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Non - Cirrhotic Portal Hypertension

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SUMMARY

During the year 1991-1992 eight patients presenting with variceal bleeding were referred to the Department of Cardiothoracic Surgery at the Shaikh Zayed Hospital, Lahore for shunt surgery. All cases were initially worked up in the Department of Gastroenterology and were found to have non-cirrhotic portal hypertension. Following surgical treatment, all patients are alive and well.

INTRODUCTION

Portal vein is formed by the union of superior mesenteric vein and splenic vein. It is 6-8 cm. long and 1.2 cm. in diameter. It has no valves and divides into right and left branches supplying the respective lobes of the liver. Portal vein blood flow is approximately 900-1200 ml/min. while hepatic artery flow is 500-700 ml/min. Portal vein carries 2/3rd of blood flow and 1/2 of oxygen delivery to the liver. Normal portal vein pressure is 5-10 mm. of Hg. Rise in the portal pressure above 12mm. of Hg. is labelled as portal hypertension. Hypertension within the portal vein and its tributaries may accompany hepatic disease or disturbance in the anatomy of the extra-hepatic vascular system. As a consequence of this elevated pressure, congestion of collateral pathways is established and may be manifested by oesophagogastric varices, splenomegaly, hypersplenism, ascites or encephalopathy. Portal hypertension due to non-cirrhotic conditions has well preserved liver function, no ascites or encephalopathy. The causes of portal hypertension may be classified as follows:

Presinusoidal (Extrahepatic)
a. Portal vein thrombosis
b. Extrinsic compression of portal vein
c. Nodes in porta hepatis
d. Cancer of the pancreas
e. Arteriovenous fistulae into portal system
f. Massive splenomegaly (occasionally)

Presinusoidal (Hepatic)
a. Congenital hepatic fibrosis
b. Sarcoidosis
c. Schistosomiasis
d. Alcoholic hepatitis
e. Precirrhotic primary biliary cirrhosis
f. Partial nodular transformation of the liver
g. Lymphomatous or other infiltrations of portal tract
h. Toxins (e.g. arsenic, vinyl chloride)

Sinusoidal
a. Cirrhosis
b. Diffuse nodular hyperplasia

Post-sinusoidal
a. Budd-Chiari syndrome
b. Hepatic veno-occlusive disease
c. Alcoholic central hyaline necrosis
d. Constrictive pericarditis
e. Cardiac temponde

PATIENTS AND METHODS

During year 1991-1992, all patients presenting with variceal bleeding were assessed for cause of portal hypertension in the Department of Gastroenterology at the Shaikh Zayed Hospital. Eight patients were diagnosed as having idiopathic non-cirrhotic portal hypertension (INCPH) on the basis of clinial and laboratory data supplemented by ultrasound and isotope liver spleen scan. One patient underwent pre-operative liver biopsy while two more had it done during the shunt surgery. Three patients were subjected to pre-operative splenoportogram.

The characteristics and presentation of these eight patients is given in Table 1.

Table 1:

Patient No.	Age (years)	Sex	Presentation
1.	12	F	Haemetemesis
2.	15	F	Haemetemesis and malaena
3.	17	M	Haemetemesis
4.	18	M	Haemetemesis and malaena
5.	15	F	Haemetemesis and malaena
6.	4 1/2	F	Haemetemesis and malaena
7.	23	F	Haemetemesis and malaena
8.	17	M	Haemetemesis and malaena

All patients had upper GI endoscopy by an experienced gastroenterologist, esophageal and/or gastric varices were sclerosed using 50% ethanol at several (2-5) sessions while awaiting shunt surgery.

The laboratory data of these patients are given in Table 2 and Table 3.

Table 2:

Patient No.	Hb (g/dl)	Platelets	Prothrombin time (sec)	Control time (sec)
1.	10.7	52,000	15.5	12
2.	10.0	83,000	14.3	12
3.	11.4	377,000	14.2	12
4.	12.3	67,000	11.6	11
5.	12.3	46,000	13.7	12
6.	8.8	158,000	11.8	11
7.	12.5	147,000	12.0	12
8.	12.2	150,000	13.0	12

All patients were referred to an experienced cardiothoracic surgeon who performed the shunt/devascularization surgery in these patients. The surgical procedures performed in our patients are listed in Table 4.

On pre-operative evaluation all patients had normal functioning liver as evident from normal liver function tests, normal ultrasound and isotope scans of the liver. One patient had a pre-operative percutaneous needle biopsy of the liver while two patients had a wedge biopsy of liver at operation. All these liver biopsies were reported as normal. Three patients had pre-operative, percutaneous splenoportogram, showing elevated splenic vein pressure, oesophagogastric varices and blocked portal vein.

Table 3:

Patient No.	Bilirubin (mg/dl)	Alk.Phos. (U/L)	SGPT (U/L)	Albumin (g/dl)
1.	0.7	365	21	4.2
2.	0.6	130	36	4.0
3.	0.9	306	16	4.5
4.	1.3	117	25	4.6
5.	1.5	100	18	4.4
6.	0.8	573	44	5.0
7.	0.7	64	21	4.2
8.	0.8	350	23	4.1

Normal values: Bilirubin, 0.2 to 1.0 mg/dl; Alk Phos, adult:64 to 306 U/L, Children, upto 644 U/L; SGPT, Upto 40 U/L; Albumin, 3.8 to 5.1g/dl.

