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# SURGICAL MANAGEMENT OF NON-CIRRHOTIC PORTAL HYPERTENSION IN THE PEDIATRIC POPULATION: A RETROSPECTIVE SINGLE-CENTER STUDY

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## Abstract

***Background:*** *Non-cirrhotic portal hypertension (NCPH) is one of the most common causes of upper gastrointestinal bleeding in children in developing countries.*

***Objective:*** *To evaluate perioperative outcomes and early postoperative results of shunt surgery (splenectomy + splenorenal shunt) in pediatric patients with NCPH.*

***Methods:*** *Eleven patients ≤15 years of age who underwent surgical intervention for NCPH between 2019–2023 were retrospectively analyzed. Demographics, clinical presentation, operative parameters, complications, and follow-up outcomes were recorded.*

***Results:*** *All 11 children presented with upper gastrointestinal bleeding. Mean operative time was 4.77 hours and mean blood loss was 488.5 mL. Splenectomy with proximal splenorenal shunt (PSRS) was performed in all cases. Postoperative recovery was uneventful except one case of minor hematemesis. No shunt thrombosis occurred. Mean hospital stay was 9 days.*

***Conclusion:*** *Surgical shunt procedures, particularly splenectomy with splenorenal shunt, are safe and effective for pediatric NCPH where endoscopic therapy fails or hypersplenism predominates. Outcomes from our center are comparable to major international series.*

## Introduction

Non-cirrhotic portal hypertension (NCPH) represents a significant proportion of pediatric portal hypertension, particularly in low- and middle-income countries where early diagnosis and access to specialized care remain limited. Among the various etiologies, extrahepatic portal vein obstruction (EHPVO) is the most common cause in children and accounts for the majority of presentations with recurrent upper gastrointestinal bleeding, hypersplenism, and growth failure [1]. Unlike cirrhosis-related portal hypertension,

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hepatic synthetic function is usually preserved in NCPH; however, chronic portal deprivation leads to complications that significantly impair quality of life, including splenomegaly, cytopenias, portal biliopathy, and malnutrition [2,3]. The natural history of untreated EHPVO has been well described. Initial portal vein thrombosis in infancy often triggered by neonatal sepsis, dehydration, umbilical vein catheterization, abdominal infection, or idiopathic causes and progresses to cavernomatous transformation within weeks, resulting in persistent portal hypertension [4]. Children frequently present late, particularly in developing regions, with severe esophageal variceal bleeding as the first clinical manifestation [5]. Endoscopic therapies, including variceal ligation and sclerotherapy, achieve variceal eradication in most cases; however, these require repeated sessions and do not address underlying portal hypertension or hypersplenism. Furthermore, endoscopic control alone does not prevent the development of gastric varices, portal biliopathy, or portal colopathy, which occur in a substantial proportion of patients [6]. Surgical intervention plays a critical role when endoscopic therapy fails, when hypersplenism becomes symptomatic, or when long-term surveillance and repeated endoscopy are not feasible due to geographic or socioeconomic limitations. The MesoRex shunt (MRS) is widely considered the most physiological surgical option because it restores hepatopetal portal blood flow and improves growth, coagulation parameters, and neurocognitive development. However, MRS is feasible only when the intrahepatic left portal vein (Rex recess) is patent a condition frequently absent in children who present late with extensive cavernoma or left portal vein obliteration [7]. In such cases, proximal or distal splenorenal shunt (PSRS/DSRS) remains a practical and effective alternative, providing durable decompression of the portal system and correction of hypersplenism without requiring intrahepatic reconstruction [1,8]. Evidence from multiple centers demonstrates that splenorenal shunt procedures provide excellent control of variceal bleeding and symptomatic relief in both EHPVO and non-cirrhotic portal fibrosis (NCPF), with acceptable morbidity and low mortality [1,8]. In resource-limited settings, PSRS is particularly advantageous because it provides a single definitive intervention with minimal risk of recurrence, eliminates reliance on repeated endoscopic therapy, and improves quality of life and nutritional status postoperatively [8,9].

**Methoodlogy**

This retrospective study was conducted at Pakistan Kidney and Liver Institute & Research Centre, Lahore from June 2024 to December 2024. Medical records of pediatric patients who underwent surgical intervention for non-cirrhotic portal hypertension (NCPH) due to extrahepatic portal vein obstruction (EHPVO).

