

## Management of parastomal varices: who re-bleeds and who does not? A systematic review of the literature

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**Abstract** Although first described almost half a century ago, parastomal varices are not easily recognised as a cause of stomal bleeding even though they occur in up to 5 % of all people who have a stoma. The main challenges associated with this condition are diagnosis and management. For that reason, the aim of the present study was to perform a systematic review of all the available literature pertaining to this topic. The primary end point was recurrent variceal haemorrhage after a particular mode of management. Several secondary endpoints focused on means of diagnosis and pathological conditions of abdominal organs that could contribute to both the formation of these varices and the rate of re-bleeding. Sixty-six articles comprising 210 patients were analysed. Parastomal varices tend to be more frequent in men manifesting with bleeding in the fifth decade of life. The majority (72.0 %) of patients who bleed from parastomal varices do so from an ileostomy. The most common pathology leading to stoma formation is ulcerative colitis (57.8 %). Liver cirrhosis is the most common cause of portal hypertension leading to the development of parastomal varices and primary sclerosing cholangitis is in second place. A third of patients with parastomal varices also have co-existent oesophageal varices. There are no pathognomonic symptoms or signs of parastomal varices and only the minority of patients have a raspberry appearance of the stoma, visibly dilated submucosal veins and bluish discoloration and hyperkeratosis of the skin around it. Venous phase contrast angiography or portal venography is the most successful radiological investigation to confirm the diagnosis. The transjugular intrahepatic

portosystemic shunt (TIPS) procedure has the highest success rate in preventing recurrent haemorrhage and local measures, either non-operative or surgical, are the least effective. Comparison of TIPS with non-operative and local surgical treatment groups produced a risk reduction in 4.60 and 3.85, respectively. Treatment of 1.37 people with a TIPS procedure prevents one person suffering from recurrent variceal bleeding and using TIPS can reduce the likelihood of re-bleeding by 78.5 %. Surgical portosystemic shunting or embolisation alone leaves patients with approximately 50 % chance of re-bleeding. Although TIPS has gained popularity over the last two decades almost three quarters of patients with parastomal varices are still treated with local measures as first-line management. Liver transplantation as a treatment of the primary cause of parastomal varices remains very rare.

**Keywords** Parastomal · Peristomal · Varices · Ileostomy · Colostomy

### Introduction

It is well known that obstruction of the portal venous blood flow and resulting portal hypertension leads to the development of pathologically enlarged venous channels at the junction between high pressure portal and low pressure systemic venous systems. There are several naturally occurring sites of portosystemic venous communication. These include: the lower oesophagus, where the tributaries of the left gastric (portal) and oesophageal (systemic) veins meet; the anal canal with the meeting of the superior rectal (portal) and middle and inferior rectal (systemic) veins; the bare area of the liver (tributaries of the portal venous system meet the systemic phrenic veins) and the paraumbilical

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region where small paraumbilical veins draining to the portal system meet systemic veins of the anterior abdominal wall. Increases in portal pressure, such as in hepatic cirrhosis, allow these portosystemic anastomoses to offer an alternative route for blood to flow. The higher pressure portal system forces abnormal dilatation of these communicating veins, which clinically are seen as varices, or in the paraumbilical area of the abdominal wall, as the *caput medusae*.

It is therefore not surprising that collateral venous channels may also form at the surgically constructed mucocutaneous junction of a colostomy or ileostomy, where the intestinal veins are iatrogenically juxtaposed with the systemic veins of the anterior abdominal wall. These so-called “parastomal varices” or “peristomal varices” develop in patients with a stoma, who also suffer from liver disease. Similar to their counterparts elsewhere in the body, parastomal varices are not painful but can bleed in a torrential and repeated manner and there have even been reports of loss of life secondary to acute haemorrhage in up to 4 % of cases.

The main challenges associated with this condition are diagnosis and management. Certain physical signs and several diagnostic tools have been described to assist in diagnosing parastomal varices, and clinicians’ awareness of the condition is clearly of key importance. Over the last 40 years, multiple different management approaches have been employed to control bleeding from parastomal varices, mostly at a local level. However, we will show that these attempts are futile unless the underlying cause is addressed.

