



OUTCOMES OF PROXIMAL SPLENORENAL SHUNT SURGERY IN EXTRA HEPATIC PORTAL VEIN OBSTRUCTION (EHPVO) – EXPERIENCE FROM A TERTIARY CENTER IN GUJARAT, INDIA.

Hepatobiliary Surgery

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ABSTRACT

Background – Extra hepatic portal vein obstruction commonly presents with variceal bleeding and splenomegaly. Management with endoscopic means provide temporary palliation. A proximal splenorenal shunt is a definitive procedure with good results. Our aim was to evaluate the outcomes of splenectomy with proximal splenorenal shunt in patients with extrahepatic portal venous obstruction.

Methods – The records of all patients with diagnosis of EHPVO who were operated for splenectomy with proximal splenorenal shunt during the period from January 2014 to august 2018 were analysed. Total of 34 patients with EHPVO were operated for splenectomy with proximal splenorenal shunt over the study period. Outcomes were evaluated in term of rebleeding, variceal regression shunt thrombosis and post shunt encephalopathy.

Results – All the patients were below 30 years age. Shunt surgery lead to reversal of various degree of cytopenias and hypersplenism. Variceal regression was seen from large size (grade III and IV) to small size (grade II) in 74% of patients. We had low incidence of shunt block (5.88%), variceal rebleeding (2.94%) and no mortality. None of the patients developed encephalopathy during follow-up.

Conclusion- Splenectomy with proximal splenorenal shunt is a single time safe procedure, with low morbidity and mortality; having low rates of variceal rebleeding and encephalopathy; with low cost in teaching hospital, thus can be considered as the main treatment approach in patients with EHPVO.

KEYWORDS

extrahepatic portal vein obstruction, proximal splenorenal shunt

INTRODUCTION

As per the APASL consensus, EHPVO is defined as "a vascular disorder of liver, characterized by obstruction of the extra-hepatic portal vein with or without involvement of intra-hepatic portal vein radicles or splenic or superior mesenteric veins".¹ Replacement of the extrahepatic portal vein by a cavernoma with or without thrombosis of the intrahepatic portal vein, splenic vein, or superior mesenteric vein is seen in EHPVO.

Variceal bleeding in EHPVO usually occurs in the first or second decade of life². Well tolerated variceal bleed and splenomegaly are common presentations of EHPVO. Growth retardation can be seen in children. Lykavieris et al²³ reported mortality of 59% without effective treatment of portal hypertension secondary to EHPVO, thus suggesting the need of potential treatment in this patient group.

Acute variceal hemorrhage episodes are treated by endoscopic management along with pharmacotherapy support. But has a recurrence rate of up to 40% in some studies³. Surgical options for these patients can be divided into portosystemic shunts or esophagogastric devascularization with splenectomy. In presence of shuntable veins and surgical expertise, shunt surgery is preferred in patients with EHPVO. This improves growth recovery and reduces development of gastric and ectopic varices.^{1,4,5} In present study, we aimed to evaluate the outcomes of patients undergoing splenectomy with proximal splenorenal shunt for EHPVO.

MATERIALS AND METHODS

This study is a retrospective case series of patients with portal hypertension diagnosed to have EHPVO in the surgical gastroenterology department at NHL Medical College, Ahmedabad. The records of all patients with diagnosis of EHPVO who were operated for splenectomy with proximal splenorenal shunt during the

period from January 2014 to august 2018 were reviewed. Total of 34 patients with EHPVO were managed over the study period.

Diagnostic criteria used for EHPVO included clinical signs of portal hypertension like gastrointestinal bleed in presence of splenomegaly, presence of portal cavernoma on doppler ultrasound , exclusion of chronic liver disease, exclusion of other causes of non-cirrhotic portal hypertension along with exclusion of cirrhosis on per operative liver biopsy. Paediatric age group patients under 12 years of age were screened with serum ceruloplasmin and 24 hour urine copper excretion levels along with ophthalmic examination for Kayser–Fleischer rings. Esophageal varices were documented on upper GI endoscopy and were graded according to Paquet's classification⁶ Grade I- Microcapillaries located in distal esophagus or oesophago-gastric junction. Grade II- One or two small varices located in the distal esophagus. Grade III- Medium-sized varices of any number. Grade IV - Large-sized varices in any part of the esophagus.

Indication for surgery was recurrent variceal hemorrhage. All patients underwent splenectomy with proximal splenorenal shunt. Under general anesthesia left subcostal incision was made extending into the left flank and/or upper midline. Liver was examined for any evidence of macroscopic cirrhosis. Spleen was dissected from all the collateral veins and diaphragm, splenic vein is then dissected at the splenic hilum and splenectomy done.

