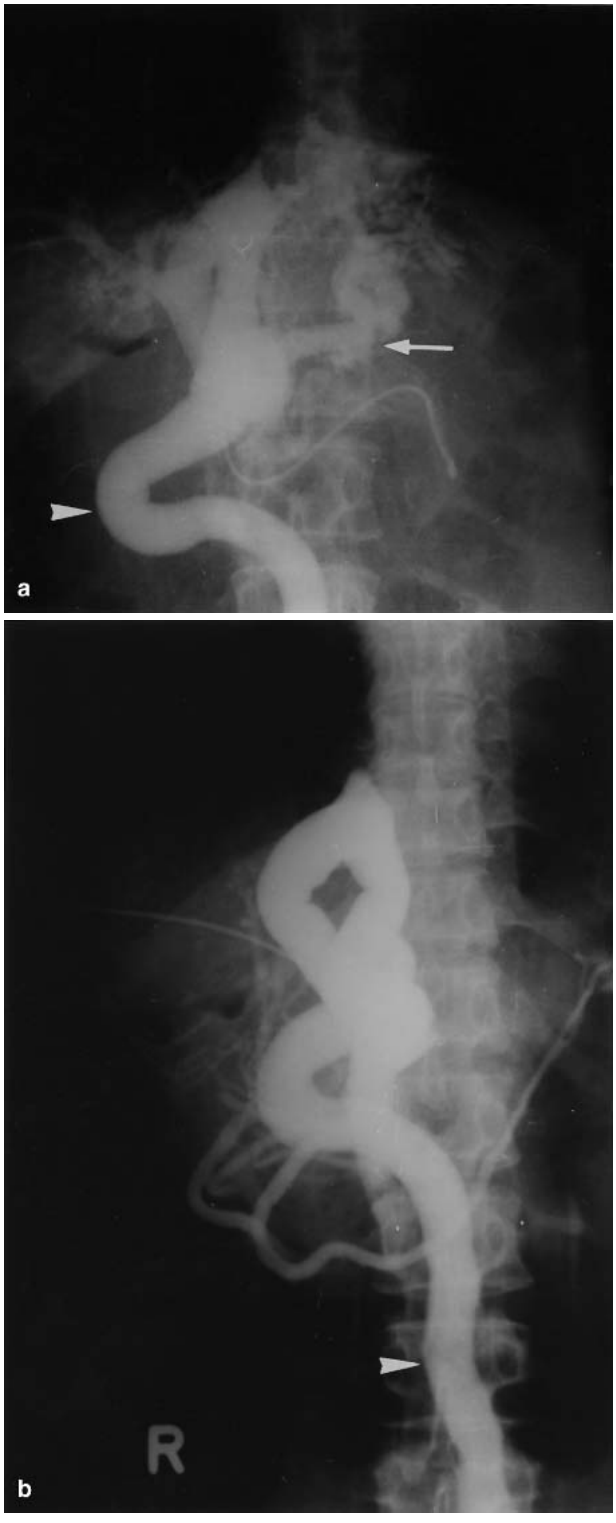

Fig. 6. a. Preoperative portogram showing esophageal varices (arrow) and a large paraumbilical vein (arrowhead). **b.** Postoperative portogram showing poor visualization of the intrahepatic portal branches and persistent drainage of the large paraumbilical vein (arrowhead).

bleeding [1, 4, 14, 16]. These observations suggest that most spontaneous shunts probably already exist, and they reopen and enlarge as a result of increased portal pressure. Moreover, the finding that a spontaneous splenorenal shunt may develop following variceal obliteration with sclerotherapy in cirrhotic patients might also help to explain this phenomenon [17]. However, based on the sparse studies on this subject, it appears that these nonsurgical portosystemic shunts do little hemodynamically to help decompress the portal system in most patients [1, 12]. Thus the risk of variceal bleeding remains the same.

Our study also revealed that there was no significant difference between the MPP before and after devascularization surgery with the shunt ligated. These data suggest that the portal pressure is not affected by the type, flow pattern, or direction of the MSPSS; and the presence or lack of a large shunt has little effect on the portal pressure. In accord with the findings of most hemodynamic studies, we also clarified the proposal that the MSPSS is clinically ineffective in protecting cirrhotic patients from variceal bleeding. In the cirrhotic liver, intrahepatic resistance may be affected by deposition of fibrous tissue and proliferation of myofibroblasts around the sinusoid and terminal hepatic venules, resulting in increased resistance and thereby contributing to portal hypertension. We postulate that intrahepatic resistance plays a more important role in the pathogenesis of variceal bleeding than does the MSPSS.

Unfortunately, the development of recurrent varices is inevitable following devascularization, as the portal system remains in a hyperdynamic state. In this study the recurrence rate of EGV was acceptable and is consistent with that in other clinical reports [11, 12]. There was no statistical difference of the incidence of recurrent varices between the two (ligation versus nonligation) groups. The results again support the earlier observation that MSPPSs had no effect on the portal pressure and thus on the occurrence of recurrent varices. MSPSS might be considered a specific alternative to portosystemic collaterals for partial or incomplete portal decompression and, ultimately, hemodynamic change, although they do not develop episodes of spontaneous bleeding as do the usual varices.

