Distal Splenorenal Shunt (DSRS) in Children with Extrahepatic Portal Hypertension

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ABSTRACT

Portal hypertension (PH) is infrequent in children as compared to adults. Currently, repetitive endoscopic ablation of varices is standard treatment. However, endoscopic facilities are not readily available in resource-limited settings. We reviewed the indications, operative morbidity, mortality, shunt patency and re-bleeding episodes in nine patients (age range 6-16 years, M:F 6:3) who had DSRS for recurrent GI bleeding not responding to endoscopic treatment and hypersplenism. DSRS is a safe and effective treatment alternative in resource limiting setting for preventing bleeding and improving hypersplenism in children with extrahepatic portal hypertension.

Key Words: Children, Portal hypertension, Esophageal varices, GI bleeding, Distal splenorenal shunt (DSRS).

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Extra-hepatic portal vein obstruction (EHPVO) is the cause in majority of cases of portal hypertension (PH) in children. It is usually idiopathic and is also reported following umbilical vessel catheterisation in infancy or biliary atresia surgery. Currently, medical and repetitive sclerotherapy is the recommended management of portal hypertension. However, up to 15% of children may require shunt surgery. Shunt surgery offers one-time solution to recurrent GI bleeding and hypersplenism.1 However, in pediatric age the procedure is technically demanding due to small calibre veins. This study evaluates indications, morbidity and patency of DSRS in children with portal hypertension.

After ERC (ethical review committee) approval (2019-0749-2321), medical records of pediatric patients (<18 years) who had DSRS from January 1995 to December 2018 at The Aga Khan University Hospital, Karachi (Pakistan) were reviewed for patient demographics, indication of procedure, imaging, laboratory investigations, operative time, morbidity, hospital stay, shunt patency and episodes of recurrent bleeding. Outcomes measures were prevention from repeat bleeding episodes and improvement in platelet counts. Data was analysed in SPSS version 22. Quantitative variables were reported as means and/or medians. Frequency was reported for categorical variables. Difference in outcome variables were assessed through paired t-test.

Median age of nine patients was 11 (6-16) years. Most were males (6:3). All patients had history of upper gastrointestinal bleeding including 4 (44.4%) patients having multiple episodes. Median sclerotherapy sessions before shunt procedures were 2 (0-4). All patients had normal liver function tests. The procedures were performed under general anaesthesia through transverse upper abdominal incision. The splenic vein was mobilised from its junction with superior mesenteric vein to a length which can easily be brought to the left renal vein to create a tension-free and supple end-to-side anastomosis. The mean operative time was 263 ±22.2 minutes. Routine postoperative monitoring and care was provided. There were no operative complications including bleeding, injury to the surrounding structures, wound infection, or encephalopathy. The mean hospital stay was 8.8 ±1.26 days. Patients were followed three monthly during first postoperative year and then annually, both clinically and with ultrasound doppler. Graft was considered patent if it showed flow from portal venous system to renal vein.

Median follow-up was 3 (1-10) years. Mean preoperative platelets improved from 57.3 ±22.2 to 94.3 ±34.4 X 109/l (p=0.005) and splenic size decreased from 15.6 ±2.39 to 15.03 ±2.46 cm (p=0.692, Table I). Most patients (88.8%) had no GI bleeding episode. One patient had bleeding 6 months after the surgery which required a session of sclerotherapy. Shunts were patent on last follow-up except in one patient whose shunt was noted to be patent.

**Table I:** Comparison of pre- and post-outcome parameters.

| Variables          | Preoperative Mean ± (SD) | Postoperative Mean ± (SD) | p-value |
|--------------------|--------------------------|---------------------------|---------|
| Bleeding episodes  | 9 (100%)                 | 1 (11.1%)                 | <0.001  |
| Platelets count (x 109 / l) | 57.4 ±22.2             | 94.4 ±34.4                | 0.005   |
| Spleen size (cm)  | 15.6 ±2.4                | 15.1 ±2.5                 | 0.692   |
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Distal splenorenal shunt (DSRS) in children with extrahepatic portal hypertension blocked at 6-month follow-up. She had protein C and S deficiency and had small calibre vein (4 mm) and dissection was difficult due to the previous episode of pancreatitis. Another patient required splenectomy, two years after the procedure for persistent hypersplenism (low platelets and increasing transfusion requirements to maintain hemoglobin levels).

EHPVO (extra-hepatic portal vein obstruction) is usually the cause of portal hypertension in children and it manifests with hematemesis and hypersplenism. Most patients are treated with repeated sclerotherapy and pharmacotherapy such as propranolol. In cases, which are refractory to endoscopic treatment, DSRS is a preferable alternative as it provides efficient and long-lasting decompression of the portal venous system. The reported cases of children managed with DSRS illustrate that it is effective in preventing recurrent bleeding and resolution of hypersplenism; and can safely be offered in a developing country where access to medical care is suboptimal. However, we observed that shunt surgery should not be offered in children with previous history of severe pancreatitis or hypercoagulable state.

There are several misconceptions about shunt surgery in pediatric age group, e.g. high failure rates in children under 10 years, particularly those whose veins are less than 10 mm in diameter. The other is that bleeding episodes in such children should be managed conservatively until they grow out of the disease by development of collateral circulation. There is no long-term data to support these impressions.²

Many studies have demonstrated that shunts can successfully be offered to children of any age with minimal morbidity as success rate around 90%, as is reported in different series.³ Orloff et al. have reported 97% patency and thrombosis in less than 5% of patients.⁴ Similarly, Lautz et al. reported remarkable outcome in terms of prevention of recurrent GI bleeding.⁵ Recently, Paloma et al. and Patel et al. have shown effectiveness of DSRS in children.¹,⁶ Moreover, shunts are cost-effective, psychologically less traumatic than the repeated endoscopic procedures, and favour normal physical growth in children.

In conclusion, though this is a retrospective study from a single centre with a small number of patients, it demonstrates that shunt surgery is safe and reliable alternative to repeated endoscopic procedures; for prevention of bleeding and to improve hypersplenism in children with extrahepatic portal hypertension.

AUTHORS’ CONTRIBUTION:
ZUR: Study concept, data collection, writing, critical review and revision; final approval of the article and accountability of all aspects of the article.
ZN: Study concept, critical review and revision; final approval of the article and accountability of all aspects of the article.

CONFLICT OF INTEREST:
Authors declared no conflict of interest.

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