Original Article

Portal hypertension: A critical appraisal of shunt procedures with emphasis on distal splenorenal shunt in children

Nitin Sharma, Minu Bajpai, Arbinder Kumar, Shashi Paul[1], Manisha Jana[2]

Departments of Pediatric Surgery, [1]Pediatrics, and [2]Radiology, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence: Prof. Minu Bajpai, Department of Pediatric Surgery, All India Institute of Medical Sciences (AIIMS), New Delhi - 110029, India. E-mail: bajpai2@hotmail.com

Access this article online

Website: www.jiaps.com

DOI: 10.4103/0971-9261.129599

Quick Response Code:

ABSTRACT

Background: Extrahepatic portal venous obstruction (EHPVO) is the most common cause of pediatric portal hypertension. We analyzed the investigative protocol and results of portosystemic shunts in this group of patients. **Materials and Methods:** A total of 40 consecutive children aged below 12 years operated with a diagnosis of extra-hepatic portal hypertension formed the study group. Historical data and clinical data were collected. All patients underwent upper gastrointestinal endoscopy, ultrasound Doppler and computed tomographic portogram pre-operatively and post-operatively. Results with respect to shunt patency, hypersplenism and efficacy of different radiological investigations were collected. **Results:** A total of 40 patients, 28 boys and 12 girls constituted the study group. Lienorenal shunt (LRS) was performed in 14 patients; distal splenorenal shunt in 21 patients and side-to-side lienorenal shunt in 4 patients, inferior mesenteric renal shunt was performed in 1 patient. Follow-up ranged from 36 to 70 months. At a minimum follow-up of 3 years, 32 (80%) patients were found to have patent shunts. Patent shunts could be visualized in 30/32 patients with computer tomographic portogram (CTP) and 28/32 with ultrasound. Varices regressed completely in 26/32 patients and in the rest incomplete regression was seen. Spleen completely regressed in 19/25 patients. Hypersplenism resolved in all patients with patent shunts. **Conclusions:** Portosystemic shunting in children with EHPVO is a viable option. While long-term cure rates are comparable with sclerotherapy, repeated hospital visits are reduced with one time surgery. Pre-operative and post-operative assessment can be performed with complimentary use of ultrasound, CTP and endoscopy.

KEY WORDS: Computer tomographic portogram, distal spleno-renal shunt, endoscopic sclerotherapy, extra-hepatic portal hypertension, extra-hepatic portal venous obstruction, hypersplenism, lieno-renal shunt

INTRODUCTION

Extra-hepatic portal hypertension (EHPH) is responsible for up to 2/3 of all the cases of pediatric portal hypertension. This is in contrast to the adults where cirrhosis is the most common underlying cause for portal hypertension.[1] The management of these patients has turned a full circle in the last 40 years. While in 1960's surgery was the only option available, late 1980's saw the development of endoscopic sclerotherapy (EST), which totally replaced surgery;[2] in 1990's a renewed interest has been ushered in the Porto systemic shunts as a viable option for pediatric EHPH.[3,4] Primary shunt procedure have an advantage over the sclerotherapy that

Cite this article as: Sharma N, Bajpai M, Kumar A, Paul S, Jana M. Portal hypertension: A critical appraisal of shunt procedures with emphasis on distal splenorenal shunt in children. J Indian Assoc Pediatr Surg 2014;19:80-4.

Source of Support: Nil, **Conflict of Interest:** None declared.

it provides a onetime solution to patients, takes care of the hypersplenism, prevents bleed due to hypertensive gastropathy/enteropathy and prevents patient from morbidity associated with repeated sclerotherapy sessions. We report our experience with Porto systemic shunts in children with EHPH from a tertiary care center. Our emphasis on the non-invasive investigations for assessment of the shunt ability and post-operative status is discussed along with an emphasis on the spleen preserving shunts.

