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Operative Technique

Splenectomy with proximal spleno-left portal shunt for extrahepatic portal vein obstruction in children☆

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ABSTRACT

Purpose: To report our initial experience with splenectomy and proximal spleno-left portal shunt as an alternative to the standard Rex shunt, when not applicable, in children with Extrahepatic Portal Vein Obstruction (EHPVO).
Methods: Patients from March 2015 till September 2018, with EHPVO not suitable for Rex shunt or whose caregivers refused to consent for Internal Jugular Vein (IJV) dissection were assessed and prepared for splenectomy with proximal spleno-left portal shunt. The operative technique includes splenectomy, freeing of the splenic vein from the pancreatic bed till its junction with the inferior mesenteric vein, and then anastomosis with the intrahepatic left portal vein at the Rex recess. A distal lieno-renal shunt was performed in one patient and was excluded from the study.
Results: A total of 14 patients (mean age: 4.6 years) underwent splenectomy with proximal spleno-left portal shunt during the study period. The mean operative time was 246 min, while the mean postoperative hospital stay was 4.1 days. The patients' follow up period ranged from 6 to 42 months (median: 19.6 months). Only two patients had a single attack of variceal bleeding, 2 and 2.5 months postoperative respectively, and required endoscopic management with no further bleeding episodes. While the rest of patients showed an improvement of their variceal grades after the surgery.
Conclusion: Splenectomy with proximal spleno-left portal shunt seems to be a valuable alternative to the standard Rex shunt in treatment of children with EHPVO unsuitable for or following unsuccessful Rex shunt.
Level of evidence: IV

Extrahepatic portal venous obstruction (EHPVO) is the commonest cause of portal hypertension in children as it accounts for 66 to 76.5% of cases [1]. Rex shunt is considered as an ideal treatment for children with EHPVO and portal hypertension as it eliminates the risk of variceal bleeding and restores the normal physio-dynamics of the portal system [2].

Classic Rex shunt entails neck dissection for harvesting the internal jugular vein (IJV) and using it as a graft. This step may lead to the risk of pseudotumor cerebri [3]. Unfortunately, IJV is sometimes unavailable being thrombosed following previous insertion of central venous catheter. Moreover, dissection of IJV is worrisome for some caregivers; thus, they do not give their approval to harvest one of the main veins to drain blood from the head and the neck to be used for Rex shunt. Therefore, finding an appropriate alternative for Rex shunt has been always a priority. Proximal splenic-left intrahepatic portal shunt [4], inferior meso-left portal shunt [5], autogenous saphenous vein graft [6], and left gastric-left portal shunt [7] are among the modifications. They have the advantage of involving a single anastomosis and avoiding neck dissection.

This study was conducted to report our initial experience with splenectomy and proximal spleno-left portal shunt as an alternative to the standard Rex shunt, when not applicable, in treating children with EHPVO.

1. Methods

During the period from March 2015 till September 2018, patients with portal hypertension secondary to EHPVO and not suitable for Rex shunt or whose caregivers refused to consent for IJV dissection, were assessed and prepared for surgery by the hepatobiliary pancreatic surgery team. The team had more than 10 years' experience in hepatobiliary pancreatic surgery and liver transplantation in the pediatric surgery department, Ain Shams University.

The study was approved by the ethical committee of the pediatric surgery department, Ain Shams University. An informed consent was signed by the parents discussing the plan of care and the possibility of

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switching to another type of shunt according to the intraoperative findings.

• Preoperative assessment

Included patients suffering from EHPVO and unsuitable for Rex shunt. Patients were complaining of recurrent attacks of hematemesis due to bleeding varices despite endoscopic management, in addition to splenomegaly and manifestations of hypersplenism. Included patients proved to be Child A. Radiology included ultrasound to evaluate homogeneity of the liver, size of the spleen, and the presence of ascites. Doppler ultrasound was used to diagnose the presence of main portal vein thrombosis, assess the patency and diameter of the left portal vein, determine the direction of flow within the left portal vein, assess the presence of collateral veins, and measure the splenic pulsatility index (PI) trying to semi-quantitatively grade the degree of portal hypertension, when possible, by measuring both PBF (portal blood flow) and PVR (portal venous resistance), at least when a sufficient visualization and a good angle of insonation is obtainable, according to Ohm's law [P (pressure) = Q (blood flow) × R (resistance)]. Portal resistance was determined using the equation (0.066 × splenic PI − 0.044)] [8]. Triphasic CT was done to assess size and number of cavernoma veins and the size and patency of the splenic vein and the coronary vein were assessed. Fibroscan was done to make sure that the patients included were stage 0 or 1 to 2. Our protocol was to exclude those with stage 3 and 4 as they are more liable to decompensation [9]. Hypercoagulable tests in the form of protein C, protein S, antithrombin III, factor V Leiden, prothrombin 20,210, and Methylene tetrahydrofolate reductase were checked in the pediatric hepatology clinic before referral to the surgical team. No abnormalities were detected in any of the checked tests in all children.

