Surgery for portal hypertension in children: A 12-year review

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Background. Portal hypertension is a common and potentially devastating condition in children. Notwithstanding advances in the non-surgical management of portal hypertension, surgery remains an important treatment modality in select patients. We report here on our experience in the past 12 years.
Objectives. To describe the profile of, indication for, and complications of shunt surgery in children with portal hypertension.
Methods. Twelve children underwent shunt surgery between 2005 and 2017. Patient records were reviewed.
Results. Fourteen procedures were performed on 12 patients during the study period. The median age at surgery was 6.5 (range 1 - 18) years. Six patients were male. Gastrointestinal bleeding that was not amenable to endoscopic control was the most common indication for surgery. Portal vein thrombosis was the most common cause of portal hypertension in our series (n=11). Two-thirds (8/12) of all patients had an identifiable underlying risk factor for portal vein thrombosis. One-third of all patients (4/12) underwent a meso-portal bypass procedure (Rex shunt), while 58% (7/12) were managed with a distal splenorenal shunt. All patients received postoperative thromboprophylaxis. We experienced a single mortality, 1 patient experienced shunt thrombosis that required revision shunt surgery, and 2 patients experienced anastomotic strictures, with one being managed with revision surgery and the other currently awaiting radiological venoplasty.
Conclusions. Surgery is a safe and important tool in the management of children with non-cirrhotic portal hypertension and those with sufficient hepatic reserve who fail to respond to more conservative methods for the treatment of side effects of portal hypertension.

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Portal hypertension is a common and serious condition in children. The management of portal hypertension in children has undergone significant change with the advent of liver transplantation, progress in interventional radiology and advances in the medical and endoscopic management of variceal haemorrhage.[1,2] We report on our experience with shunt surgery over a 12-year period and provide a brief overview of literature.

Methods

Ethics approval was obtained from the University of the Witwatersrand Human Research Ethics Committee (ref. no. M170647). All patients that underwent shunt surgery at Chris Hani Baragwanath Academic Hospital, Charlotte Maxeke Johannesburg Academic Hospital, Wits Donald Gordon Medical Centre and MediClinic Sandton between 2005 and 2017 were included in the study. All candidates for inclusion were identified by a review of the databases of the Department of Paediatric Surgery at the University of the Witwatersrand. Demographic data, medical data, and data on surgical technique and outcome were collected by means of a retrospective record review. Descriptive statistics were calculated using Microsoft Excel.

Results

A total of 14 shunt procedures were performed on 12 patients between 2005 and 2017. Unfortunately, complete records on shunt surgery performed within the academic public hospitals in Johannesburg were only found for 2 patients. The remainder of cases performed within public hospitals were untraceable due to data gaps resulting from poor record keeping. The Department of Paediatric Surgery has instituted a system for electronic record management with the aim to address this problem. Half of all patients (6/12) were male. The median age (range) at surgery was 6.5 (1 - 18) years. Gastrointestinal haemorrhage that was not amenable to endoscopic control was the primary indication for surgery in 83% (10/12) of cases. The majority of patients in our series demonstrated features of hypersplenism. Portal vein thrombosis was the most common cause of portal hypertension, accounting for 92% (11/12) of cases. Two-thirds (8/12) of patients had some identifiable risk factor for portal vein thrombosis. Seven patients underwent a distal splenorenal shunt (DSRS), 4 underwent a meso-portal bypass procedure (MRB, Rex shunt), and 1 had a mesocaval bypass. All patients received post-operative thromboprophylaxis in the form of aspirin, except for 2 patients with known protein C deficiency who were initially given a low-molecular-weight heparin (enoxaparin) instead. A single patient that initially underwent a MRB procedure experienced shunt thrombosis at 3 years post surgery. This patient subsequently underwent a DSRS. We experienced 2 cases of shunt stenosis due to anastomotic stricture. One such stricture occurred in a patient that underwent mesocaval bypass and required revision of the anastomosis at 1 year post surgery. This patient demised from systemic sepsis 6 weeks after relook surgery, and was the only mortality in our series. The second patient with an anastomotic stricture is currently awaiting radiological venoplasty. Both anastomotic strictures occurred in patients with known protein C deficiency. These data are summarised in Tables 1, 2 and 3 and Fig. 1 shows the spleen size v. platelet count by means of a scatter plot.