MATERIALS AND METHODS

This was a retrospective study, which included forty patients out of a total of 44 patients who were referred for management of upper gastrointestinal (GI) bleed secondary to EHPH from 1998 to 2012 and received a portosystemic shunt. Indications for surgery were portal hypertension, splenomegaly and hypersplenism. Four cases which underwent shunt secondary to sclerotherapy were excluded. Ethical clearance was obtained from the institute's ethics committee. The case records of these cases were analyzed for the historical details (history for umbilical sepsis, catheterization and age at first GI bleed, number of bleeding episodes, blood transfusions, jaundice, bleeding tendency (epistaxis), previous treatments and assessment of access to health-care), demographic profile, splenic size, presence of hypersplenism (hypersplenism if present was graded as "mild" [platelet count <100,000 and total leukocyte count (TLC) 4000] or "severe" [platelet count <50,000 or TLC <3000]) and type and details of the surgical procedure. Pre-operative ultrasound Doppler (USGD) and Computer tomographic portogram (CTP) with intravenous contrast was used to obtain data on esophageal varices and paraesophageal collaterals, congestive gastropathy, splenic and superior mesenteric vein size and volume of retroperitoneal collaterals.[5,6] Upper GI endoscopy (UGIE) was performed under sedation and varices were graded according to the published protocols.[7] Post-operatively, the follow-up was carried out according to a uniform protocol. All patients received a USGD on post-operative day 5 or 6 to assess shunt patency before discharge. A follow-up USGD and a CTP was performed 3-6 months post-operatively. UGIE was performed at 6 months and yearly thereafter. Any episode of UGI bleed was recorded and an UGIE and radiological investigations were performed promptly. Secondary intervention EST was performed for patients who had re-bleed and a shunt block. Shunt was defined as clinically patent when patient did not have any GI bleed and the varices had regressed on UGIE; and radiologically patent when it could be visualized unequivocally on either USGD or CTP. The data at the time of last follow-up was compiled and analyzed.

RESULTS

Out of the 40 children, which formed the study group, 28 were boys and 12 were girls. The age range at the time of surgery was 10 months to 12 years with a median of 7 years. The follow-up period ranged from 2 to 5 years with a mean of 56 months. There was no mortality and all the children were seen at least once in last 3 months at the time of last follow-up.

Type of shunt

Lienorenal shunt (LRS) was performed in 14 patients; distal splenorenal shunt (DSRS), side to side lienorenal shunt (SSLR) and inferior mesentric renal shunts were performed in 21, 4 and 1 cases. Patients with very large spleens or with obvious large infarcts (total-14) were not considered for a spleen preserving shunt and they received a LRS.

Shunt patency

Overall 32 shunts (80%) were found to be patent at follow-up. None of these patients had any GI bleed post-operatively and varices regressed in all patients, though not completely in some patients. Radiologically, 30/32 clinically patent shunts were seen well on either USGD (28/32) or CTP (30/32). Only 1 patent SSLR and 1 DSRS could not be seen on either of the investigations.

When patency was assessed in context of the type of the shunt procedure; 11/14 (78.5%) of the LRS, 17/21 (80.9%) of the DSRS and 3/4 (75%) of SSLR were patent while 1/1 inferior mesentricorenal shunt was found patent.

USGD findings

Pre-operatively splenic vein could not be identified in 6 patients, but it could be well-seen per-operatively. Post-operatively shunt could be well-seen in 28/32 patients with patent shunt, while 2 SSLR, 1 LRS and 1 inferior mesentricorenal shunt could not be seen.

CTP findings

Pre-operatively splenic vein could be well-seen and size could be estimated in 36/40 patients. Azygos vein dilatation, enhancing paraesophageal collaterals, congestive gastropathy and varices [Figure 1] were well-seen in all the pre-operative scans, but none of them had significant biliopathy. Post-operatively, patent shunts could be visualized in 30/32 patients with a clinically patent shunt. In other 2 patients indirect evidence of patent shunts could be seen such as regression of esophageal varices and paraesophageal collaterals, normalization of the azygos vein size, decrease in retroperitoneal collaterals [Figure 2] and resolution of congestive gastropathy.

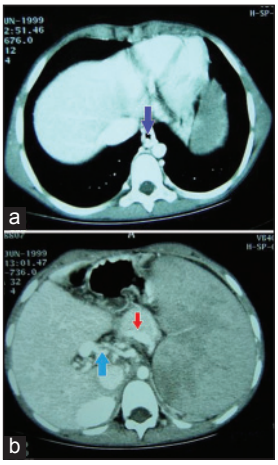

Figure 1: (a) Esophageal varices and peri-esoph collaterals, (b) Blue arrow-RP collaterals; Red arrow-splenic vein

