

## Surgical management of extrahepatic portal vein obstruction at University Hospital

Shah S, Ghimire B, Kansakar P B S, Bhandari R S, Lakhey P J, Vaidya P, Singh Y P

Department of GI and General Surgery, Tribhuvan University Teaching Hospital, Institute of Medicine, Manarajgunj, Nepal

**Correspondence:** Dr. Yogendra P. Singh, Professor and Head, Department of GI and General Surgery

**E-mail:** yp\_singh@hotmail.com

### Abstract

**Introduction:** Extrahepatic portal vein obstruction (EHPVO) is a second most common cause of portal hypertension which causes upper gastrointestinal (GI) bleeding. Primary management of upper GI bleeding is endoscopic therapy. However, surgery is performed as a secondary management of upper GI bleeding and if patients fail to respond endoscopic management or complications of EHPVO develops. The aim of the study was to determine the perioperative outcome of surgery done for EHPVO.

**Methods:** This is retrospective observational study of all the patients of EHPVO, who were undergone surgical management at Tribhuvan University Teaching Hospital in between April 2015 to March 2017. Data were collected from case sheets of the patients. The demographic and clinical characteristics of the patients, and perioperative and short term outcome of the surgical management of EHPVO patients were analysed.

**Results:** Total 34 patients were included in the study including 20 males (58.8%) and 14 (41.2%) females with median age of 17 years (4 to 45 years). Most common presentation of EHPVO were fullness in upper abdomen (34/34) upper GI bleeding (29/34). Most of the patients had anemia (33/34), splenomegaly (34/34) and hypersplenism (28/34). Shunt surgery was performed in 20 patients and modified Hassab's procedure in 14 patients. There was no post-operative severe complication except one mortality in Hassab's procedure. The median duration of surgery was higher in shunt surgery group compared to devascularisation group (240 minutes versus 180 minutes). There were no significant differences in intraoperative blood loss, total hospital stays and recurrent upper GI bleeding in both surgery groups.

**Conclusions:** Surgical management for EHPVO have good perioperative and short term outcome.

**Keywords:** Esophageal varices, Hypersplenism, Portal biliopathy, Portal vein obstruction

### Introduction

Extrahepatic portal vein obstruction (EHPVO) is the second common cause of the portal hypertension after cirrhosis. The causes of EHPVO are unknown in most of the cases. The etiology and clinical presentation between children and adults are different.(1,2) Patients with EHPVO most commonly presented with upper gastrointestinal (GI) bleeding and splenomegaly.(3) In 1960's, surgery was the only option available thereafter development of endoscopic sclerotherapy totally replaced surgery.(4) In 1990's interest in portosystemic

shunt has been ushered again.(5) Recently, surgery is primarily indicated in cases of variceal bleeding secondary to EHPVO refractory to medical/endoscopic management and/or EHPVO related complications like hypersplenism and portal biliopathy.(6) Shunt surgery is not possible for 10% to 15% of patients with EHPVO due to non-shuntable vein or thrombosed splenoportal and mesentericoportal axis. In these cases, splenectomy with esophagogastric devascularisation procedures for example, modified Hassab's procedure can decrease blood flow in the gastrosplenic region and enhances

liver perfusion. Thus, whenever shunt surgery is not feasible, devascularisation remains an effective surgical options in the management of variceal bleeding to prevent re-bleeding.(7,8)We reviewed our experience with surgical management of EHPVO in Tribhuvan University Teaching Hospital (TUTH), a tertiary care center in Nepal. The main objective of this study was to analyse and describe perioperative and short term (at least 6 months) postoperative outcomes of EHPVO patients after surgical management.

