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Sugiura procedure for children with hemorrhagic portal hypertension secondary to extrahepatic portal vein occlusion

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Summary

From October 1990 to September 1994, eight children with the hemorrhagic extrahepatic portal hypertension (EPH) secondary to portal vein thrombosis (PVT) underwent the Sugiura operation in our department. All of the patients were male. The mean age was 7.4 years. The cause of PVT was idiopathic in 7 patients and due to polysplenia in one case. The Sugiura procedure (SP) was carried out in one stage via the thoracic route. The mean follow up period was 22 months. No patient had died and no rebleeding was found. We conclude that SP is the effective treatment for EPH caused by PVT.

Key words: *Portal hypertension, portal vein thrombosis, polysplenia, esophageal varices, bleeding, nonshunt operation, child*

Introduction

The portocaval shunting procedure for the surgical treatment of portal hypertension was first reported by Eck in 1877. This procedure was established as the standard decompressive method by Whipple and Blakemore in 1945 [13]. Subsequently portosystemic shunting has been adapted as the most effective method of controlling hemorrhage from bleeding varices. However, there has been a widespread belief that portal systemic encephalopathy and late hepatic failure occur in a significant proportion of patients with shunts [13,17].

In 1973 Sugiura and Futagawa [13] described a procedure consisting of transthoracic esophageal devascularisation, esophageal transection and reanastomosis, and paraesophagogastric devascularisation with splenectomy performed in one or two stages. From 1971 to 1977, another 191 patients have been treated with the same procedure by Sugiura and Futagava. They have achieved better results in controlling bleeding from esophageal varices without compromising hepatic functions and causing hepatic encephalopathy [14]. We report here our results on 8 children treated by the Sugiura procedure (SP).

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Material and Methods

From October 1990 to September 1994, eight patients with hemorrhagic portal hypertension were operated on with the Sugiura procedure (SP). The cause of the esophageal varices was extrahepatic portal hypertension (EPH) in 8 patients. The causes were confirmed by splenoportogram in 7 patients (Figure 1) and by histopathologic examination of the liver. The diagnosis of portal obstruction was made by ultrasound in one patient (polysplenic patient). Most patients received therapy before beign referred to us; this involved emergency and elective treatments to control bleeding. Emergency therapy to control variceal hemorrhage, consisting of intravenous vasopressin, and/or esophageal balloon tamponade; however, all patients had recurrence of variceal bleeding. The indications of operation were severe recurrent bleeding from esophageal varices in 6 patients and dangerous esophageal varices with hypersplenism in 2 patients whose platelet counts were below 70.000/mm^3. The number of variceal bleeding episodes ranged from 1 to 7 (median 3).

Nonshunt operation: We performed a one-stage transthoracic, transdiaphragmatic SP. A standard left lateral thoracotomy incision is made, and the in-

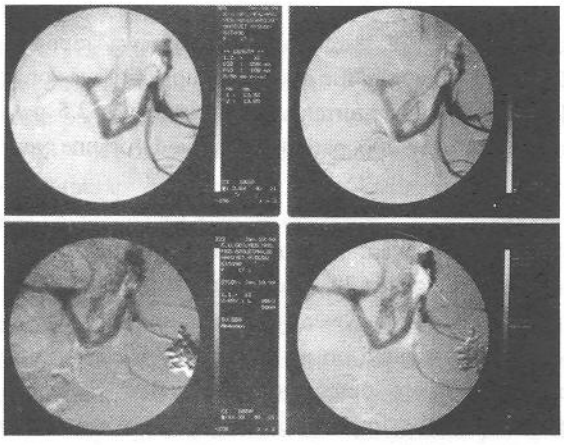

Figure 1. Portal hypertension with large portal vein (12 mm. in diameter), collaterals and large esophagogastric varices.

ferior mediastinum is opened. All of the dilated collateral veins were ligated and divided, with care taken not to damage the truncus vagalis. Upon completion of devascularization, the esophagus is doubly clamped with pairs of specially ordered esophageal clamps of the noncrushing type and esophageal transection is performed at the level of the diaphragm. The anterior muscular layer left intact.

