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Original Article

Outcomes of Portosystemic Shunts in Children with and without Liver Transplantation

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ABSTRACT

Purpose: Limited data exist regarding outcome and morbidity associated with portosystemic shunts in the pediatric transplant population. Our study assesses the outcomes of pediatric patients who underwent a portosystemic shunt procedure, both with and without liver transplantation (LT).
Methods: This study retrospectively reviewed the medical records of pediatric patients aged 0–19 years who underwent shunt placement between 2003 and 2017 at a tertiary care center. The analysis included cases of shunt placement with or without LT.
Results: A total of 13 pediatric patients were included in the study with median age of 8.8 years. Among the cases, 11 out of 13 (84.6%) underwent splenorenal shunt, 1 (7.7%) underwent a mesocaval shunt, and another 1 (7.7%) underwent a Modified Rex (mesoportal) shunt. Additionally, 5 out of 13 (38.5%) patients had LT, with 4 out of 5 (80.0%) receiving the transplant before shunt placement, and 1 out of 5 (20.0%) receiving it after shunt placement. Gastrointestinal bleeding resulting from portal hypertension was the indication in all cases. A total of 10 complications were reported in 5 patients; the most common complication was anemia in 3 (23.1%) patients. At the most recent follow-up visit, the shunts were functional without encephalopathy, and no deaths were reported.
Conclusion: Shunt placement plays a crucial role in the management of patients with portal hypertension. Our study demonstrates favorable long-term outcomes in pediatric patients who underwent shunt placement. Long term shunt outcomes were similar and unremarkable in patients with LT and without LT.

Keywords: Hypertension, portal; Liver transplantation; Thrombosis; Biliary atresia; Portosystemic shunt, surgical

INTRODUCTION

Portal hypertension (PH) results from obstruction to the venous blood flow in the hepatic venous outflow tracts, within hepatic sinusoids or proximally in the portal venous system [1]. The spectrum of etiologies is wide [1]. If left untreated, it can lead to ascites, hepatic encephalopathy, exudative enteropathy, gastrointestinal (GI) bleeding and hypersplenism

Conflict of Interest
The authors have no financial conflicts of interest.

[2-5]. The PH can be decreased by surgically creating a portosystemic shunt, typical shunts include, mesocaval, splenocaval and splenorenal [6].

There is data in the literature regarding outcome and morbidity secondary to portosystemic shunts in the pediatric population. However, the literature is deficient on the impact that liver transplantation (LT) can have on pre-existing shunts and vice versa. The aim of this study was to review the outcomes of portosystemic shunts in patients with and without LT. A secondary aim was to compare the outcomes of patients who underwent transplantation in addition to a shunt compared to patients who underwent shunt without history of transplantation.

MATERIALS AND METHODS

This retrospective study was conducted at MedStar Georgetown University Hospital (Washington, D.C., USA) with approval for review of patient data from the review board. All pediatric patients between the age of 0–19 years of age who underwent shunt placement with and without liver transplantation were included in the study. Charts of these patients were reviewed and data collected included patient's age, sex, chief complaint, diagnosis, history of transplant, age when the transplant was done, type of transplant (liver, foregut, colon, kidney), co-morbid past conditions, age when the shunt was placed, type of shunt that was placed, number of days between transplant and shunt placement, length of hospital stay, post shunt dilation for stenosis, redo of shunt surgery, complications (infections, upper and lower GI bleeding, retroperitoneal bleeding, anemia, fever, ascites, elevated liver function tests (LFTs), shunt stenosis, transplant rejection), management of complications, change in patient's height and weight following shunting, current condition of the shunt, use of current medications for shunt encephalopathy, platelets count values at the time of shunt placement, and at 30 days after the shunt placement. The data was gathered and analyzed using Microsoft Office Excel version 2007 (Microsoft).

RESULTS

A total of 13 (n=13) pediatric patients, 8 males (61.5%) and 5 females (38.5%), who had shunt placement were included in this study. The age of the patients ranged from 1.5 to 18.8 years at the time of shunt placement with a median age of 8.8 years. Eleven (84.6%) out of 13 patients underwent splenorenal shunt, 1 patient (7.7%) underwent mesocaval shunt and 1 (7.7%) underwent failed splenorenal shunt followed by Modified Rex (mesoportal) shunt placement. Five patients (38.5%) also had LT of which 4 (80.0%) had the transplant before shunt placement while 1 (20.0%) had the transplant after shunt placement; all the five patients had splenorenal shunt placement. The time between transplant and subsequent shunt placement ranged from 315 to 3,000 days with a median of 1,504.5 days. Gastrointestinal bleeding due to PH was the indication in all our cases. Mean duration of hospital stay for the procedure was 7.9 days. The height velocity ranged from 0 to 9.4 cm/yr with a median of 5.5 cm/yr and all patients gained weight since shunt placement with a range of 9.4 kg to 39.6 kg and median of 14.5 kg. None of the patients were on any medication for shunt encephalopathy at the time of this study. At day 30th, platelets count improved in our patient with an improvement range of 12 K/cumm to 164 K/cumm and median of 60.5 K/cumm. Five (38.5%) patients developed complications of which 3 (60.0%) had transplant (2 before shunt placement and 1 after shunt placement) versus 2 (40.0%) patients who had not had a transplant. A total of

Table 1. Comparison of shunt outcomes in patients with and without history of liver transplantation