Table 4:

Patient No.	Surgical Procedures
1.	Splencetomy and Proximal Splenorenal Shunt.
2.	Splenectomy and Proximal Splenorenal Shunt.
3.	Splenectomy and Proximal Splenorenal Shunt.
4.	Splenectomy and Proximal Splenorenal Shunt.
5.	Splenectomy and Proximal Splenorenal Shunt.
6.	Oesophageal Transection (Sigura Procedure).
7.	Splenorenal Shunt with Graft (Dacron)
8.	Warren Shunt.

Splenectomy was performed in five patients as they were symptomatic with massive splenomegaly, had features of hypersplenism and surgery was considered technically difficult without resorting to splenectomy. Patient no.6 had oesophageal transection and re-anastomosis as an emergency procedure for recurrent variceal bleeding, despite repeated sessions of endoscopic sclerotherapy and also because she was considered too young for shunt surgery. Patient no. 7 was found to have a congenital malformation of the splenic vein in the form of an aneurysmal sac related to it which was excised and a graft interposed between the splenic vein and the left renal vein.

RESULTS

All eight patients who underwent surgical treatment for non-cirrhotic portal hypertension are alive and well. They have been followed up for a period of 6-15 months. Although longer follow-up is needed to evaluate the results, short-term outlook has been excellent. In our series, the mean age of the patients was 15.1 years and they all had idiopathic portal vein thrombosis as the cause of portal hypertension. The post-operative course was uneventful except in one patient (patient no.6) who developed left sided empyema, requiring repeated chest aspirations and antibiotics. The average total hospital stay was 12.8 days and the average post-operative stay was 8.5 days.

DISCUSSION

Most of the reported work regarding selective or non-selective decompressive shunts for bleeding oesophagogastric varices has been carried out in patient with cirrhosis of liver. Although, all these trials show that survival is not increased, the incidence of rebleeding is greatly reduced following shunt surgery[1-5]. Warren and his colleagues have demonstrated that endoscopic sclerotherapy, despite a high rebleeding rate, gives better liver function and improved survival when compared with the distal splenorenal shunt[6]. The use of shunt surgery has been limited by the advent of liver transplantation, which is technically complicated in the presence of shunt, especially porta-caval shunt[7]. In view of these facts, the reported number of shunt operations in patients with cirrhosis has fallen dramatically in the last decade.

The role of shunt surgery in patients with non-cirrhotic portal hypertension has not been assessed as extensively as in cirrhosis because of the fact that the former condition is not so common in the western world. In our country non-cirrhotic portal hypertension is fairly common but the causative factors remain speculative. Neonatal umbilical cord sepsis leading to portal vein thrombosis may be an important aetiological factor. Exposure to certain tropical infections and toxins like arsenic and vinyl chloride have also been implicated but in 50% of cases no cause can be identified.

Non-cirrhotic portal hypertension is commonly seen in children and young adults. It often presents as upper GI bleeding from oeophagogastric varices. Splenomegaly and features of hypersplenism are commonly seen. The treatment in such patients is aimed at reducing portal pressure by employing one of the following treatment options:

a. Pharmacological agents like propranolol.
b. Endoscopic variceal sclerotherapy.
c. Shunt surgery or devascularization procedures.

The role of propranolol and other pharmacological agents in reducing portal pressure and preventing rebleeding remains controversial and this form of therapy cannot be generally recommended[8-13]. Based on present data, endoscopic variceal sclerotherapy seems to be an effective form of treatment for acute variceal bleeding and to prevent recurrent bleeding. There are however, early and late treatment failures and such patients should be identified and surgical treatment offered to them[14-19].

Shunt surgery in patients with non-cirrhotic portal hypertension is a safe procedure and long term survival figures of upto 100% have been reported[20]. Shunt failure is rare and can be managed by repeat surgery. Oesophageal transection with re-anastomosis and other devascularization operations are usually performed as emergency procedures in patients in whom bleeding cannot be controlled by any other measures. In such patients, however, operative mortality is high[19].

In Pakistan, facilities for endoscopic sclerotherapy are available only in larger towns and patients have to travel long distances to come to these centres. Thus, some of our patients bleed to death before reaching hospitals for the same reason. Because of these reasons we recommend shunt surgery to all our patients with non-cirrhotic portal hypertension and as already mentioned the results of surgery in these patients are excellent. Those who refuse surgery, long-term injection sclerotherapy is continued until the varices are obliterated and surveillance endoscopy is planned on 6 monthly basis. Indian studies also show good long term survival and better outcome after shunt surgery[21,22].

In conclusion, we believe that although endoscopic variceal sclerotherapy remains the initial treatment of choice for all forms of variceal bleeding, we recommend early shunt surgery for patients with non-cirrhotic portal hypertension because of the delay involved in their presentation to a centre with emergency endoscopic variceal sclerotherapy service

following an episode of G.I bleeding. Moreover, the results of surgery in this group of patients are excellent.