Retropancreatic segment of the splenic vein was meticulously dissected and mobilized. Left renal vein identified and dissected for adequate length and the patient was heparinized with 5000 units of heparin. Side occluding satinsky vascular clamp was applied to the left renal vein. Venotomy made in the left renal vein and end to side anastomosis done using 7-0 prolene as shown in image number 1. Operation was completed with one abdominal tube drain in the splenic bed.

All the patients were followed up at an interval of three months during the first year & six months in the second year. During follow up, we performed routine haemogram, liver function test and Doppler ultrasound to assess the shunt patency, UGIE to assess variceal regression. The long-term outcome parameters were measured and compared with EHPVO related bleed, decrement in grade of varices, shunt related complications (encephalopathy), and shunt patency. Statistical analysis – All data was entered into Microsoft excel worksheet. The results were tabulated for better presentation.

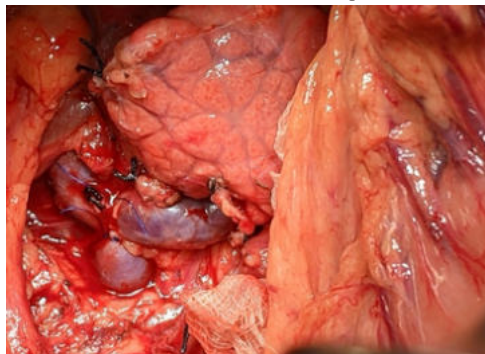


Image number 1 – Showing PSRS anastomosis

RESULTS

A total of 34 patients were included in the analysis. All the patients were below 30 years. Average duration of symptoms was 5.15 +/- 2.52 years. The most common presenting symptom was upper gastrointestinal bleed manifesting as hematemesis. The clinical, laboratory, imaging and endoscopic parameters are detailed in Table 1.

Table number 1 – Showing Clinical Characteristics of patients with EHPVO

Patients clinical characteristics	Values
Total Patients	34
Age (Mean± SD)	16.70 +/- 6.53 years
Age distribution	
<10-years	7 (20.58%)
11–20 years	16(47.05%)
21–30 years	11 (32.355)
Above 30 years	None
Gender	
Male	19 (54.9%)
Female	15 (45.09%)
Clinical presentation (%)	No. Of Patients n
Hematemesis	34 (100 %)
Melena	4 (11.7 %)
Awareness Of Abdominal Lump	22 (64.70 %)
Pain Abdomen	8 (23.52 %)
Palpable lump (Splenomegaly)	34 (100 %)
Jaundice	1 (2.94%)
Hepatic Encephalopathy	None
Preopaerative Investigations	Mean + /- SD
Haemoglobin(gm/dl)	8.82 +/- 0.92
WBC (cells/mm ³)	4.16+/- 1.09
Platelets (cells/mm ³)	0.76+/-0.21
Total Bilirubin(mg/dl)	0.97 +/- 0.30
SGOT(IU/L)	43.39 +/-10.67
SGPT (IU/L)	44.67 +/- 15.54
ALP(IU/L)	69.44 +/- 55.75
S.Albumin(gm/dl)	3.59 +/- 0.36
INR	1.14 +/- 0.12
Imaging Characteristics	Value
Spleen size	16.59 cm
Largest	23 cm
Smallest	15.2 cm
Splenic vein at hilum	9.82 +/-2.20 mm
Left renal vein	9.61 +/- 1.25 mm
Ascites	None
Esophageal Varices Grading	N (%)
Grade I	None
Grade II	3 (8.82%)
Grade III	19 (55.88%)
Grade IV	12 (35.29%)

Varying degrees of cytopenias suggesting hypersplenism were observed with presence of normal liver functions. In all patients on imaging there was cavernomatous transformation of portal vein, normal liver parenchymal echotexture without any typical cirrhotic changes and patent hepatic veins in all patients. Table number 2 shows intraoperative and postoperative outcomes.

Table number 2 – Intra Operative outcomes and Postoperative outcomes

Parameters	N(%)
Grossly non – cirrhotic liver surface	100 %
Shunt Diameter (mm) (mean +/- SD)	9.26 +/- 1.08
Operative time (min) Median (Range)	243 +/- 17
Blood loss (ml) Median (Range)	210 +/- 36
Hospital stay (days) Median (Range)	7 (6 -11)
SSI	3 (8.82%)
Fever	2 (5.88 %)
Cardio Respiratory Complication	1 (2.94%)
Hepatic Encephalopathy	None
Mortality	None

All the patients were in follow up for a median period of 2 years (range 1-5 years). During follow up, two patients (5.88 %) had shunt blockage and one of them developed rebleeding (2.94%) after shunt surgery, which was managed conservatively. Decrement in variceal grade IV was noted from 35.29% to 5.88 % and in grade III from 55.88% to 17.64%. Table 3 shows detail follow up outcomes.