Patient details	No history of transplant before shunt placement	History of transplant before shunt placement
Number of patients	9	4
Sex distribution		
Female	5 (55.6)	0 (0.0)
Male	4 (44.4)	4 (100.0)
Reason for portal hypertension		
Portal vein thrombosis	3	4
Cavernous transformation	3	-
Congenital hepatic fibrosis	1	-
Type of shunts		
Splenorenal	7 (77.8)	4 (100.0)
Mesocaval	1 (11.1)	-
Mesoportal	1 (11.1)	-
Mean age at shunt placement (mo)	102.6	103
Median time of shunt placement after transplant (d)	-	1,504.5
Mean length of hospital stay (d)	7.2	9.3
Post shunt dilation for stenosis	1 (splenorenal shunt dilation twice, after 2 and 4 years)	0
Redo shunt surgery	0	0
Current shunt condition	Unremarkable 9/9 (100.0)	Unremarkable 4/4 (100.0)

Values are presented as number only or number (%).

Table 2. Post shunt placement complications

Type of complication	No history of transplant before shunt placement (n=9)	History of transplant before shunt placement (n=4)
Infections	0	0
UGI bleed	1	0
LGI bleed	0	0
Retroperitoneal bleed	1	0
Anemia	2	1
Fever	0	1
Chylous ascites	0	1
Transaminitis	0	1
Shunt stenosis	1	0
Transplant rejection	-	1

Values are presented as number only.
UGI: upper gastrointestinal, LGI: lower gastrointestinal.

10 complications were reported in these 5 patients. The most common complication reported was anemia in 3 (23.1%) patients. Other complications reported include upper GI bleed (1 patient, 7.7%), retroperitoneal bleed (1 patient, 7.7%), Fever (1 patient, 7.7%), chylous ascites (1 patient, 7.7%), and elevated LFTs (1 patient, 7.7%). One patient (7.7%) also underwent post-shunt twice dilation due to decrease flow, once at 2-year mark and again at 4 years mark. Another had Liver transplant rejection in the follow-up period. The follow-up period since shunt placement ranged from 0.1 to 7.4 years with a median of 5.8 years. The condition of the shunts at the last follow-up visit was unremarkable for all the patients. **Table 1** summarizes the comparison of shunts in patients with and without liver transplantation and **Table 2** summarizes complications post shunt placement in patients with and without transplantation.

DISCUSSION

In the pediatric population the most common cause of PH is extrahepatic portal vein thrombosis followed by biliary atresia [7]. PH occurs when there is either poor blood flow due to increased resistance or an increase in portal blood flow or both presenting together,

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and the hepatic venous pressure gradient (HVPG) greater than 5 mmHg is defined as PH. Also, HVPG greater than 10 mmHg is associated with esophageal varices, and HVPG greater than 12 mmHg is associated with ascites and bleeding of the varices [8]. Portal pressure is not checked routinely. The approach to variceal bleeding can be either primary prophylaxis or secondary prophylaxis. Primary prophylaxis aimed at reducing bleeding in patients with esophageal varices who have never had bleeding; this is achieved by surveillance and by using beta-blockers that reduce the portal venous pressure by constriction of the splanchnic vasculature [2,9-12]. The aim of secondary prophylaxis is to reduce the recurrence of variceal bleeding in patients with existing esophageal varices and who have bled previously. This is achieved by endoscopic variceal ligation or by sclerotherapy [2,13].

Surgical options are reserved for patients who fail endoscopic therapies. Liver transplantation is the best option for pediatric patients with progressive liver disease and who fail endoscopic therapies [8]. Surgical shunting is an excellent alternative option for patients whose liver disease is stable and does not require transplantation soon [8]. The types of shunts include portal bypass (e.g., Meso-Rex shunt) and portosystemic shunts (splenocaval, splenorenal and mesocaval) [6]. Splenorenal are the most common type of shunts followed by meso-Rex [6]. Eleven out of thirteen patients in our study had splenorenal shunts. Blachman-Braun et al. [14] reported a case of a 3-year-old girl who developed PH secondary to portal vein thrombosis and was successfully treated with revision of the meso-Rex bypass by utilizing the collateral vein.

Surgical shunts have good outcomes and control bleeding from varices in 90% of the patients [15-18]. Our patients underwent splenorenal, mesocaval and mesoportal shunts and all thirteen patients in our study had good long-term outcomes. There were 9 patients with no history of LT before shunt placement while 4 patients had a history of LT before the shunt placement (**Table 1**). When comparing both populations, there was no difference noted in terms of shunt outcomes. Similarly, Slowik et al. [1] retrospectively reviewed 34 pediatric patients with PHTN who underwent transjugular intrahepatic portosystemic shunts and concluded the procedure to be effective and safe with improvement in hematocrit, stabilization of the other laboratory values and decreased in HVPG.

Failure to maintain the shunts patent is a known long-term complication of the shunts which can lead to PHTN and associated sequelae [19-22]. One of our patients with splenorenal shunt required dilation twice post-placement; once at 2-year mark and again at 4 years mark. Stein et al. [4] in their study concluded that interventional radiologist lead percutaneous endovascular interventions are safe and effective in maintaining the patency of the portosystemic and mesoportal shunts.

In conclusion, shunt placement is an important aspect of managing patients with portal hypertension. Our study concludes a 100% good long-term outcome in patients who underwent shunt placement. In addition, there was no difference noted in terms of shunt outcomes between the LT group and non-LT group.

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