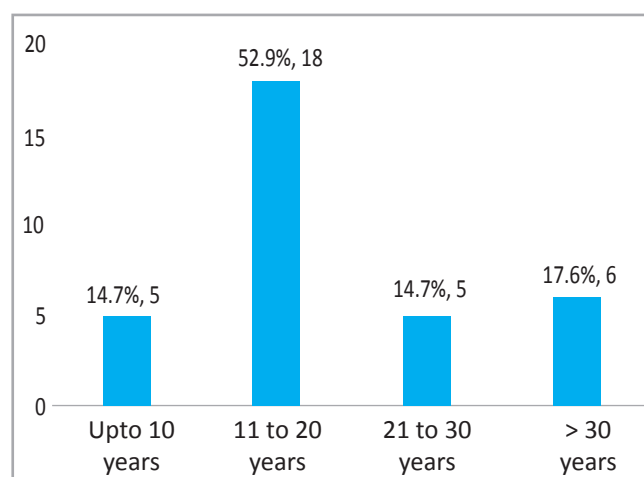
## Methods

This was a retrospective descriptive study conducted at Tribhuvan University Teaching Hospital (TUTH) from April 2015 to March 2017. All patients with EHPVO who had undergone surgical management at TUTH were included in the study. Data of patients demographic, clinical course, investigations, type of surgical management and perioperative outcome were retrieved from case-sheets available at medical record section and analysed. Those patients who had not undergone surgical management were excluded from the study. Preoperative diagnosis was confirmed with ultrasound abdomen and Doppler study of the portal system to look for portal venous system, cavernous transformation of portal vein, pattern of portal blood flow (hepatopedal or hepatofugal), status of liver and spleen. Preoperative computed tomographic portography were performed in all cases to assess splenic vein, extend of thrombosis or obliteration of portal venous system, left renal vein, and status of the kidney. Hematological and biochemical blood tests were checked preoperatively to assess the general status of the patients. Categorical variables were presented in frequency and percentage (proportion) and quantitative variable were presented in median (range). Differences between different surgical management groups were analysed using Chi-square test or Fisher exact test for categorical variable and student t-test or Mann-Whitney U-test for continuous variables. Statistical analysis was performed by using IBM SPSS statistics version 23.

## Results

There were 34 patients of EHPVO who had undergone surgical management at TUTH during study period. The median age of the patients was 17 years (ranges from 4 to 45 years). Most of the patients with EHPVO were in second decades of their life (Figure 1). The

EHPVO was predominantly present in males (20/34) compared to females (14/34). Patients with EHPVO were commonly presented with abdominal fullness (34/34) and upper gastrointestinal bleeding (29/34). The most common physical examination findings were anemia (33/34) and splenomegaly (Figure 2). The esophageal varices were present in 88% (30/34) patients and grade III and/or above esophageal varices were present in 70.6% (24/34). The rectal varices were present in 14.7% (5/34) patients (Figure 3). Portal biliopathy were present in 55.9% (19/34) patients. But symptomatic biliopathy was only 17.6% (6/34) patients. The portosystemic shunt surgeries (proximal splenorenal shunt with splenectomy in 19 patients, and proximal spleno-adrenal vein shunt with splenectomy in one patient) were performed in 58.8% (20/34) and modified Hassab's procedure in 41.2% (14/34) patients. The overall median duration of surgery, intraoperative blood loss, and hospital stay was 210 minutes, 300 ml and 10 days respectively. Among 34 patients, two patients presented with recurrent upper GI bleeding, one had undergone proximal splenorenal shunt with splenectomy and one had undergone modified Hassab's procedure. Recurrent upper GI bleeding was managed with endoscopic variceal band ligation. There was one mortality due to postoperative cytopenia and sepsis who had undergone modified Hassab's procedure. Portal biliopathy was significantly common in patients who were undergone modified Hassab's procedure (Table 1). The duration of surgery was significantly longer in patients who were undergone portosystemic shunt compared to the patients who was undergone modified Hassab's procedure (Table 2).