Hepatic encephalopathy has been reported with varied frequency in cirrhotic patients with an MSPSS because these large portosystemic communications resulted in enough of a shunt to facilitate the development of hepatic encephalopathy in some patients [1, 4, 9]. Previous studies have revealed that large esophageal varices are significantly less common in MSPSS patients with encephalopathy, although the risk of variceal bleeding remains the same [1, 16]. Moreover, the development of encephalopathy is mainly related to the diameter of the portal vein, not the shunt diameter [13]. In our series, three patients of the ligation group had clinical signs of hepatic encephalopathy before surgery, the degree of which was classified as mild to moderate. Dramatically, the episode of hepatic encephalopathy did not reappear during the 4 years of follow-up. Moreover, in the nonligation group, persistent drainage of large amounts of portal blood via the MSPSS and decreased intrahepatic portal perfusion were seen by postoperative portography. We believe that preservation of the harmless MSPSS might reduce perfusion of intrahepatic portal flow and increase the risk of developing hepatic encephalopathy. Thus, ligation of an MSPSS may be beneficial in patients with hepatic encephalopathy owing to the possible increase in the intrahepatic portal flow.

Conclusions

An MSPSS may be regarded as an alternative to a shunt channel, as it is insufficient to decompress portal hemodynamics and is unable to protect cirrhotic patients from variceal bleeding. MSPSSs may prevent the development of large EGV but not variceal hemorrhage. Ligation of the MSPSS is recommended during surgery for EGV in cirrhotic patients.

Résumé. Le rôle des shunts spontanés portosystémiques (MSPSS) chez le patient cirrhotique porteur d'hypertension portale n'est pas claire. Le but de cette étude a été d'explorer l'évolution clinique et les changements hémodynamiques du système porte après ligature des shunts MSPSS pendant la chirurgie de dévascularisation. On a réalisé une portographie et une gastroendoscopie avant et après la chirurgie afin de constituer des études hémodynamiques et de suivi. On a mis en évidence trois types de MSPSS par portographie: 22 shunts porto-ombilicaux (PUS), 18 shunts spléno-rénaux (SRS) et deux shunts mésentérico-cavaux inférieurs (IMCS). Quarante patients MSPSS avec des varices oesogastriques (EGV) hémorragiques ont eu un acte chirurgical. Parmi eux, 26 ont eu une ligature des MSPSS alors que les 14 autres ont servi de groupe témoin. Ni la pression porte moyenne (MPP) préopératoire ou postopératoire n'ont été différentes de façon significative entre les groupes ligature ou pas ($p > 0.1$). Aucune différence significative n'a été observée en ce qui concerne la mortalité chirurgicale, la récidive des varices ou la survie cumulative entre les deux groupes. Cependant, la portographie postopératoire a démontré un drainage persistant du débit portal et une diminution de la perfusion porte intrahépatique en cas de non ligature. Les signes cliniques d'encéphalopathie hépatique ont diminué après ligature des MSPSS chez trois patients. Puisque quelques patients peuvent tirer un bénéfice de la prévention d'encéphalopathie par MSPSS, la ligature des MSPSS est recommandée pendant la chirurgie de dévascularisation pour EGV chez le cirrhotique.

Resumen. El papel del shunt portosistémico espontáneo masivo (SPSEM) en pacientes cirróticos con hipertensión portal aún no ha sido dilucidado. El propósito del presente estudio fue investigar el resultado clínico y los cambios hemodinámicos luego de la ligadura del SPSEM en el curso de la cirugía de desvascularización. Se practicó portografía y gastroendoscopia antes y después de la cirugía para estudios hemodinámicos y de seguimiento. Tres tipos de SPSEM fueron demostrados portográficamente, incluyendo 22 shunts portoumbilicales, 18 shunts espleno-renales y 2 shunts mesentérica inferior-cava. Cuarenta pacientes con SPSEM y sangrado por várices esofagogástricas fueron llevados a cirugía; en 26 se practicó ligadura del SPSEM y los 14 restantes constituyeron el grupo de no ligadura. Ni la presión portal preoperatoria ni la postoperatoria aparecieron significativamente diferentes entre los grupos con ligadura y sin ligadura ($p > 0.1$). Tampoco se observó diferencia significativa en cuanto a mortalidad quirúrgica, várices recurrentes y tasa acumulativa de supervivencia. Sin embargo, la portografía postoperatoria demostró drenaje persistente del flujo portal y perfusión intrahepática portal en los pacientes no ligados. Los signos clínicos de encefalopatía hepática cedieron luego de la ligadura del SPSEM en 3 pacientes. Puesto que algunos pacientes pueden beneficiarse en cuanto a la prevención de encefalopatía por el SPSEM, la ligadura del SPSEM es recomendada en el curso de desvascularización por várices esofagogástricas en pacientes cirróticos.

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