The condition is not rare—it affects up to 5 % of all people who have a stoma. Despite this it represents a clinical entity relatively poorly reported in the literature. For that reason, the aim of our study was to carry out a systematic review of the available literature with and perform a meta-analysis of all reported cases to date to guide clinicians with the diagnosis and successful management of parastomal varices.

## Materials and methods

### Literature search

In December 2010, relevant articles were sourced from National Health Service (NHS) evidence, a website combining various databases, including Embase and Medline ([www.evidence.nhs.uk](http://www.evidence.nhs.uk)).

Similar searches were applied to NextBio ([www.nextbio.com](http://www.nextbio.com)). The search terms included the words ‘bleeding’, ‘stoma’, ‘stomal’, ‘varices’, ‘peri’, ‘para’, ‘ectopic’ and

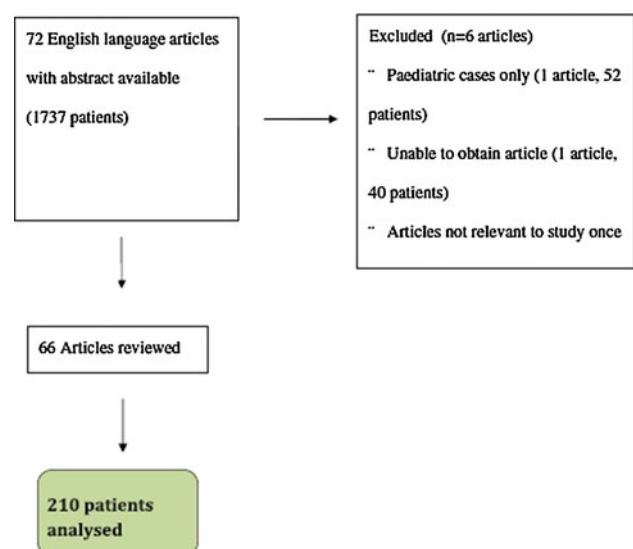
‘portal hypertension’, either alone or in combination. Relevant articles referenced in publications were also reviewed for possible inclusion. The literature search was carried out by both authors.

### Article selection and data extraction

Articles without an abstract were excluded. Abstracts of all English language articles were reviewed and subsequently the full text of the articles was obtained. At the time of the search, one article was available ahead of publication on-line only and the printed version was obtained subsequently.

In total, 72 articles comprising 235 patients were reviewed by both authors. Six articles were excluded for reasons outlined in the flowchart on the selection of articles (Fig. 1). This brought the final number of cases available for analysis to 210.

An Excel database was then devised to collect available data for comparison into a single data collection form. Data were extracted both on the primary and secondary endpoints of the study. The primary endpoint was whether or not a patient suffered recurrent variceal haemorrhage after undergoing a particular mode of management. The volume of blood loss during each episode of bleeding was impossible to quantify as this detail was not routinely recorded in the articles reviewed. We accepted that where the authors saw fit to report parastomal variceal haemorrhage this was likely to be troublesome enough for the patient and to warrant medical attention. There were several secondary endpoints, which focused on means of diagnosis as well as on underlying pathological conditions of abdominal organs that could have contributed to both the formation of parastomal varices and the recurrence of bleeding.



**Fig. 1** Selection of articles’ flowchart

## Statistical analysis

Forty-four variables from the selected articles were critically mapped to allow integration of the reported quantitative findings and provided a numerical estimate of the overall effect of interest. We excluded variables from the analysis if that particular variable was reported in only 15 or less cases out of the 210 analysed. We explored patterns using cross-tabulation and corresponding  $\chi^2$  tests. We also computed risk measures. Statistical analysis was carried out using IBM SPSS, version 18.0, computer software (SPSS, Chicago, Illinois, USA).

## Results

### Patient demographics and general information

Out of all patients with documented gender ( $n = 139$ ), 61 % were male and 39 % female. The relative risk (RR) of suffering from parastomal varices if you were male was 2.5. The mean age at presentation with bleeding was 50.8 years with a range of 18–82 years ( $n = 179$ ).