The divided esophageal varices are not ligated because of the divided varices often cause postoperative stenosis. About 40-50 continued sutures of 4-0 prolein (Ethicon Co.) are placed and the divided varices are occluded with sutures. The muscle layer of esophageal anastomosis is completed, and a nasogastric tube is left in the stomach. Abdominal procedures were carried out via diaphragmatic route. After performing portal venography and manometry, splenectomy is carried out.

The abdominal esophagus and the cardia are devascularized from the greater curvature and the posterior of the stomach to the esophagus. Devascularization of the lesser curvature of the stomach and the abdominal esophagus is completed, and the cardioesophageal branches of the left gastric vessels are ligated and divided. The esophagus and the cardia are completely mobilized and freed from the adjacent structures. One pyloroplasty and two extramucosal pyloromyotomy were performed because of division of the gastric vagus nerves. A drain is inserted in the splenic cavity. All these patients were immunized with pneumococcal vaccine preoperatively, and benzatil penicillin has been prescribed for every month until 20 years of age.

Table I. Clinical data on patients who underwent the Sugiura procedure

Number of variceal bleeding episodes	
Median	3
Range	1-7
Blood transfusion (unit)	
Median	5.5
Range	1-8
Splenic pulp pressure (cm saline)	
Median	32.5
Range	29-38
Total bilirubin (mg/dl)	
Median	1.3
Range	08-1.7
Albumin (gr/dl)	
Median	4.2
Range	3.9-4.9
SGOT (IU/L)	
Median	29
Range	14-32
Alkaline phosphatase (IU/L)	
Median	67.5
Range	57-171

Results

The eight children with portal vein thrombosis (PVT) have been treated with SP. The mean age of the patients was 7.4 years and all the patients were male. The causes of EPH were PVT in 7 patients and polysplenia (18 spleen) in one. Liver function studies revealed the following values: albumin 4.2 (range 3.9-4.9) mg/dl, total bilirubin 1.3 (range 0.8-1.7) mg/dl, serum glutamic oxaloacetic transaminase (SGOT) 29 (range 14-32) IU/L, and alkaline phosphatase 67.5 (range 57-171) IU/L, splenic pulp pressure was 32.5 (range 29-38) cm in saline solution. No ascites was found in these patients (Table I). The patients were operated upon electively and SP was completed in one stage. The mean follow-up time was 22 months. To date, there have been no recurrent bleeding and no episodes of sepsis.

Discussion

The PVT consititutes about 7 percent of causes of portal hypertension in adults [2]. PVT is the main cause of portal hypertension in children. It is not possible to obtain any cause of PVT in the majority of cases [1]. Orloft et al reported that the cause of PVT was unknown in 68 %, neonatal omphalitis in

12 %, umbilical vein catheterization in 8 %, trauma in 4 %, and coagulopathy in 2 % [8]. In our cases, there were 7 idiopathic (unknown) PVT. There was a case whose EPH associated with polysplenia in this series. The polysplenia is usually associated with situs inversus, cardiac abnormalities and extrahepatic biliary atresia. There is no clear relation with pure polysplenia and EPH [1,4].

Controversy exists regarding the management of variceal bleeding secondary to PVT. Some authors have recommended only supportive measures during hemorrhagic episodes because bleeding is generally well tolerated by patients who are usually young and with good hepatic function [9]. However, hemorrhagic episodes tend to be repetetive and others have estimated that the mortality rate would be about 10 percent without specific treatment [16]. Similarly, one group has described nine patients with rebleeding out of series of 11 patients with hemorrhagic portal hypertension secondary to PVT who did not have any treatment [2]. Since the portacaval shunt procedure was established as the standard decompressive measure by Whipple and Blakemore in 1945 numerous shunt configurations have been developed [13].