Splenectomy prophylaxis was applied to all patients enrolled in the study in the form of vaccines against encapsulated organisms 2 weeks before surgery [10].

• Operative technique

The procedures on all children were performed by the same team. The recanalized umbilical vein was exposed and catheterized for direct visualization of the left portal vein by injecting a contrast agent under fluoroscopy. This was followed by dissection of the falciform ligament towards the most distal part of the left portal vein exposing its left branches. The pressure within the left portal vein was directly measured as it was cannulated using 25 g Chiba needle that was connected to monitor (Dräger® Infinity Delta). On the other hand, we did not try to dissect or cannulate the main portal vein due to the accompanying excessive number of collateral veins. We relied on the rough preoperative estimation of the main portal pressure depending on the Ohm's law and the splenic pulsatility index. After measuring the left portal pressure, a lateral clamp was placed at the most distal part of the vein. Splenectomy was then performed with ligation of all retroperitoneal shunts around the spleen and along its attachments to avoid steeling of blood towards the systemic circulation. After ligating the splenic vein and artery at the splenic hilum, the splenic vein was then completely lifted off the pancreatic bed after ligation of its short pancreatic branches till its junction with the inferior mesenteric vein. The splenic vein was then passed upwards retro-gastric to the Rex recess. End to side vascular anastomosis using Prolene 6/0 was done between the end of the most proximal part of the splenic vein and the side of the left portal vein Fig. 1. Following the anastomosis, intraoperative Doppler ultrasound was conducted to check the flow across the anastomosis.

In one patient, left portal vein was completely occluded at the Rex recess as was proved by intraoperative venography. An intraoperative decision was made to dissect the left renal vein and a distal end to side vascular anastomosis was fashioned between the end of the distal splenic vein and the side of the left renal vein using Prolene 6/0. This patient was excluded from the study.

• Postoperative follow-up

All patients received low molecular weight heparin for 3 months. Low dose aspirin was added if platelet count exceeded 6×10^5/mm. Postoperative Doppler ultrasound was performed weekly for 1 month, every other week for 2 months then every 6 months to predict the left portal vein pressure depending on portal flow measurement, and to confirm patency of the shunt and disappearance of the collateral vessels. One week after the procedure, pressures within the new main portal (splenic) and left portal veins were directly measured under sedation. The procedure could be safely accomplished by percutaneous transhepatic puncture of splenic and left portal veins using 25 g Chiba needles, which were connected to monitor (Dräger® Infinity Delta). Indirect indices such as; absence of the collateral veins, improved main and left portal veins flow and diameters, and the congestion index that was applied for the splenic shunt, were used for further follow up by Doppler ultrasound.

CT portography was done 6 months after surgery. Blood count, coagulation profile, and synthetic function of the liver were checked routinely during hospital stay and then every month. Varices status was checked by upper gastrointestinal endoscopy 3 months after surgery.