REFERENCES

1. **Rikkers LF, Burnett DA, Volentine GD, Buchi KN, Cornier RA.** Shunt surgery vs. endoscopic sclerotherapy for long term treatment of variceal bleeding. Early results of a randomized trial. *Ann Surg 1987;* **206**: 261-71.
2. **Teres J, Bordas JM, Bravo D, et al.** Sclerotherapy versus distal splenorenal shunt in the elective treatment of variceal hemorrhage: a randomized controlled trial. *Hepatology 1987;* **7**: 430-6.
3. **Paquet K-J, Kalk J-F, Koussouris P.** Prospective evaluation and long term results of mesocaval interposition shunts. *Acta Chir Scand 1987;* **153**: 423-9.
4. **Paquet K-J, Mercado MA, Kalk J-F, Koussouris P, Siemens F, Muting D.** Analysis of a prospective series of 100 mesocaval interposition shunt for bleeding portal hypertension. *Hepatogastroenterology 1990;* **37**: 115-20.
5. **Cello JP,Crass R, Trunkey DD.** Endoscopic sclerotherapy vs. esophageal transection of Child class C patients with variceal hemorrhage. Comparison with results of portacaval shunt: preliminary report. *Surgery 1982;* **91**: 333-8.
6. **Warren WD, Henderson JM, Millikan WJ, et al.** Distal splenorenal shunt versus endoscopic sclerotherapy for long-term management of variceal bleeding. Preliminary report of a prospective randomized trial. *Ann Surg 1986;* **203**: 454-62.
7. **Brems JJ, Hiatt JR, Klein AS, et al.** Effect of a prior portasystemic shunt on subsequent liver transplantation. *Ann Surg 1989;* **209**: 51-6.
8. **Lebrec D, Novel O, Corbic M, et al.** Propranolol:a medical treatment for portal hypertension. *Lancet 1980;* **2**. 180-2.
9. **Sheen IS, Chen TY, Liaw YF.** Randomized controlled study of propranolol for prevention of recurrent esophageal varices bleeding in patients with cirrhosis. *Liver 1989;* **9**: 1-5.
10. **Burroughs AK, Jenkins WJ, Sherlock S, et al.** Controlled trial of propranolol for the prevention of recurrent variceal hemmorrhage in patients with cirrhosis. *N Engl J Med 1983;* **309**: 1539-42.
11. **Villeneuve JP, Pomier-Layrargues G. Infante-Rivarde C, et al.** Propranolol for the prevention of recurrent variceal hemmorhage: a controlled trial. *Hepatology 1986;* **6**: 1239-43.
12. **Queuniet AM, Czernichow P, Lerebours E et al.** Controlled trial of propranolol for the prevention of recurrent gastrointestinal bleeding in patients with cirrhosis. *Gatroenterol Clin Biol 1987;* **11**: 11-7
13. **Garden OJ, Mills PR, Birnie GG, et al.** Propranolol in the prevention of recurrent variceal hemorrhage in cirrhotic patients. A controlled trial. *Gastroenterology 1990;* **98**: 185-90.
14. **Terblanche J, Bornman PC, Kahn D, et al.** Failure of repeated injection sclerotherapy to improve long-term survival after oesophageal variceal bleeding: a five-year prospective controlled clinical trial. *Lancet 1983;* **2**: 1328-32.
15. **The Copenhagen Esophageal Varices and Sclerotherapy Project.** Sclerotherapy after first variceal hemorrhage in cirrhosis: a randomized multi-center trial. *N Engl J Med 1984;* **311**: 1594-600.
16. **Westaby D, Macdougall BR, Williams R.** Improved Survival following injection sclerotherapy for esophageal varices: final analysis of a controlled trial. *Hepatology 1985;* **5**: 827-30.
17. **Korula J, Balart LA, Radvan G, et al.** A prospective, randomized controlled trial of chronic esophageal variceal sclerotherapy. *Hepatology 1985;* **5**: 584-9.
18. **Soderlund C, Ihre T.** Endoscopic sclerotherapy v. conservative management of bleeding oesophageal varices: a 5-year prospective controlled trial of emergency and longterm treatment. *Acta Chir Scand 1985;* **151**: 449-56.
19. **Paquet K-J, Mercado MA, Gad HA.** Surgical procedures for bleeding esophagogastric varices when sclerotherapy fails: a study. *Am J Surg 1990;* **160**: 43-7.
20. **Myburgh JA.** Selective shunts: Johannesburg experience. *Am J Surg 1990;* **160**: 67-74.
21. **Pande GK, Reddy VM, Kar P, et al.** Operations for portal hypertension due to extrahepatic obstruction. Results and 10 years follow up. *Br Med J 1987;* **29**: 1115-7.
22. **Mitra SK, Kumar V, Datta DV, et al.** Extrahepatic portal obstruction. A review of 70 cases. *J Pediatr Surg 1978;* **13**: 51-4.

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