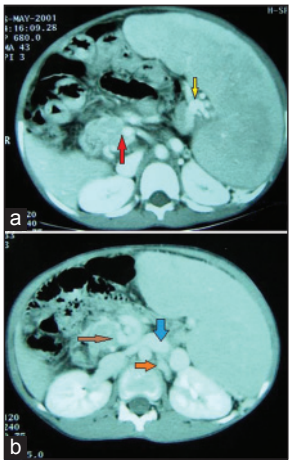

Figure 2: (a) Splenic vein (yellow) and retroperitoneal collaterals (red) (b) Post distal splenorenal shunt (DSRS) shunt — Patent DSRS (blue arrow), dilated left renal vein (orange arrow) and minimal regression of RP collaterals (brown arrow)

Overall, all 30 patients had direct or indirect evidence of patent shunts, which was marginally better than USGD. Interestingly, in patients who had received a DSRS, the retroperitoneal collaterals did not show any regression while the esophageal varices and paraesophageal collaterals regressed. In patients with the other types of non-selective shunts, the retroperitoneal collaterals regressed in volume.

UGIE findings

Varices regressed to some extent in all 32 patients with patent shunts. In 21 patients complete regression occurred and in rest 11 the grades regressed. In 8 patients with blocked shunts the varices persisted. Similar findings were seen in congestive gastropathy; it resolved completely in 26 patients and improved in 6 patients with patent shunts.

Splenomegaly and hypersplenism

Out of the 25 (21 DSRS and 4 SSLR) patients who had undergone a spleen preserving shunt, 24 were eventually able to retain the spleen (one patient with DSRS developed splenic vein thrombosis on post-operative 1 day and required splenectomy). Another patient with a spleen preserving shunt had a blocked shunt and persistent splenomegaly. In 4 other patients, the splenic size did not regress post-operatively (3 DSRS and 1 SSLR), all had splenomegaly >5 cm pre-operatively. Spleen completely regressed in 19 patients.

13 (32.5%) patients had hypersplenism pre-operatively, 8 had severe and 5 other had mild hypersplenism. 4 patients with severe hypersplenism received a LRS and 4 DSRS. Of the 5 with mild hypersplenism, 4 underwent a DSRS and 1 underwent a SSLR. At follow-up, 2 patients had hypersplenism and both had blocked shunts (1 DSRS with splenic vein thrombosis and 1 SSLR). Thus the resolution rate of hypersplenism was 85%.

Complications

We did not encounter any mortality in the present series at the time of last follow-up. 8/40 (20%) of patients had shunt block and three of them re-bled and required EST. One patient with DSRS developed splenic vein thrombosis on 2nd day of surgery and required dismantling of the shunt and splenectomy. She later re-bled and required EST. One patient developed ascites post-operatively, which was managed with repeated taps and spironolactone. Three patients had minor chest infections and 1 patient had a minor wound infection.

DISCUSSION

Extrahepatic portal venous obstruction is one of the common causes of portal hypertension in childhood; non cirrhotic portal fibrosis is also among the important causes. Children with EHPH and upper gastro-intestinal bleed present a difficult clinical challenge. Hematemesis, ascites and obvious splenomegaly have been known to be a common presenting symptom among the cases of portal hypertension in children. With the advent and refinement of the endoscopic therapies, they are regarded as the mainstay of the management of variceal bleed in children with EHPH.[2,8,9]Conventional wisdom states that the children with EHPH stop bleeding as they grow up, but there are no long-term data to support this notion. Proponents of the EST state that the bleeding episodes are not life-threatening in children with EHPH; shunts are difficult and have an associated morbidity of a major surgical procedure. There is also a concern about the chance of hepatic encephalopathy due to a long life span ahead of these children.

On the other hand, large series have demonstrated excellent shunt patency rates and minimal morbidity.[3] Patent shunts have been achieved even with 4 mm sized veins.[10] In children with EHPH who have received portosystemic shunts either selective or nonselective, the long-term chances of encephalopathy have been found to be negligible.[3,11]

Various management protocols are available for the management of portal hypertension and each has its own ifs and buts. EST is associated with variceal bleed recurrence rate of 12-30% in large series with long follow-up.[8,9] EST is not the panacea of the management of the children with EHPH and surgery may be the only option available for children with uncontrolled bleeding and the failure of EST, severe hypersplenism and growth failure.[8]