Table number 3 – Showing outcomes at follow up

Parameters	N(%)
Variceal Rebleeding	1 (2.94%)
Variceal Regression	
Grade I	None
Grade II	26 (76.47%)
Grade III	6 (17.64%)
Grade IV	2 (5.88 %)
Status of Shunt	
Patent	32(94.11%)
Thrombosed	2 (5.88 %)
Hemoglobin(gm/dl)	12.44 +/- 1.24
WBC (cells/mm ³)	6.88 +/- 1.33
Platelet (cells/mm ³)	2.54 +/- 0.71
Encephalopathy	None

DISCUSSION

EHPVO is a common cause of portal hypertension and variceal bleeding in children and young adults. The etiology of EHPVO is largely idiopathic with omphalitis, umbilical vein catheterization and intra-abdominal sepsis responsible in some of the cases¹. Lykavieris et al²³ reported the risk of variceal bleeding increases from 49% at 16 years to 76% at 24 years and mortality of 59% after mean follow up of 8 years(range 3-22 years) without effective treatment of portal hypertension secondary to EHPVO, thus suggesting the need of potential treatment in this patient group.

Portosystemic shunts and esophagogastric devascularization are surgical options for EHPVO. Shunt surgery is indicated in a group of patients where variceal bleeding fails to respond to endoscopic management. At times shunt surgery can also be considered for symptomatic hypersplenism and portal biliopathy^(4,8). Portosystemic shunts divert blood flow from the high pressure portal circulation to low pressure systemic circulation. Proximal splenorenal shunt, side to side splenorenal shunt, distal splenorenal shunt and mesocaval shunt are various types of portosystemic shunts.

Since 1947, being first reported by Dr. Robert Linton⁹ the PSRS with splenectomy has become a established one time procedure in EHPVO patients that prevents variceal bleeding with rebleeding rate of 0–2%, no mortality and no encephalopathy in the postoperative period, and low incidence of post splenectomy infection^{7,8,9}. Ramamurthy et al¹⁰, showed 80% regression to obliterated varices from grade III after PSRS, whereas in our study also comparable variceal regression was seen from large size (grade III and IV) to small size (grade II) in 74% of patients. Saluja et al¹⁸ also reported significantly better decrement in grade of varices in patients undergoing PSRS 68.7% than splenectomy 33.3%.

PSRS is also more effective in reducing rebleeding rates compared to

devascularization as reported by Mukund et al¹⁹, Ramamurthy et al¹⁰ (rebleeding rate of 17% for devascularization plus endotherapy and zero after PSRS). Finally shunt surgery also performs better than sclerotherapy, as Wani et al²⁰ reported rebleeding rates of 22.6% in the sclerotherapy group and 3.3% in the shunt surgery group.

During the follow-up after PSRS surgery shunt patency was 94.11% in our study which was 78.5% in a study by Sharma et al¹⁴, 90% in study by Rao KL et al¹⁵, 93% by Bismuth et al¹⁶, 97.3% by Agarwal AK et al.¹³ The size of the splenic vein has a significant impact on the shunt patency rate, the diameter of vein should be at least 10 mm for a shunt to be effective and remain patent^{21,22} which was 9.82 mm (mean splenic vein diameter) in our study. Additionally imaging modality also significantly influences shunt patency rates. In a study by Pk Mishra et al¹⁷ overall shunt patency rates were 70%, 40% and 60% using clinical parameters, color Doppler and dynamic CT portography respectively. They noted superiority of dynamic CT to UGIE and ultrasound doppler study to visualize shunt patency. We did not use CT portography because of the cost factor, a limitation of our study.

Good outcomes are possible for Splenectomy with PSRS in hands of experienced surgeons having adequate understanding of surgical anatomy and dissection expertise even without availability of advanced haemostatic equipment. Performing PSRS with good shuntable vein (sufficient size and thrombus free vein) and strictly avoiding patients with possibility of liver cirrhosis helped us to achieve good outcomes like low incidence of shunt block (5.88 %), variceal rebleeding (2.94%) and no mortality. Also we did not come across any case developing hepatic encephalopathy or post splenectomy sepsis after splenectomy with proximal splenorenal shunt in our case series.

Additionally Splenectomy with PSRS is also effective in relieving any associated hypersplenism, portal hypertensive gastropathy, portal biliopathy and improved quality of life in majority of patients and it does not need any use of natural or synthetic grafts unlike mesocaval shunt.^{3,11,12}

CONCLUSION

Splenectomy with proximal splenorenal shunt is a single time safe procedure, with low morbidity and mortality; having low rates of variceal rebleeding and encephalopathy; with low cost in teaching hospital, hence, we suggest splenectomy with proximal splenorenal shunt should be considered as the main treatment approach in patients with EHPVO.

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