Discussion

Normal portal pressure ranges from 0 to 10 mmHg and is usually slightly higher than the pressure in the inferior vena cava.[3] The pressure gradient between the portal and caval systems, or the hepatic venous pressure gradient (HVPG), is normally <4 mmHg. It is low owing to the high compliance and low resistance of the portal vein,

hepatic sinusoids and hepatic veins. As such, portal hypertension (PHT) is defined as either: (*i*) a hepatic venous pressure gradient >5 mm Hg, or (*ii*) hepatic venous wedge pressure >10 mmHg. Clinically, portal hypertension becomes relevant at a HVPG of >10 mmHg, as this is the threshold gradient associated with variceal haemorrhage in adults.[4] Typically, portal hypertension is defined by its clinical manifestations: (*i*) variceal gastrointestinal haemorrhage, (*ii*) splenomegaly and/or hypersplenism, (*iii*) portal hypertensive gastropathy, (*iv*) ascites, (*v*) portopulmonary disorders, (*vi*) growth abnormalities, or (*vii*) encephalopathy or learning disorders.[2,4] Although age at presentation and the specific history of presentation are variable and dependent on the underlying disease process, up to two-thirds of all patients with portal hypertension present with a history or features of upper gastrointestinal haemorrhage and splenomegaly. This constellation of symptoms is considered pathognomonic for portal hypertension.[4]

Portal hypertension can be classified into two categories, namely (*i*) portal hypertension caused by primary liver pathology with associated fibrosis, or (*ii*) portal hypertension owing to primary vascular abnormalities.[3] Both groups can be further subdivided according to the nature of the underlying disease process. Regardless of aetiology, the underlying pathophysiology ultimately involves (*i*) an increase in resistance to flow through the portal tract, (*ii*) an increase in flow through the portal tract, or (*iii*) a combination of both.[2,3] Understanding the aetiology of portal hypertension impacts significantly on the surgical strategies available for its management and potential complications.[2,3]

Pre-surgical management of portal hypertension in children is focussed on the management of variceal haemorrhage and is largely based on adult trials.[4] Management may be thought of in terms of primary prophylaxis, secondary prophylaxis, and emergency treatment of variceal bleeding. All management options involve surveillance endoscopy, administration of beta–blockers and/or somatostatin analogues, and endoscopic band ligation or sclerotherapy.[4]

Up to 15% of children with portal hypertension ultimately require shunt surgery. While shunt surgery may be classified as elective or emergent, improvements in the endoscopic management of variceal bleeding means that emergency shunt surgery to control upper gastrointestinal bleeding is now a rare occurrence.[2,4] In the modern era, shunt surgery is reserved for patients with extrahepatic portal hypertension in whom the procedure may be curative, and those with sufficient hepatic reserve where a risk benefit analysis favours shunt surgery over liver transplantation. More specific indications for elective shunt surgery are presented in Table 4.[1,3,5]

Table 1. Demographic data and risk factors

Patient no.	Age (years)	Gender	Risk Factor
1	11	F	None
2	15	M	Neonatal umbilical catheter
3	8	F	None
4	13	F	Neonatal umbilical catheter
5	5	M	Neonatal umbilical catheter
6	1	F	Previous liver transplant
7	10	M	Cystic fibrosis
8	1	F	Protein C deficiency
9	2	F	None
10	7	M	Protein C deficiency
11	6	F	None
12	5	M	Neonatal umbilical catheter

F = female; M = male.

Table 2. Patient characteristics

Patient no.	Spleen size (cm)	INR	Platelet count (x 10^9/L)	Presenting symptoms
1	11	1.1	30	GI haemorrhage
2	15	1	90	GI haemorrhage
3	8	1.2	86	GI haemorrhage
4	13	1	77	GI haemorrhage
5	5	1	98	GI haemorrhage
6	1	1.2	68	Ascites
7	10	1.4	33	GI haemorrhage
8	1	1	40	GI haemorrhage
9	2	1	90	GI haemorrhage
10	7	1.1	82	GI haemorrhage
11	6	1.2	78	GI haemorrhage
12	5	1	198	GI haemorrhage

INR = international normalised ratio; GI = gastrointestinal.

Table 3. Diagnosis, surgical procedure, and outcome

Patient no.	Diagnosis	Procedure	Outcome
1	PVT	Distal splenorenal	Patent till present
2	PVT	Mesoportal	Thrombosed, DSRS 3 years post initial surgery
3	PVT	Mesoportal	Patent till present
4	PVT	Mesoportal	Patent till present
5	PVT	Distal splenorenal	Patent till present
6	PVT	Distal splenorenal	Patent till present
7	Cystic fibrosis	Distal splenorenal	Patent till present
8	PVT	Mesocaval	Anastomotic stenosis, distal anastomosis revised 1 year after primary surgery, demised from sepsis 6 weeks post revision
9	PVT	Distal splenorenal	Patent till present
10	PVT	Distal splenorenal	Anastomotic stenosis, awaiting venoplasty
11	PVT	Mesoportal	Patent till lost to follow up 2 years post surgery
12	PVT	Distal splenorenal	Patent till present

PVT = portal vein thrombosis; DSRS = distal splenorenal shunt.