We offered portosystemic shunts to these children as most of these had a limited access to health-care and lived in far off villages. We strongly believe that providing a patent shunt to these cases is a onetime solution to their problem and saves them from the morbidity associated with repeated sessions of sclerotherapy. A significant number of patients in our series had hypersplenism and 4 were EST failures (these were excluded from the study group). We agree with Orloff *et al*.[3] that the tendency to bleed does not go away as these children grow-up. Though not a representative group, most of these children had their first bleed at less than 5 years of age and many had received multiple blood transfusions, at least 2 in all patients. The risk of multiple transfusions, hypersplenism, risk of injury to enlarged spleen and growth retardation were also the factors considered. Further repeated sessions of EST are required, which are a financial and psychological burden on the family. Most of the patients in this study group belonged to remote places with limited access to the health-care facilities and offering shunt was a onetime solution for them. The advantage of shunt was that it took care of the hypersplenism and bleeds from hypertensive gastropathy, which was otherwise not addressed by the EST.[4]

Most of the cases who underwent shunt had minimal complications as shown in Table 1. Our shunt patency rates (80%) are similar to other large studies and our re-bleed rate is comparable with EST.[3,8,9] Due to apprehensions of attendants to the asplenic state, our emphasis was towards spleen preserving shunts unless the spleen was very large or contained obvious infarcts in which we observed that splenomegaly might not regress even in patients with patent shunts. No regression was seen in 5 such patients and these children still had a palpable spleen at 2 years follow-up. These children had a pre-operative splenic size more than 5 cm below costal margin. Hypersplenism regressed in all patients with patent shunts (except two [who had blocked shunt] who had a persistent mild hypersplenism). These results were in accordance with the literature.[3,12-16] as shown in Tables 2 and 3.

In 21 patients complete regression of varices was seen and in rest of 11 the grades regressed and completely disappeared at 2 years follow-up.

In the present study, pre-operative assessment of suitability for shunt surgery and post-operative evaluation of shunt patency could be very well-assessed non-invasively by complimentary use of USGD and CTP. Pre-operatively splenic vein could not be identified in 6 patients and in these it could be well-seen per-operatively indicating that it is worthwhile to keep a

Table 1: Complications after shunt

Parameters	Complications	Bismuth *et al*.[10]	Dhiman *et al*.[18]	Shinde *et al*.[17]	Vang *et al*.[13]	Our study
Number of cases of shunt		90	53	8	25	40
Mortality		1	1	Nil	4	0
Morbidity	Intra-abdominal hemorrhage	3	Nil	Nil	1	0
	Encephalopathy	2	3	Nil	2	0
Late results	Thrombosis of the shunt	5	5	1	2	8
	Recurrent bleeding	2	3	1	6	3
	Encephalopathy	4	1	Nil	2	0
	Mortality	6	4	Nil	1	0

Table 2: Comparative description of resolution of hypersplenism following Warren shunt

Study	Total no. of patients	Number of cases with hypersplenism	Resolution of hypersplenism after shunt *n* (%)	Shunt thrombosis/ stenosis	Rebleed	Mortality
Moon *et al*.[12]	15	15	14 (93)	3	0	0
Vang *et al*.[13]	25	Not specified	Insignificant	2	6	4
Hase *et al*.[14]	15	15	15 (100)	2	3	1
Klein *et al*.[15]	25	16	13 (81.2)	0	0	0
Shilyansky *et al*.[16]	11	9	7 (77.7)	0	0	0
Our study	40	13	11 (84.6)	2	0	0

Table 3: Regression rate of spleen after shunts

Study	Number of cases with palpable spleen	Resolution of spleen	Percentage regression
Moon *et al.*[12]	15	7	46.6
Vang *et al.*[13]	13	11	84.6
Hase *et al.*[14]	5	4	80
Klein *et al.*[15]	16	16	100
Shilyansky *et al.*[16]	9	9	100
Our study	24	19	79.1

possibility of splenorenal shunt even if the splenic vein may not be identified on pre-operative studies.

CTP provided a direct visualization of patent shunt in 30 patients while in two patients with a patent shunt there were other indirect signs of shunt patency such as regression of esophageal varices, paraesophageal collaterals, congestive gastropathy, azygos vein normalization and retroperitoneal collaterals. In these respects CTP was superior to USGD and UGIE, as these could not provide any indirect measures of shunt patency. We did not required any invasive study to assess shunt patency. Another important observation was the regression of retroperitoneal collaterals in patients with LRS. Although regression of variceal decompression was excellent in DSRS, regression of retroperitoneal collaterals was not seen, indicating the selective nature of this shunt. Our group has previously reported the advantages of CTP such as early pick-up of varices, visualization of paraesophageal collaterals and importance of documenting azygos vein dilatation. As surgeons we emphasize the additional advantage of CTP in helping direct visualization of the shunt without any invasive test.