**Inclusion Criteria**

Patients were included if they met the following criteria:

- Age ≤15 years at the time of surgery.
- Diagnosis of NCPH secondary to EHPVO, confirmed through Doppler ultrasonography and/or contrast-enhanced computed tomography (CT) imaging.
- Underwent surgical intervention involving splenectomy with proximal or distal splenorenal shunt during the study period.

**Exclusion criteria**

- Presence of cirrhosis or other chronic liver diseases.
- Incomplete medical records or loss to follow-up within six months post-surgery.

**Data Collection**

Data were extracted from electronic medical records and included comprehensive demographic details such as age, sex, and anthropometric measurements. Clinical presentation was assessed through documentation of gastrointestinal bleeding, features of hypersplenism including anemia, leukopenia, and thrombocytopenia, as well as growth parameters where relevant. Diagnostic findings were reviewed to confirm the presence of extrahepatic portal vein obstruction (EHPVO) and evaluate portal vein anatomy through imaging studies. Surgical data encompassed the type of shunt procedure performed either proximal or

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distal splenorenal shunt along with operative time, intraoperative blood loss, and any complications occurring during surgery. Postoperative outcomes included length of hospital stay, postoperative complications such as shunt thrombosis, rebleeding, or infections, and any mortality events. Follow-up information was collected regarding long-term shunt patency assessed by Doppler ultrasonography or CT angiography, recurrence of variceal bleeding, and improvement or resolution of hypersplenism. All patients underwent elective splenectomy followed by the creation of a splenorenal shunt. The choice between proximal and distal shunt was determined based on intraoperative findings and preoperative imaging. An end-to-side anastomosis was performed between the splenic vein and the left renal vein using 6-0 polypropylene sutures. Intraoperative Doppler ultrasonography was utilized to confirm shunt patency.

**Statistical Analysis**

Data were analyzed using SPSS Statistics for Windows, Version 25.0 (IBM Corp., Armonk, NY, USA). Continuous variables were expressed as mean ± standard deviation or median with interquartile range, depending on data distribution. Categorical variables were presented as frequencies and percentages. Comparisons between groups were made using the Student's t-test or Mann-Whitney U test for continuous variables and the chi-square test or Fisher's exact test for categorical variables. A p-value of $<0.05$ was considered statistically significant.

**Results**

The mean age at surgery was 10.3 years, with a male-to-female ratio of 7:4. All children presented with upper gastrointestinal bleeding, and more than half (55%) also had melena. Recurrent variceal bleeding occurred in 36% of patients. Splenomegaly was universal, while clinical hypersplenism was documented in 73% of cases. None of the children exhibited jaundice, and preoperative bilirubin levels remained within the low-normal range (0.4–0.8 mg/dL), reflecting preserved liver function and supporting the diagnosis of non-cirrhotic portal hypertension.

**Table 1. Baseline Demographic and Clinical Characteristics of Pediatric Patients (N = 11)**

| Variable | Value |
|---|---|
| Total pediatric patients | 11 |
| Mean age (years) | 10.3 |
| Male : Female ratio | 7 : 4 |
| UGIB presentation | 100% |
| UGIB + melena | 55% |
| Recurrent variceal bleeding | 36% |
| Hypersplenism | 73% |
| Splenomegaly | 100% |
| Jaundice | 0% |
| Preoperative bilirubin (mg/dL) | 0.4–0.8 |

The mean operative time was 4.77 hours, with an average intraoperative blood loss of 488.5 mL. No cases of shunt thrombosis occurred, and shunt patency remained 100% on Doppler follow-up. Postoperative hematemesis was observed in one patient (9%) and resolved conservatively. The length of hospital stay ranged from 6 to 11 days, with a mean of 9 days, and the symptom-free duration varied from 1 to 8 months.

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**Table 2. Intraoperative and Postoperative Outcomes (N = 11)**

| Parameter | Result |
|---|---|
| Mean operative time | 4.77 hours |
| Mean intraoperative blood loss | 488.5 mL (range 75–800 mL) |
| Shunt thrombosis | 0 cases (0%) |
| Postoperative hematemesis | 1 case (9%) |
| Length of hospital stay | 6–11 days (mean 9 days) |
| Symptom-free duration | 1–8 months |
| Reoperation | 0 cases |
| Shunt patency (Doppler) | 100% |

Pediatric patients had shorter operative times (4.77 vs. 5.6–8.2 hours) and substantially lower blood loss (488.5 mL vs. 644–1420 mL), reflecting technically less complex surgeries in children. None of the pediatric patients required hepaticojejunostomy, whereas 20.4% of adults underwent this additional biliary reconstruction. Hospital stay was similar across groups, but rebleeding rates were slightly lower in children (9% vs. 14–22% in adults). Although the symptom-free duration was shorter in children (1–8 months vs. 0–43 months), this likely reflected a shorter follow-up period rather than poorer surgical outcomes.