**Figure 1: Distribution of patients according to age groups.**

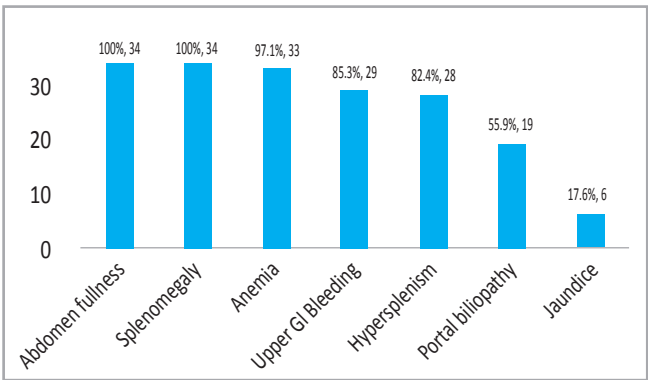


Figure 2: Clinical characteristics of the patients.

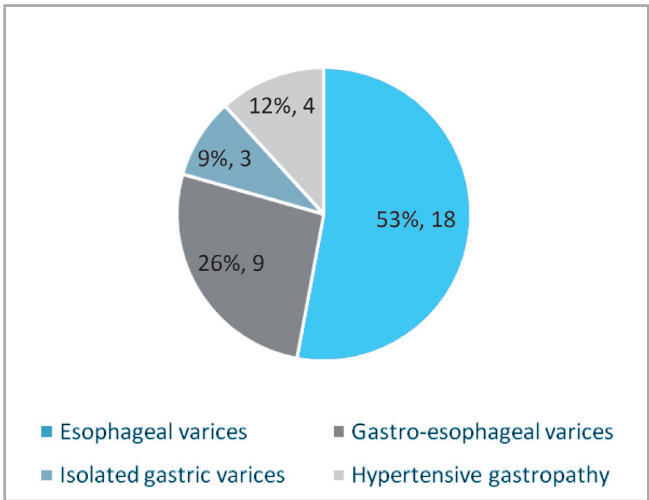


Figure 3: Type of Varices in cases of EHPVO.

Table 1: Differences in demographic, clinical characteristics among both surgical management groups.

| Characteristics                | Portosystemic shunt (n=20) | Modified Hassab's procedure (n=14) | p-value |
|--------------------------------|----------------------------|------------------------------------|---------|
| Age, median (range) in years   | 17 (5-35)                  | 19 (4-45) years                    | NS      |
| Gender                         |                            |                                    | NS      |
| Male (number)                  | 14 (70%)                   | 6 (30%)                            |         |
| Female (number)                | 6 (43%)                    | 8 (57%)                            |         |
| Upper GI bleed (number)        | 15 (75%)                   | 14 (100%)                          | NS      |
| Abdominal fullness (number)    | 20 (100%)                  | 14 (100%)                          | NS      |
| Jaundice (number)              | 3 (15%)                    | 3 (21.4%)                          | NS      |
| Splenomegaly (number)          | 20 (100%)                  | 14 (100%)                          | NS      |
| Anemia (number)                | 19 (95%)                   | 14 (100%)                          | NS      |
| Hypersplenism(number)          | 17 (85%)                   | 11 (79%)                           | NS      |
| Portal biliopathy(number)      | 7 (35%)                    | 12 (85.7%)                         | 0.005   |
| Varices (≥ Grade III) (number) | 12 (60%)                   | 12 (86%)                           | NS      |

p-value <0.05 is significant.

NS = Non-significant.

Table 2 Differences in perioperative and short term postoperative outcomes among both surgical management groups.

| Characteristics                     | Portosystemic shunt (n=20) | Modified Hassab's procedure (n=14) | p-value |
|-------------------------------------|----------------------------|------------------------------------|---------|
| Duration of surgery, median (range) | 240 (165-480) minutes      | 180 (105-255) minutes              | 0.003   |
| Intraop blood loss, median (range)  | 300 (100-400) ml           | 325 (100-650) ml                   | NS      |
| Hospital stay, median (range)       | 10 (6-34) days             | 9.5 (4-30) days                    | NS      |
| Recurrent UGI bleeding, number      | 1 (5%)                     | 1 (7%)                             | NS      |

p-value <0.05 is significant.