CONCLUSION

We believe that portosystemic shunting in children with EHPH is a viable option. Although long-term cure rates can be comparable with EST, repeated hospital visits are reduced with one time surgery. Primary shunt surgery is a viable and strong option for these patients in a developing country like us where people have limited access to the health-care facilities and lack proper infrastructure for repeated sessions of sclerotherapy. In patients who receive a shunt, both pre-op and post-operative assessment can be performed with complimentary use of USGD, CTP and UGIE. CTP is an excellent investigation suited for the purpose of direct visualization of the vessels.

REFERENCES

1. Mitra SK, Kumar V, Datta DV, Rao PN, Sandhu K, Singh GK, *et al*. Extrahepatic portal hypertension: A review of 70 cases. J Pediatr Surg 1978;13:51-7.
2. Howard ER, Stringer MD, Mowat AP. Assessment of injection sclerotherapy in the management of 152 children with oesophageal varices. Br J Surg 1988;75:404-8.
3. Orloff MJ, Orloff MS, Rambotti M. Treatment of bleeding esophagogastric varices due to extrahepatic portal hypertension: Results of portal-systemic shunts during 35 years. J Pediatr Surg 1994;29:142-51.
4. Maksoud JG, Gonçalves ME. Treatment of portal hypertension in children. World J Surg 1994;18:251-8.
5. Gulati MS, Paul SB, Arora NK, Berry M. Evaluation of extrahepatic portal hypertension and surgical portal systemic shunts by intravenous CT portography. Clin Imaging 1999;23:377-85.
6. Gulati MS, Paul SB, Arora NK, Mathur P, Berry M. Esophageal and gastric vasculature in children with extrahepatic portal hypertension: Evaluation by intravenous CT portography. Clin Imaging 2000;24:351-6.
7. Japanese Research Society for Portal Hypertension. The general rules for recording endoscopic findings on esophageal varices. Jpn J Surg 1980;10:84-7.
8. Molleston JP. Variceal bleeding in children. J Pediatr Gastroenterol Nutr 2003;37:538-45.
9. Zargar SA, Yattoo GN, Javid G, Khan BA, Shah AH, Shah NA, *et al*. Fifteen-year follow up of endoscopic injection sclerotherapy in children with extrahepatic portal venous obstruction. J Gastroenterol Hepatol 2004;19:139-45.
10. Bismuth H, Franco D, Alagille D. Portal diversion for portal hypertension in children. The first ninety patients. Ann Surg 1980;192:18-24.
11. Alagille D, Carlier JC, Chiva M, Ziadé R, Ziadé M, Moy F. Long-term neuropsychological outcome in children undergoing portal-systemic shunts for portal vein obstruction without liver disease. J Pediatr Gastroenterol Nutr 1986;5:861-6.
12. Moon SB, Jung SE, Ha JW, Park KW, Seo JK, Kim WK. The usefulness of distal splenorenal shunt in children with portal hypertension for the treatment of severe thrombocytopenia and leukopenia. World J Surg 2008;32:483-7.
13. Vang J, Simert G, Hansson JA, Thylen U, Bengmark TS. Results of a modified distal spleno-renal shunt for portal hypertension. Ann Surg 1977;185:224-8.
14. Hase R, Hirano S, Kondo S, Okushiba S, Morikawa T, Katoh H. Long-term efficacy of distal splenorenal shunt with splenopancreatic and gastric disconnection for esophagogastric varices in patients with idiopathic portal hypertension. World J Surg 2005;29:1034-6.
15. Klein J, Král V, Aujeský R, Utíkal P, Frysák Z, Vomácka J. Effect of portosystemic anastomosis on the degree of splenic inhibition. Rozhl Chir 2001;80:246-9.
16. Shilyansky J, Roberts EA, Superina RA. Distal splenorenal shunts for the treatment of severe thrombocytopenia from portal hypertension in children. J Gastrointest Surg 1999;3:167-72.
17. Shinde RS, Adyanthaya K, Yadav K. Extrahepatic portal hypertension - Review of 40 cases. Bombay Hosp J 2012;54:273-80.
18. Dhiman RK, Chawla Y, Vasishta RK, Kakkar N, Dilawari JB, Trehan MS, *et al*. Non-cirrhotic portal fibrosis (idiopathic portal hypertension): Experience with 151 patients and a review of the literature. J Gastroenterol Hepatol 2002;17:6-16.