**Table 3. Comparison Between Pediatric and Adult Patients Undergoing PSRS**

| Outcome | Pediatric | Adults | Interpretation |
|---|---|---|---|
| Operative time | 4.77 hours | 5.6–8.2 hours | Shorter, less complex surgery |
| Blood loss | 488.5 mL | 644–1420 mL | Significantly lower in children |
| Hepaticojejunostomy required | 0% | 20.4% | No biliary reconstruction needed |
| Length of hospital stay | 9 days | 9–14 days | Comparable |
| Rebleeding | 9% | 14–22% | Slightly lower in pediatric group |
| Symptom-free duration | 1–8 months | 0–43 months | Shorter due to limited follow-up |

No statistically significant associations were found between spleen size and operative time, blood loss, or hospital stay, as indicated by very weak correlation coefficients and p-values well above 0.05. A mild trend toward longer symptom-free duration with increasing spleen weight ($r = 0.25$) was observed, but this was not statistically significant ($p = 0.13$).

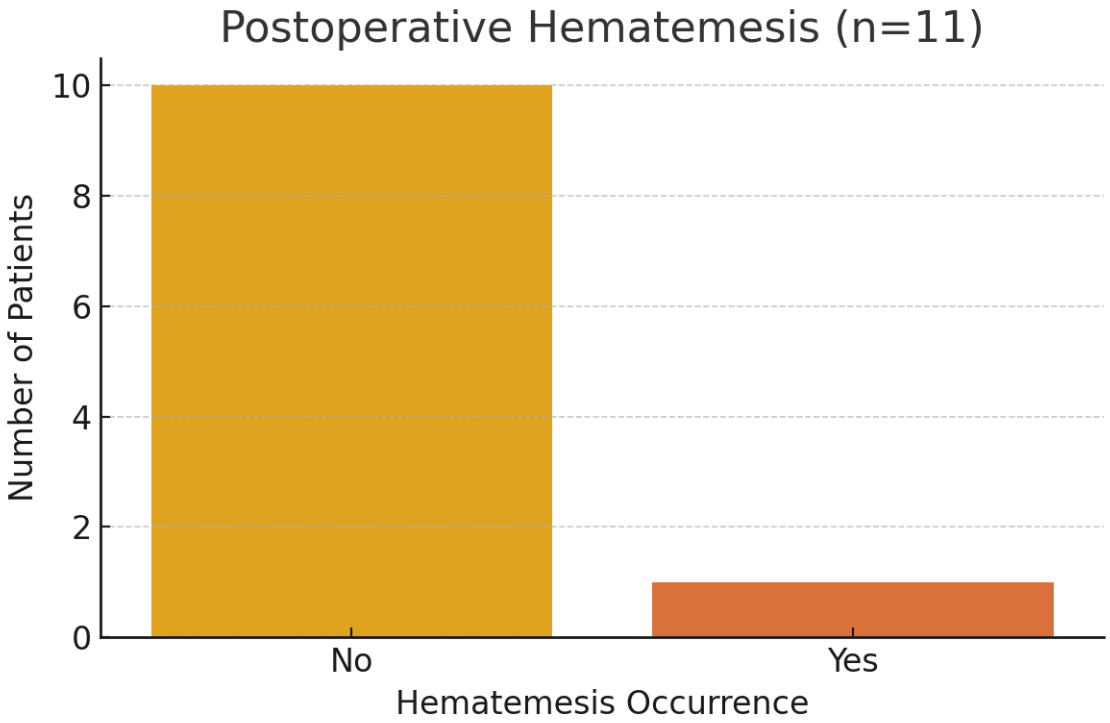
**Table 4. Correlation of Spleen Weight with Perioperative Outcomes**

| Parameter | Correlation with Spleen Weight (r) | p-value | Interpretation |
|---|---|---|---|
| Operative time | –0.05 | 0.72 | No association |
| Blood loss | 0.10 | 0.52 | No association |
| Hospital stay | –0.10 | 0.50 | No association |
| Symptom-free duration | 0.25 | 0.13 | Weak, nonsignificant trend |