Intraop = Intraoperative, NS = Non-significant, UGI = upper gastrointestinal bleeding.

## Discussion

Extrahepatic portal venous obstruction is defined as obstruction of extrahepatic portal vein with or without involvement of the intrahepatic portal vein or other segments of the splanchnic venous axis. It does not include isolated thrombosis of the splenic vein or superior mesenteric vein.(9) Although EHPVO is not common in developed countries. It is responsible for about 54% of overall cases of portal hypertension in developing countries and upper GI bleeding in 68% to 84% patients.(10) EHPVO is common in childhood however it may present in adulthood. In study by Shah Syed Khalid et al, median age of EHPVO patients was 12 years (ranges from 3 to 50 years) and majority of patients were less than 20 years with males (70%) predominant.(11) In our study, median age of the patients with EHPVO was 17 years (ranges from 4 to 45 years) and majority were age of less than 20 years (67.6%) with male predominant (58.8%).

Clinical presentation depends on the onset and extent of the obstruction and development of collateral circulation. Common clinical presentation of EHPVO was upper GI bleeding (85% to 90%) splenomegaly (88%) and hypersplenism (28%). There were 90% of grade III varices, and rectal varices in 24% patients. If a young child has upper GI bleeding and splenomegaly in a normal liver function, the diagnoses of EHPVO is most likely and needs to be excluded. (12,13) In our study, upper GI bleeding was present in 85.3%, and splenomegaly in 100%. As splenomegaly was present in all cases in our study, abdominal fullness was present in all cases and hypersplenism in 82.4% of patients. Grade III or above varices were present in 70.6% of patients in our study.

Portal biliopathy is defined as anatomical and functional alterations of the intra-/extra-hepatic bile ducts in patients with portal hypertension due to EHPVO. Portal biliopathy developed as a result of compression and ischaemia of bile duct due to dilated venous collaterals around the bile duct originating from paracholedochal venous plexus of Saint and pericholedochal venous plexus of Petren. The change in portal biliopathy includes indentation in bile duct due to collaterals, strictures, angulations, focal narrowing, stones, and irregular walls. Portal biliopathy is recognized in 90% to 100% of the cases, however only few patients are symptomatic, usually in the adult age groups and reflects advance disease which is regressed after lowering portal pressure surgically or

radiologically.(14,15) Portal biliopathy was present in 55.9% of patients in our study, however, only 17.6% were symptomatic presented with jaundice.

Ultrasound abdomen allows to assess the echogenicity and size of liver and spleen, presence of cavernous transformation of the portal vein, and patency of portal and splenic veins and to determine the direction of blood flow. Computed tomography (CT) and magnetic resonance imaging is used to assess the anatomy of the portal system where surgery is considered. Shunt patency can be evaluated using clinical parameters, color Doppler or dynamic CT portography.(16,17) In our study, we also used ultrasound/color Doppler to diagnose the cases of EHPVO and CT portography before surgery.

Endoscopic therapy is indicated for primary prophylaxis and when there are no veins available for shunting due to extensive thrombosis or too small veins and patients not fit for shunt surgery. Shunt surgery is preferred as a secondary prophylaxis as a one-time treatment options to avoid repeated follow-up and morbidity associated with it.(18) In our study, patient with acute upper GI bleeding were managed endoscopically and surgery were performed as a secondary prophylaxis. Surgery was indicated due to hypersplenism with enlarged spleen in 22 (64.7%), portal biliopathy with enlarged spleen in 6 (17.6%) and failed endoscopic therapy to control variceal bleeding in 6 (17.6%) patients.