Table 1 shows the incidence of various diseases of abdominal organs necessitating stoma construction where they were clearly identified in the literature ( $n = 133$ ). As expected, the most common pathology leading to stoma formation was ulcerative colitis (57.8 %), followed by carcinoma of the rectum (23.3 %) and carcinoma of the urinary tract (9.0 %). Table 2 shows the incidence of underlying liver diseases leading to portal hypertension ( $n = 182$ ). The majority (72 %) of patients who bled from parastomal varices did so from an ileostomy ( $n = 165$ ). This figure reflects the high proportion of patients who underwent colectomy for ulcerative colitis. Nineteen percent of patients bled from a colostomy and 8 % from an ileal conduit. The remaining 1 % consisted of two patients, one with an ureterocolostomy and the other with both an ileostomy and a sigmoid colostomy. The average time from stoma formation to the first documented episode of bleeding was 73.6 months, (range 1–480, median 48 months) ( $n = 140$ ).

Many patients (42.9 %) required blood transfusion as a direct result of bleeding from parastomal varices. Portal venous pressures were documented only in a small group of patients undergoing transjugular intrahepatic portosystemic shunt (TIPS) procedure ( $n = 24$ ). The average drop in portal pressure after a TIPS procedure was 12.3 mmHg (range 6–22 mmHg). Co-existent oesophageal varices were clearly documented in 66 cases (31.6 %). Their association with bleeding parastomal varices was previously explained by the concept of splanchnic compartmentalisation due to non-uniform directional blood flow patterns in portal

**Table 1** Incidence of abdominal pathology leading to stoma formation

Abdominal pathology	Incidence	
	Number	%
Ulcerative colitis	77	57.8
Carcinoma of rectum	31	23.3
Carcinoma of urinary tract	12	9.0
Crohn's disease	6	4.5
Colonic diverticular disease	5	3.7
Carcinoma of anus	2	1.5
HIV-related lymphoma	1	0.8
Carcinoma of uterus	1	0.8
Necrotising enterocolitis	1	0.8
Benign rectal stricture	1	0.8
Adenomatous polyposis coli	1	0.8
Streptococcus viridans septicaemia	1	0.8
Gastric ulcer and ileus	1	0.8
Hypotonic bladder	1	0.8

**Table 2** Incidence of underlying liver disease

Liver disease	Incidence	
	Number	%
Cirrhosis (any cause)	81	44.5
Primary sclerosing cholangitis	59	32.4
Hepatitis (any cause)	19	10.4
MELD stage 3 and 4	10	5.5
Liver metastases	5	2.7
Nodular hyperplasia	2	1.1
Cryptogenic liver disease	1	0.5
Cholestatic liver disease	1	0.5
Hepatocellular carcinoma with portal vein thrombosis	1	0.5
Portal vein thrombosis (unspecified)	1	0.5
Actinomyces liver abscess	1	0.5
Unknown	1	0.5

*MELD* The model for end stage liver disease, where the exact nature of the liver disease was not recorded. Some authors simply cited “MELD stage 3–4”

hypertension [1]. Some authors suggested that patients who bled from parastomal varices were less likely to bleed from their oesophageal varices due to the “safety valve” theory [2, 3]. Unfortunately, documentation of bleeding from oesophageal varices was poor, which precluded any meaningful conclusion pertaining to this subtopic. A similar problem with analysis of small numbers was applied to the Child-Pugh score of portal hypertension, which was recorded in only 13 (6 %) out of 210 patients.

## Diagnosis of parastomal varices

In many cases, patients reported severe bleeding from their stoma, which settled with pressure before or at the time of the examination by a clinician. The classical findings on physical examination were well documented in a third (33 %) of all patients. These included bluish discoloration of the skin, raspberry appearance of the stoma, visibly dilated submucosal veins, *caput medusae* around the stoma and hyperkeratosis of the skin. Some patients described irritation of the skin, which bled on trivial contact. Removal of the stoma appliance with careful examination of the surrounding skin was, therefore, advocated.

Authors reported which tool they used to successfully diagnose parastomal varices in 77 cases. Venous phase mesenteric angiography was cited in 40.2 %, portal venography was successfully employed by 37.7 % and Doppler ultrasound helped to reach the diagnosis for 14.3 % of authors. Computerised tomography (CT) scan was reported as providing the correct diagnosis in 7.8 % of all reported cases. Transstomal endoscopy proved unreliable. Within the analysis, 24 authors reported the use of transstomal endoscopy. In these reports, correct endoscopic diagnosis of parastomal varices occurred in only 5 cases.