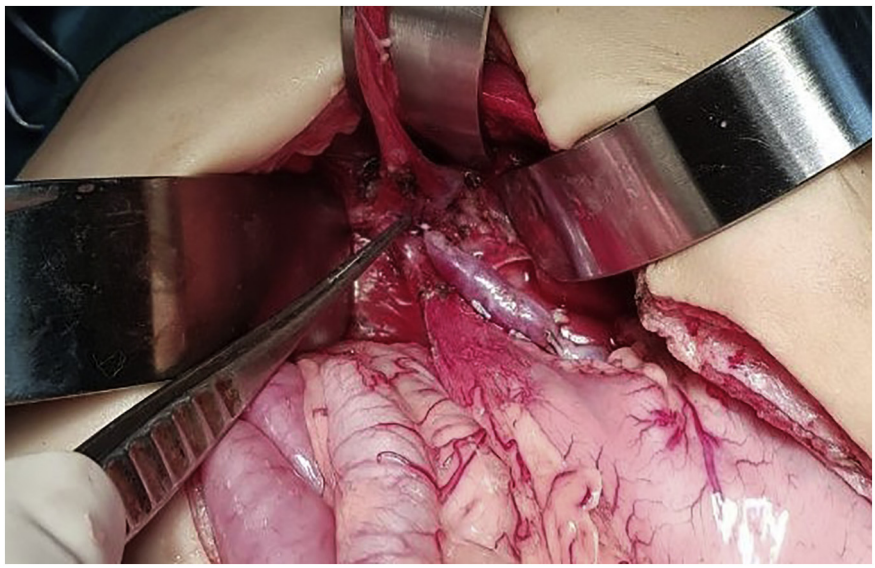

Fig. 1. End to side spleno-left portal anastomosis.

Table 1
Direct measures of intraoperative left portal venous pressure and postoperative splenic (new portal) vein and left portal vein pressures.

	Intraoperative Left Portal vein pressure (cm H_2O)	One week after surgery	
		Splenic (new portal) vein pressure (cm H_2O)	Left Portal vein pressure (cm H_2O)
Patient 1	7.2	12.2	15.2
Patient 2	5.6	14.3	18.1
Patient 3	9.4	13.8	16.4
Patient 4	12.1	16.1	15.3
Patient 5	8.7	14.9	19.4
Patient 6	5.4	12.8	14.7
Patient 7	10.3	15.2	16.4
Patient 8	6.2	12.7	13.4
Patient 9	5.7	18.3	29.1
Patient 10	6.5	16.5	17.3
Patient 11	7.2	13.4	15.5
Patient 12	11.4	17.5	14.7
Patient 13	5.6	14.8	15.2
Patient 14	8.1	13	13.3
Mean +/− SD	7.81 +/− 2.17	14.67 +/− 1.81	16.71 +/− 3.8

2. Results

Data are presented as mean (standard deviation) unless otherwise specified.

During the study period, 14 patients (9 boys and 5 girls) were operated by splenectomy with proximal spleno-left portal shunt. Their mean age was 4.6 years (range: 2–12 years). Eleven patients were not candidates to Rex shunt due to thrombosis of the right IJV proved by Doppler US following previous IJV catheterization during past NICU admission, while the remaining 3 patients' guardians refused to consent for IJV dissection. Three of our patients had previous history of umbilical vein catheterization during neonatal intensive care admission. The remaining patients had irrelevant past history. The mean operative time was 246 min (range: 236–290 min), while the mean postoperative hospital stay was 4.1 days (range: 3.6–7 days). All patients were followed up for a period ranged from 6 to 42 months (median: 19.6 months).

The predicted values of preoperative PV pressure, depending on Doppler measurements of PBF and PVR, ranged from 24.3 to 56.4 cm H_2O, reflecting in a semi-quantitative way a high grade of portal hypertension. On the other hand, direct measure of the postoperative splenic vein (new PV) pressure was 14.67 (+/− 1.81) cm H_2O. The left portal venous pressure was 16.71 (+/− 3.8) cm H_2O 1 week after surgery, in contrast to 7.81 (+/− 2.17) cm H_2O as recorded by intraoperative measurement. (Table 1).

All patients had a sort of thrombocytosis with platelet count exceeding 4×10^5/mm. Six patients had platelet count exceeding 6×10^5/mm necessitating low dose aspirin administration for 6 months (Table 2), while 2 of them required to continue low dose aspirin for extra 2 months until their platelet count dropped to the normal range.

All patients showed mild to moderate degree of postoperative ascites, which improved spontaneously. Three patients had minor collections at the tail of pancreas that resolved spontaneously. Two of them had an associated obliteration of the costophrenic angle. Both patients were managed conservatively. Nine patients had postoperative elevation of serum amylase and lipase. Seven of these patients returned to normal on the fourth day postoperative, while 2 continued to suffer from elevated pancreatic enzymes. They were asymptomatic with soft and lax abdomen. They were tolerating oral feeding and were sent home on low fat diet. Their pancreatic enzymes returned back to normal 2 weeks after discharge.