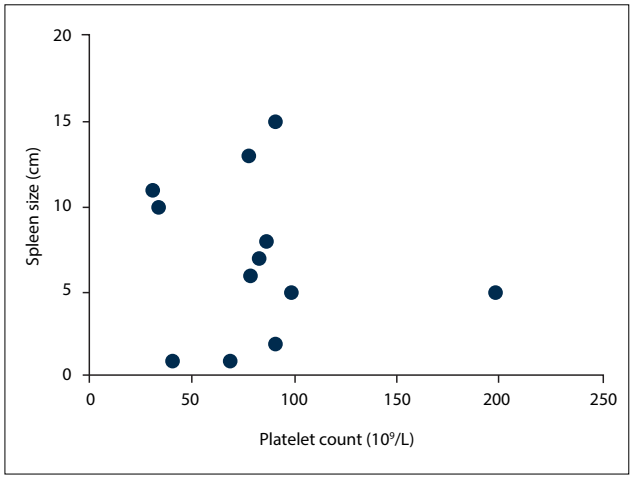

Fig. 1. Spleen size v. platelet count.

Table 4. Indications for shunt surgery in portal hypertension

Uncontrolled bleeding from oesophageal varices*
Bleeding from gastric or ectopic varices
Hypersplenism or massive splenomegaly
Isolated extrahepatic portal vein obstruction
Lack of access to endoscopy
Symptomatic biliary obstruction due to choledochal varices
Neurological impairment
Patient choice

*That is, no response to at least 2 sessions of endoscopic treatment.

Shunt surgery may be thought of as physiological or non-physiological. The only true physiological shunt is the meso-portal bypass shunt (MRB, Rex shunt) in which an obstructed portal vein is bypassed to restore normal hepatopetal flow.[3,6,7] Non-physiological shunts result in the diversion of blood from the mesenteric and portal systems to the systemic circulation. Non-physiological shunts can be further divided into selective and non-selective shunt groups, with the non-selective group being further divided into total and partial shunts. Total non-selective shunts, such as the portocaval, mesocaval and proximal splenorenal shunts, create direct communication between the portal and systemic circulation. Depending on the specific technique, these shunts can create a full diversion of flow with a consequent fall in portal pressure. While this significantly reduces the risk of bleeding from varices, it is also associated with a marked increase in the risk of encephalopathy.[2-4] In contrast, partial non-selective portosystemic shunts (such as the Sarfeh shunt) maintain some portal flow and are thus associated with lower rates of post-operative encephalopathy. This benefit comes at the cost of higher rates of shunt thrombosis and recurrent variceal bleeding.

Selective shunts such as the DSRS (also known as the Dean Warren shunt) are intended to only decompress the compartment associated with variceal bleeding. In this way these shunts maintain some portopetal flow and avoid the encephalopathy associated with non-selective shunts. With time, this selectivity is lost and selective shunts progressively centralise.[2,3,6,7]

Prehepatic portal hypertension is the most common cause of portal hypertension in children. In most cases this is due to idiopathic portal vein thrombosis, with <25% of patients reported to have some history of umbilical vein catheterisation, trauma, previous surgery or peritonitis.[4] The MRB is an example of a portoportal shunt and was first described in 1992 as a technique to treat portal vein thrombosis post liver transplantation.[7] This procedure utilises a graft patch between the superior mesenteric vein and the left portal vein at the level of the Rex recesses to bypass portal vein obstruction and restore flow to the liver.[1,3,7] The MRB restores portal venous flow, thus restoring normal physiology. In this way, it resolves portal hypertension in patients with isolated extrahepatic portal vein obstruction (EHPVO) and, as such, is the only curative surgical shunt. Successful MRB facilitates the reversal of the effects of portal hypertension including: (*i*) restoration of hepatopetal flow; (*ii*) resolution of portosystemic collaterals; (*iii*) reversal of hepatopulmonary syndrome; (*iv*) resolution of hypersplenism; (*v*) improved neurocognitive function; and (*vi*) decreased risk of focal nodular hyperplasia in the liver.[7]

Shunt thrombosis and anastomotic stricture are significant post-operative complications, though patency rates of 97% and thrombosis rates of <5% are reported within the international literature.[1-3] Outcomes are better in patients undergoing Rex bypass earlier in the disease process and current recommendations are for children with EHPVO to undergo MRB prior to the onset of portal hypertension.[3]