## Management of parastomal varices

Over the decades, many methods have been employed to control bleeding from parastomal varices. In order to facilitate meaningful analysis, these methods have been divided into 6 groups:

**Table 3** Non-operative treatment of parastomal varices

Non-operative treatment	First-, second- and third-line management attempts (numbers reported)		
Compression	38	8	5
Ligation	35	25	7
Unspecified “conservative” measures	33	7	1
Unspecified “local measures”	9	0	0
Percutaneous embolisation	9	3	0
Propranolol	7	4	4
Sclerotherapy	4	4	4
Cautery	1	2	0
Vasopressin infusion	1	0	0
Vitamin K	1	1	0
Octreotide infusion	1	0	0
Argon beam	1	0	0
Overall	140	54	21

- Non-operative treatment (NOT) (Table 3).
- Local operative treatment (LOT) (Table 4).
- Surgical portosystemic shunts (SPSS) (Table 5).
- Embolisation (transhepatic or transjugular) (Table 6).
- Transjugular intrahepatic portosystemic shunt (TIPS) (Table 6).
- Liver transplant (Table 6).

We looked at first-, second- and third-line management attempts for these patients to see if patients were being offered alternative management techniques when one line of treatment failed, or if clinicians were persevering with unsuccessful techniques regardless of the outcome (perhaps as it has not been clearly established what constitutes the best management of this condition).

Table 3 shows that compression and other conservative measures make up the majority of the first management options offered to patients. When these measures fail, however, they are less likely to be offered as second- and third-line therapy. The overall trend demonstrates that non-operative local measures are popular at the initial

**Table 4** Local operative treatment of parastomal varices

Local operative treatment	First-, second- and third-line management attempts (numbers reported)		
Revision and resiting of stoma	13	24	9
Local treatment via laparotomy	3	1	0
Muco-cutaneous disconnection	1	10	1
Circumferential suture technique	2	0	0
Overall	19	35	10

**Table 5** Surgical portosystemic shunt procedures for parastomal varices

Operative shunt	First-, second- and third-line management attempts (numbers reported)		
Mesocaval	1	5	1
Portocaval	0	6	3
Splenorenal	2	3	8
Overall	3	14	12

**Table 6** Treatment of parastomal varices with TIPS, embolisation and liver transplant

Procedure	First-, second- and third-line management attempts (numbers reported)		
Embolisation	7	13	9
TIPS ± embolisation	16	20	6
Liver transplant	0	0	5

*TIPS* Transjugular intrahepatic portosystemic shunt

presentation. Of the local operative measures available, stoma revision and relocation remain widely used. The described technique of mucocutaneous disconnection is based on a similar principle where the stoma is disconnected from the dermal interface and each individual varix is isolated and tied off. Circumferential suture technique involves placement of large sutures over visible veins and tissue around the stoma. The lack of treatment of the primary cause meant the success from local surgical techniques is short lived as new varices form in the same pathological manner.

Table 5 shows a general trend to create surgical porto-systemic shunts if other measures fail. Shunts are successful in achieving long-term control of variceal haemorrhage but like any invasive surgical procedure they are not without risks of morbidity and mortality. The TIPS procedure has been gaining favour over recent years although is still infrequently used. A liver transplant is only offered when other methods have failed.

Re-bleeding rates after different types of management

Prevention of the recurrent variceal haemorrhage was considered the most important long-term outcome of treatment. Therefore, re-bleeding rates were analysed as a measure of success of the various management options (Table 7). Patients in whom re-bleeding rates were not clearly documented were discounted from the analysis, which accounts for different total numbers of patients as compared with Tables 3, 4, 5 and 6.