Two patients (14.3%) suffered from recurrence of symptoms in the form of a single episode of upper gastrointestinal bleeding necessitating endoscopic management. Postoperative pancreatic enzymes were normal in both of them. Routine upper endoscopic surveillance done 3 months postoperative for the rest of patients showed either improvement in the grade of varices or their disappearance.

Eleven patients were followed up for a period longer than 1 year including 2 patients with recurrent symptoms. Our protocol was to assess patency of shunts by Doppler ultrasound twice yearly. Radiologists measured the difference between hepatopetal intrahepatic left portal flow before and after the shunt. This method proved patent shunts in all patients. However, Doppler results were not correlated with the results of the CT portography images done 6 months later. The radiologists stated that neither the number of detectors used nor the protocol used for contrast injection were suitable to detect the shunt patency.

3. Discussion

Extrahepatic portal venous obstruction is usually caused by extrahepatic portal vein thrombosis (PVT). It represents the most common cause of upper gastrointestinal bleeding in children. Traditional proposed therapeutic modalities for treatment of recurrent attacks of bleeding include pharmacologic agents, endoscopic management, esophagogastric devascularization, splenectomy, and fashioning of portosystemic or distal lienorenal shunts. None of the previous procedures has been proven to be curative. In addition, they have had their own sets of complications, namely; re-bleeding, shunt thrombosis, and post-shunt encephalopathy [11]. Children with EHPVO have normal sinusoid pressure which favors the establishment of hepatopetal circulation. Therefore, construction of hepatopetal bypass is commonly used to reestablish the normal hepatic flow.

In 1992, DeVille de Goyet et al. [12], introduced using the IJV to perform a jump mesenterico-left portal shunt (Rex Shunt) to treat portal hypertension. The shunt proved effective in restoring normal hepatopetal

Table 2
Platelets count preoperatively, 1 and 6 weeks postoperatively.

	Pre-operative platelets (*10^3/ml)	One-week Post-operative platelets (*10^3/ml)	Six months Post-operative platelets (*10^3/ml)
Patient 1	42	487	232
Patient 2	31	512	190
Patient 3	58	763	673
Patient 4	28	496	156
Patient 5	79	671	621
Patient 6	64	803	322
Patient 7	35	476	223
Patient 8	54	892	178
Patient 9	72	531	198
Patient 10	42	508	213
Patient 11	35	721	409
Patient 12	69	579	327
Patient 13	38	662	213
Patient 14	110	586	207
Mean +/− SD	54.07 +/− 22.2	620 +/− 128.52	297.29 +/− 157.68

venous flow and relieving symptoms related to extrahepatic portal hypertension. They used the superior mesenteric vein as the inflow vessel.

Although original meso-rex shunt has succeeded in curing the sequelae of portal hypertension in children, efforts to modify the procedure have been continuous. These efforts are attributed to the development of pseudotumor cerebri after dissection of the neck and interruption of the venous drainage of the head and neck due to the usage of the IJV as a graft [3]. Also, using the superior mesenteric vein as an inflow vessel carries the risk of failing to maintain the influx of blood as it is frequently thrombosed or of a very small caliber. Salzedas-Netto et al. in 2011 [13] described the use of an internal jugular conduit between the distal splenic vein and the left portal vein in a case when they found the superior mesenteric vein was thrombosed. Alternative bypass veins include inferior mesenteric vein [5], coronary vein [7], and splenic vein [4]. Similar to the IJV, the saphenous vein was used as a conduit [6]. Nevertheless, all previous options entail 2 incisions and 2 anastomoses rendering the operation longer and the graft more liable to thrombosis.

In 1992, Chen et al. [4] was the first to report the use of the proximal splenic-left intrahepatic portal shunt for management of EHPVO together with splenectomy. The spleno-left portal bypass in previously splenectomized children was also described by Chiu et al. in 2007 [6]. In 2015, Zhang et al. [11] performed spleen preserving proximal splenic-left intrahepatic portal shunt with ligation of splenic artery for treatment of PVT in 4 children below 6 years of age. They highlighted the lack of evidence to support their technique in cases with huge spleen or in teenagers and above. In 2010, Warshaw stated that ligation of the main splenic vessels with preservation of the collateral circulation between short gastric and left gastroepiploic vessels has limitations in supplying a large sized spleen [14]. He reported 2% postoperative splenic infarctions in his whole series. Warshaw declared that early intraoperative evaluation of the size of the spleen and the number of short gastric vessels should be performed before decision making whether to preserve the spleen or not.