Initial reports with surgical procedures showed disappointing results in the control of rebleeding. Grauer and Schwartz found 14 percent of rebleeding and PVT without hepatic disease who had nonselective shunts [7]. Voorhees et al found 13 percent of rebleeding in patients with PVT and mesocaval shunts, 44 percent with nonselective sphenorenal shunts and 48 percent with direct esophagogastric procedures [16]. In another study it was found that there was an incidence of rebleeding of 70 percent with nonselective splenorenal shunt and 75 percent with mesocaval shunts [18]. Some authors have reported no operative mortality, while others have found the mortality rate to be from 9 to 20 percent with nonselective shunts or direct esophagogastric procedure [2,3,7].

The Sugiura procedure was described in 1973. The essential element of the SP is agressive disconnection of the portal flow to the esophagus, namely, extensive devascularization around the distal esophagus, with esophageal transection and devascularization of the upper half of the stomach with splenectomy [13]. Sugiura and Futagawa reported 276 cases, in whom the operative mortality was only 4.3 percent and recurrence of varices was 2.5 percent with over 200 patients observed for one year [14].

All survivors were free of encephalopathy. In interpreting these results, one should note that 31 percent of the patients were noncirrhotic, only 52 patients being treated on an emergency basis, and 60 operations were done prophylactically. Futagawa emphasizes the importance of patient selection in avoiding a high operative mortality, recommending the procedure only for those patients with a total bilirubin value less than 5.0 mg and in whom medical control of ascites is not difficult. Orozco et al reported that fourty-five patients were treated with SP as a one stage or two stage procedure [9].

The operative mortality rate for the emergency thoracic operation was 41 percent and for the abdominal operation 42 percent. The overall operative mortality rate in the elective group was 10.8 percent. Encephalopathy developed in only one. Orozco and co-workers operated on 27 patients with hemorrhagic portal hypertension secondary to extrahepatic portal vein thrombosis without associated hepatic disease [10]. There was only one clinical encephalopathy and there were two cases of rebleeding and two deaths. One of the disadvantages with SP is that it generally must be done in two surgical stages to facilitate better tolerance of surgical trauma. However in young patients without hepatic disease, it may be possible to perform the complete procedure in only one operation in selected instances, as was done for nine patients without operative mortality or rebleeding [10]. If the trunks of the vagus are preserved carefully it is not necessary to add an antireflux procedure or a drainage measure such as pyloroplasty. In this series, one pyloroplasty and two extramucosal pyloromyotomy were performed because of vagal injuries.

Recently, endoscopic sclerotherapy has become a popular option in the therapeutic management of esophageal varices. However recurrent bleeding requiring repeated injections and the need for additional hospitalization and general anesthesia, es-

pecially in patients under 10 years of age are drawbacks. Eran et al reported 16 EPH patients who had bleeding oesophageal varices [6]. First they treated these patients with endoscopic sclerotherapy, but in 7 (43.7 %) cases, in spite of multiple attempts the sclerotherapy was ineffective and these patients had been managed by Sugiura operation.

They concluded that the sclerotherapy had been effective in the short term follow-up period, but as the follow-up period lasted longer, these patients needed surgical repair. Complications of sclerotherapy include esophageal stricture, ulceration, and perforation. A long-term risk of sclerosant use -esophageal carcinoma- has been reported. Moreover, rebleeding from esophageal varices after sclerotherapy is not rare (approximately 5 % to 30 %) [15,19]. For these reasons we have not performed endoscopic sclerotherapy in our EPH cases.

Susceptibility to severe sepsis, especially with encapsulated organisms, has been described after splenectomy [11]. The higher risk has been correlated with age, additional immunosuppressive factors, unplanned or incidental splenic ablation and multisystem illness or injury [5,12]. Benzatil penicillin has been prescribed to prevent infection for every month until 20 years. To date there have been no episodes of sepsis.

We conclude that the SP is an effective alternative operation for treatment of EPH caused by PVT and can be performed upon all patients without anatomic requirements.

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