It was obvious that managing a patient with non-operative local measures would almost certainly lead to further episodes of bleeding. One hundred and thirty-nine (85 %) of 164 patients managed in this way re-bled. Local surgical procedures such as ligation of the varices or re-siting the stoma were also associated with high levels of re-bleeding in 29 (81 %) of 36 patients in this review. Statistical analysis showed that the RR of re-bleeding after non-operative treatment as opposed to a local surgical procedure was 1.2. TIPS was a successful means of controlling variceal bleeding with only 20 % of these patients suffering from re-bleeding. Comparison of the non-operative

**Table 7** Re-bleeding rates according to category of management

Management technique	Re-bleeding rate % (number of patients who re-bled/total number treated by that means)
Non-operative treatment	84.76 (139/164)
Local surgical treatment	80.56 (29/36)
Embolisation	45.45 (10/22)
Surgical porta-systemic shunt	38.46 (5/13)
TIPS	20.00 (4/20)

TIPS Transjugular intrahepatic portosystemic shunt

treatment group with the TIPS group produced an RR of 4.6 making patients almost five times less likely to re-bleed following a TIPS procedure. When local operative treatment was compared with TIPS, the RR was 3.85. We also looked into re-bleeding prevention with TIPS instead of conservative management and calculated that the number needed to treat was 1.37 and the relative risk reduction was 78.5 %. Therefore, treatment of 1.37 people with a TIPS procedure prevents one person from suffering from recurrent variceal bleeding and using TIPS can reduce the likelihood of re-bleeding by 78.5 %.

Changes in the management of parastomal varices through the decades

In the 1960s, management was restricted to non-operative procedures with surgical measures gaining popularity in the 1970s. The 1980s saw the beginning of embolisation but it was not until the 1990s that the TIPS procedure was readily available, with 12.5 % of patients being managed in this way. This figure rose to 24.1 % in the 2000s, highlighting recognition of the success seen with this minimally invasive procedure. These changes are shown in Table 8.

Discussion

It is estimated that some 3–5 % of all patients who have the dual pathology of portal hypertension and stoma will develop significant morbidity associated with bleeding from parastomal varices. In 1968, Resnick et al. [4] were the first to observe this clinical entity, which developed as a complication of colonic bypass surgery for chronic hepatic encephalopathy in 3 out of 19 patients with hepatic cirrhosis. Since then it has been reported sporadically in the literature, usually as isolated case reports [5–15] or small case series [16, 17]. Three publications were literature reviews [18–20]. Two of the articles referred to small series of parastomal varices observed during controlled trials of

**Table 8** Changes in the management of parastomal varices over the last five decades

	NOT	LOT	SPSS	TIPS	Embolisation	Liver transplant	Total
1960s	8	0	0	0	0	0	8
1970s	16	1	1	0	0	0	18
1980s	32	8	2	0	2	1	45
1990s	46	0	0	7	2	1	56
2000s	37	4	0	14	3	0	58
Total	139	13	3	21	7	2	185

NOT non-operative treatment, LOT local operative treatment, SPSS surgical portosystemic shunt, TIPS transjugular intrahepatic portosystemic shunt

surgical colonic bypass in chronic hepatic encephalopathy [4] and medical treatment of primary sclerosing cholangitis (PSC) [21]. There were neither randomised controlled trials of therapeutic modalities of parastomal varices nor meta-analyses pertaining to this topic. Naturally we encountered a lack of uniformity between publications with respect to selected variables and quality of data available, which led to inevitable limitations of our study. The articles were published from the 1960s on which made the practicalities of contacting authors for further information somewhat difficult. We did not find particular problems with heterogeneity of the primary endpoint as all articles were focussed on definitive control of variceal bleeding. However, heterogeneity was a problem when we examined the secondary endpoints, hence the decision to discount infrequently reported factors from statistical analysis.

The aetiology of underlying liver disease in patients who develop parastomal varices is varied. Cirrhosis is the most common cause, frequently related to alcohol. It is also recognised that a high proportion of these patients suffer from PSC which itself can be associated with ulcerative colitis (UC). Up to 15 % of all patients with UC, develop PSC in their lifetime [22]. Fucini reported a 53 % incidence of bleeding stomal varices in individuals who suffer from PSC related to ulcerative colitis [23].

Formation of the stoma offers an unusual anatomical location for the development of abnormal communications between the high pressure portal system (via mesenteric vessels) and the relatively low pressure systemic circulation (via veins of the abdominal wall). The resultant parastomal varices can bleed with only minimal trauma to the mucocutaneous junction of the colostomy or ileostomy. Bleeding is often self-limiting and managed by patients in the community by simple orthostatic measures or gentle pressure. Occasionally, it can become severe enough to prompt presentation to the emergency department. Cases of death due to exsanguination from parastomal varices have also been reported [24].