There are significantly lower re-bleeding rates (3.3% versus 22.6%), less treatment failure (6.7% versus 19.4%) and good quality of life after shunt surgery than that after endoscopy therapy. The re-bleeding after shunt surgery is usually easily managed with endoscopy therapy.(19,20) In our study 2 patients had re-bleeding after surgery for EHPVO which was managed with endoscopic therapy.

Surgical options are broadly divided into portosystemic shunt, esophagogastric devascularisation and more recently Rex shunt. Options for portosystemic shunts includes proximal splenorenal shunt (PSRS), side to side splenorenal shunt, distal splenorenal shunt (DSRS) and mesocaval shunt. DSRS is preferred in cirrhotic patients to minimize encephalopathy. However, it does not offer any advantage in EHPVO as encephalopathy does not occur even after non-selective portosystemic shunts. Though DSRS and side to side splenorenal shunt preserve spleen, hypersplenism may take a long time to reverse or may not reverse at all; and it is also not

suitable for patients with massively enlarged spleen. A mesocaval shunt usually requires the use of a vascular conduit and has high rates of shunt thrombosis.(21,22) Rex shunt (mesenterico left portal vein bypass (MLPVB)) is more physiological shunt for EHPVO which increases hepatic blood flow thus improves hepatic functions and coagulation. But it requires the presence of a patent superior mesenteric vein, intrahepatic left portal vein, and internal jugular vein. However, Rex shunt is not popularised in developing countries due to extensive involvement of portal venous system.(23) In our study, all the patients had enlarged spleen with hypersplenism in 82.4% and left portal vein was not patent. Thus, DSRS, side to side splenorenal shunt or Rex shunt were not suitable in our study.

PSRS is the most commonly performed shunt with splenectomy which take care of hypersplenism, prevents bleeding from hypertensive gastropathy/enteropathy. (17,24) PSRS has a good long term results with 15 years survival of >96%, re-bleeding rate of 2% to 11%, shunt patency rate 90% to 95%, without development of encephalopathy, and mortality <2%. PSRS divert blood flow from portal system to systemic circulation to decrease portal pressure as well as relieves the patient from symptomatic enlarged spleen and effects of hypersplenism.(8,22,25) Splenectomy with esophagogastric devascularisation (Hassab's procedure) is performed as salvage therapy in case of variceal bleeding not controlled with endoscopic therapy, and a suitable size vein is not available for a shunt procedure. Splenectomy with esophagogastric devascularisation has re-bleeding rate of 11%, control of bleeding in 96%, without encephalopathy, and overall survival was 95%.(26,27)

In our study, 58.8% (20/34) patients underwent proximal splenorenal shunt and 41.2% (14/34) patients underwent modified Hassab's procedure in which shuntable vein were not present or extensive collateral precluding identification of splenic vein. All the characteristics of the patients in these surgical groups were not different other than portal biliopathy, which was significantly more frequent in modified Hassab's procedure group ( $p = 0.005$ ). This could have been because of presence of extensive collateral veins. The median duration of surgery in portosystemic shunt was significantly higher ( $p = 0.003$ ) than with modified Hassab's procedure (240 minutes versus 180 minutes). The intraoperative blood loss (300ml versus 325ml), total hospital stays (10 days versus 9.5 days) and recurrent upper GI bleeding

after surgery (0.05% versus 0.07%) were not statistically different in both surgical groups. Perioperative morbidity and mortality were also not statistically different in both surgery groups. This study was limited in identifying shunt patency rate and regression of varices due to unavailability of follow up Doppler study or CT portography and upper GI endoscopy respectively. Long term follow-up is required to know the long term outcome of these patients.

## Conclusions

Both of proximal splenorenal shunt with splenectomy and modified Hassab's procedure have good perioperative and short term outcome in case of EHPVO as a secondary prophylaxis and in patients with symptomatic enlarged spleen with hypersplenism.

**Conflict of interest:** None declared.

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