With advances in microsurgical techniques, and through the experience gained in vascular and transplant surgery, shunt surgery (including MRB) is now successfully performed in children as small as 4 kg.[3] Prerequisites for a successful Rex procedure are (*i*) the absence of intrinsic liver pathology, (*ii*) a patent intrahepatic portal tree, and (*iii*) the presence of a suitable conduit (usually the internal jugular vein (IJV)). The choice of conduit impacts patency rate, with IJV grafts having the highest patency rate and cadaveric iliac vein grafts the lowest patency rates.[3] Shunt patency must be checked postoperatively within the first week, using Doppler ultrasonography, and post-operative thromboprophylaxis with dual antiplatelet therapy (aspirin and dipyridamole) is recommended for 3 - 6 months.[2,3]

The basic work-up of patients requiring shunt surgery involves: (*i*) an assessment of the cause of portal hypertension; (*ii*) an assessment of the patient's hepatic reserve; and (*iii*) the anatomy of the portal venous tree.[1-3,6,7] Correct delineation of the anatomy of the portal tree is critical to surgical planning and can be achieved through Doppler ultrasonography, contrasted computed tomography, magnetic resonance venography or, most definitively, through formal portal venography. Imaging must demonstrate the anatomy (including length and separating distance) and patency of the portal vein, superior mesenteric vein, splenic vein, left renal vein, inferior vena cava, and the left portal vein.[1-3,6,7] Doppler ultrasonography may also be used to assess the patency and calibre of the internal jugular veins if they are considered as conduits for bypass.[3] In cases of primary liver pathology with associated fibrosis, the pre-operative workup should include a percutaneous liver biopsy.[3] Hepatic reserve is measured by various criteria, the most common being the Child's Pugh or Paediatric End Stage Liver Disease (PELD) scoring systems. Both systems estimate hepatic reserve by scoring patients on hepatic synthetic function as measured by albumin, International Normalised Ratio (INR), and total bilirubin levels. Preoperative assessment of coagulation profiles is essential in nontransplant vascular surgery such as shunt surgery. Given that coagulopathy is a risk for intra- and post-operative haemorrhage and shunt thrombosis, patients with a history of venous thrombosis (portal or other) must be thoroughly screened to identify those with genetic hypercoagulability.[3]

Alternatives to shunt procedures include devascularisation procedures such as the Sugiura procedure.[2-4] All devascularisation procedures involve splenectomy and in the era of endoscopy and liver transplantation these procedures are considered obsolete. In selected cases of severe hypersplenism, splenectomy or splenic artery

embolisation has been described. Splenic artery embolisation reduces flow into the portal system, thereby decreasing portal pressures and bleeding from varices, although this is a temporary achievement.[2-4]

Splenic artery embolisation becomes relevant in instances of thrombosis or stenosis of DSRS and persistent hypersplenism. Adverse consequences of splenic artery embolisation or splenectomy include post-splenectomy sepsis and intra-abdominal adhesions that increase risks associated with future shunt surgery.[2-4] As such, surgical or radiological splenectomy are considered last resorts in children with portal hypertension.[2] Children with intrinsic liver disease and insufficient hepatic reserve should not be considered candidates for shunt surgery. In such cases, the rate of complications of shunt surgery prevail over the potential benefits and these children should rather be considered for primary liver transplantation.[3]

Conclusion

The management of portal hypertension in children has undergone significant improvements with advances in the available medical, endoscopic, surgical and radiological interventions for the treatment of portal hypertension and its complications. These advances include: (*i*) beta-blockers in the pre-primary and primary prophylaxis of variceal haemorrhage; (*ii*) endoscopic band ligation and sclerotherapy in the setting of acute variceal haemorrhage and as secondary prophylaxis; (*iii*) advances in liver transplantation techniques increasing feasibility of the procedure in children; and (*iv*) advances in interventional endovascular radiological procedures. Collectively, these advances have significantly decreased the need for emergency and elective shunt and non-shunt surgery in the management of portal hypertension. This said, shunt surgery remains relevant in patients with non-cirrhotic portal hypertension, in whom shunt surgery may be curative, and in those with sufficient hepatic reserve who fail to respond to more conservative treatment options as a bridge to transplantation. The choice of shunt surgery depends on the underlying pathology, overall health of the patient (including a measure of hepatic reserve), patient choice, and the available technical expertise. With advances in surgical technique, shunt surgery is a feasible option in children with well compensated cirrhosis or EHPVO regardless of their age or size. Shunt surgery prevents GI bleeding, corrects splenomegaly and hypersplenism, portal biliopathy, ascites and can alleviate growth retardation.[2-4,6]

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Conflicts of interest. None.

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