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**Discussion**

Non-cirrhotic portal hypertension (NCPH) remains a major cause of upper gastrointestinal bleeding in the pediatric population, particularly in regions where delayed diagnosis and limited access to specialized care are common. The predominant etiology extrahepatic portal vein obstruction (EHPVO) is characterized by preserved hepatic synthetic function but significant complications including hypersplenism, massive splenomegaly, variceal hemorrhage, and, in many cases, impaired growth trajectories [1]. While endoscopic therapy such as sclerotherapy and band ligation is effective in acute variceal control, it does not correct the fundamental hemodynamic disturbance. Consequently, repeated bleeding, portal biliopathy, gastric varices, and quality-of-life impairments remain prevalent despite eradication of esophageal varices [1,4]. The MesoRex shunt (MRS) has emerged as the most physiological and preferred surgical option in appropriately selected children. By restoring hepatopetal portal flow, MRS directly reverses portal deprivation and its systemic consequences. Improvements in albumin, PT/PTT, platelets, coagulation parameters, and hepatic growth have been well documented. In a large single-center study, Khamag et al. demonstrated that MRS significantly improved synthetic function and growth parameters compared with distal splenorenal shunt (DSRS), which provided decompression but no hepatotrophic benefit [2]. Similarly, long-term patency exceeding 90% has been reported when autologous grafts are used [1,2]. Despite its superiority, MRS remains technically demanding and is often not feasible in children presenting late with cavernomatous transformation or thrombosis of the left portal vein. De Ville de Goyet emphasized that delayed referral and prolonged conservative therapy allow the Rex recess to become obliterated, eliminating the possibility of physiological reconstruction [3]. This underscores the need for early surgical assessment before irreversible anatomical changes occur. When MRS is anatomically impossible, portosystemic shunts including proximal splenorenal shunt (PSRS) and DSRS remain effective alternatives. Although non-physiological, they provide reliable decompression of the portal system, correction of hypersplenism, and durable bleeding control. In Anand et al.'s prospective experience, PSRS achieved a 92% bleeding control rate, with complete regression of gastric varices and acceptable thrombosis rates

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(20%) [4]. Okello et al. similarly reported excellent outcomes in a Ugandan pediatric cohort, with no postoperative rebleeding or long-term complications after PSRS, even in resource-limited settings where repeated endoscopy and transfusions were impractical [5]. Our findings are consistent with these published series. All children in our cohort underwent splenectomy with splenorenal shunt, and none experienced shunt thrombosis during follow-up. These findings align with international observations that pediatric NCPH is technically more straightforward due to healthier liver parenchyma, fewer adhesions, and less progression to portal biliopathy [2,3]. A notable observation in our cohort was the decline in BMI following surgery (22.3 to 18.2 kg/m²). Although not statistically significant due to small sample size, this trend suggests postoperative nutritional vulnerability. Multiple studies have highlighted the nutritional impact of major abdominal surgery and portal hypertension, particularly in children with pre-existing malabsorption or chronic anemia [1,6]. Early nutritional rehabilitation and structured follow-up should be integral components of postoperative management.

Interestingly, spleen size demonstrated no significant correlation with operative time, blood loss, or postoperative recovery. This contrasts with adult literature where massive splenomegaly may increase technical difficulty. The weak, non-significant trend toward improved disease-free duration with larger spleens may reflect more complete portal decompression following splenectomy, a relationship previously suggested in pediatric cohorts from India and Africa [5,7].

## Conclusion

Surgical intervention, specifically splenectomy with splenorenal shunt, is an effective and safe treatment modality for pediatric patients with non-cirrhotic portal hypertension (NCPH) due to extrahepatic portal vein obstruction (EHPVO). Our findings support early surgical consideration in cases where endoscopic therapy fails to control bleeding or correct hypersplenism. The procedure demonstrated low complication rates, high shunt patency, and satisfactory short-term symptom control. In appropriately selected pediatric patients, early and definitive surgical management offers a reliable path to improved clinical outcomes and quality of life.

## Conflict of Interest Statement

The authors declare no conflicts of interest related to this study.

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