Miura and his colleagues [15] retrospectively tested the long-term hemodynamics in splenogastric circulation after ligation of splenic vessels in adult patients underwent pancreatectomy with preservation of the spleen. Their study included 10 patients who were followed up by computed tomography scans with intravenous contrast and endoscopy every 1–2 years, none of them had liver disease. They found perigastric varices in 7 cases (70%) and submucosal varices in 2 patients (20%). Another patient, without gastric varices, showed gastrorenal shunt on CT. They considered the presence of increased gastric venous flow as the cause of localized venous hypertension and hence varices, but they could not interpret the increased venous pressure in the absence of splenic artery. We believe that leaving a huge spleen totally dependent on the short gastric circulation alone is a troublesome.

The immunological function of the spleen and the overwhelming post-splenectomy infection (OPSI) are well recognized; therefore, both of them were thoroughly discussed with our patients' caregivers. Our perspective is that the incidence of OPSI can be dropped to minimal with strict application of NICE guidelines [11] for splenectomy prophylaxis. Davidson and Wall [16] stated that with proper family education, vaccination against capsulated organisms, and antibiotic prophylaxis OPSI occurrence rate dropped to 0.23%. NICE guidelines [10] for splenectomy prophylaxis were applied to all our patients. None of the patients had either OPSI or pneumococcal infections during our follow-up period. Moreover, most of our patients benefited from splenectomies as they reported dragging pain due to their huge size spleen, in addition to the immediate improvement in platelets count.

We acknowledge that by using the splenic vein as an inflow to the liver (proximal spleno-left portal shunt), we omit the option of using the splenic vein to decompress the portal circulation by anastomosing it to the left renal vein (selective porto-systemic shunt) in case of failure of the porto-portal shunting, yet we did not face this problem in our patients. Although this might represent a point of weakness to the proposed procedure, the option of nonselective portosystemic shunt is still available.

In this study, we report the use of proximal splenic vein to restore intrahepatic portal venous flow through anastomosing it to the left intrahepatic portal vein after splenectomy in patients not suitable for Rex shunt. Indications of splenectomy in our series were the huge size of the spleen, hypersplenism, and the small caliber of the coronary vein. We suggest that coronary-left portal shunt is the best technique to use in children with EHPVO because it requires one anastomosis and allows preservation of the spleen and its main vessels. Nevertheless, using the coronary vein is limited in children as it is usually of small diameter which is not suitable for such technique.

Zhang and Cheng defined postoperative recurrence as occurrence of melena or hematemesis after surgery. They reported 19% recurrence rate for different modifications of Rex shunt [17]. In our series 2 patients (14.3%) had recurrent symptoms in the form of single episode of upper gastrointestinal bleeding that needed endoscopic management despite Doppler ultrasound confirmed patency and adequacy of the graft. Hematemesis occurred after 2 months in one patient and 2.5 months in another patient. None of them required reoperation and were doing fine thereafter.

Postoperative development of ascites and elevated pancreatic enzymes were attributed to the dissection of the splenic vein which is embedded in the substance of the pancreas. Furthermore, the short pancreatic veins draining blood from the pancreas to the splenic vein were ligated and divided in order to free the whole length of the splenic vein. Follow up ultrasound revealed spontaneous disappearance of the ascetic fluid. Serum levels of pancreatic enzymes were back to normal levels in all patients within 3 weeks after surgery.

We acknowledge that the method used in our study to estimate the preoperative PV pressure, according to Bolognesi et al. [8], is insufficient to predict its precise value. Yet, it is a noninvasive maneuver and the range of values obtained denote, in a semi-quantitative way, an elevated PV pressure in all patients. Further limitations in our study were the small number of patients, the short duration of follow up, and the absence of control group.

4. Conclusion

Although the number of patients in this study was not big, it is one of the largest reported compared to previous studies describing a successful alternative for Rex shunt due to the rarity of cases. Splenectomy with proximal spleno-left portal shunt seems to be a safe and effective alternative in treatment of children with EHPVO if Rex shunt is not applicable or following unsuccessful Rex shunt.

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