Diagnosis is challenging in these patients as often the acute episode has stopped by the time of physical examination by a clinician and endoluminal examination of the bowel is negative in most cases. It is important to remove the stoma appliance to examine the surrounding skin. Approximately, 25 % of patients will exhibit the classic bluish discolouration of the skin and/or peristomal *caput medusae*. Doppler ultrasound and contrast-enhanced CT can be useful in the diagnosis [25], as can portal vein venography [26], but venous phase mesenteric angiography offers the best diagnostic accuracy and, therefore, should be the investigation of choice [27–30].

Management of this condition has evolved since its first description more than 40 years ago. Initially local conservative and surgical measures were tried—pressure, cautery,

sclerotherapy, drugs such as propranolol, suture ligation and resiting of stoma [31–38]. Unfortunately, the rate of re-bleeding following the above treatments is high and many patients will undergo multiple procedures with limited success. With the benefit of hindsight, we can now say with certainty that these measures do not offer a cure but merely a temporary solution, as they do not address the underlying problem of portal hypertension. Exploratory laparotomy has been reported infrequently and was negative in each case, sometimes resulting in patients dying in the perioperative period. Surgical portosystemic shunting was appealing only because of the fact that this cohort of patients tends to consist of poor surgical candidates [23, 39–43], and it is not without its operative risks [44]. A group at the Mayo Clinic recognised this problematic condition and formed an ileo-anal pouch in 40 cases, thus avoiding a stoma entirely. Although 55 % of patients did experience some kind of long-term complications after ileo-anal pouch formation, there were no cases of perianastamotic ileal varices and no cases of bleeding [45].

The evolution of minimally invasive endovascular techniques has greatly influenced the management. Thus in 1989, Samaraweera et al. [46] were the first to publish 4 cases of successful transhepatic embolisation of bleeding stomal varices. Over the ensuing three decades, embolisation has continued to be used with good results [47–49]. A proportion of these patients do re-bleed after embolisation because the abnormal variceal veins can re-canalise [50–52].

Eight years later, TIPS was introduced to manage bleeding from parastomal varices [53]. Since then no fewer than eleven authors have reported their positive experiences using TIPS with or without simultaneous embolisation [54–64]. Our critical review also suggests that TIPS is by far the most effective modality for managing bleeding from parastomal varices as it ultimately reduces the portal pressure. The procedure has its limitations being unsuitable for patients with primary and metastatic liver cancer. In these patients, a technique of direct percutaneous embolisation of parastomal varices with glue or coils has been used under ultrasound or fluoroscopic guidance [34–36, 65].

It must be noted that none of the above techniques address the primary pathological liver condition causing the portal hypertension. In that respect, only liver transplantation offers the gold standard treatment to these patients. However, the reality shows that just over 5 % of all patients are suitable candidates [21, 66].

## Conclusions

Parastomal varices tend to be more frequent in men manifesting with bleeding in the fifth decade of life. They are

more likely to be associated with an ileostomy after surgical treatment of ulcerative colitis. Liver cirrhosis is the most common cause of portal hypertension leading to the development of parastomal varices and primary sclerosing cholangitis is in the second place. A third of patients with parastomal varices also have co-existent oesophageal varices. There are no pathognomonic physical symptoms or signs of parastomal varices and only the minority of patients have a raspberry appearance of the stoma, visibly dilated submucosal veins and bluish discoloration and hyperkeratosis of the skin around it. Venous phase contrast angiography or portal venography are the most helpful radiological investigations to confirm the diagnosis. TIPS has the highest success rate in preventing recurrent haemorrhage and local measures, either non-operative or surgical, are the least effective. Surgical portosystemic shunting or embolisation alone leaves patients with approximately 50 % chance of re-bleeding. Although the TIPS procedure has gained popularity over the last two decades, almost three quarters of patients with parastomal varices are still treated with local measures as first-line management. Liver transplantation as a treatment of the primary cause of portal hypertension remains very rare.

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**Conflict of interest** None.

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