

# Stoma-Related Variceal Bleeding: An Under-Recognized Complication of Biliary Atresia

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● The medical records of 52 children with biliary atresia treated by portoenterostomy and evaluated for liver transplantation were reviewed to determine the frequency of stoma variceal bleeding and the optimal strategies for prevention and treatment. Eighteen patients had had prior stoma closure, four by preperitoneal closure without takedown from the abdominal wall. Three of the four developed occult variceal bleeding from the stoma closure site. Twenty-two patients had a stoma present at evaluation. All 22 patients with stomas (100%) had at least one bleeding episode requiring transfusion. Treatment included transfusion and local pressure (9), suture ligation of the bleeding site (5), and stoma closure and/or takedown (11). Local treatment led to recurrences in eight of 14 (57%) of the cases. To reduce the high mortality in patients with biliary atresia awaiting liver transplantation, multiple variceal bleeding episodes should be prevented. To eliminate one source, stoma variceal bleeding, the stoma, whether functioning or nonfunctioning should be taken down and closed. Preperitoneal closure alone does not prevent stoma bleeding.

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**INDEX WORDS:** Biliary atresia; portal hypertension; stoma varices; liver transplantation; portoenterostomy.

**P**ATIENTS with failed portoenterostomy develop progressive liver disease and die unless liver transplantation can be provided. Seventy-three percent to 90% of children with successful portoenterostomy develop significant biliary cirrhosis with portal hypertension.<sup>1-4</sup> Ultimately, many of these patients also become liver transplant candidates. Unfortunately, 27% of pediatric liver transplant candidates die while awaiting transplantation.<sup>5</sup> Repeated episodes of variceal bleeding contribute to this high mortality. Prevention of bleeding from portal hypertension may decrease the high pretransplant death rate. Although esophageal varices are the most common source of blood loss, bleeding from portosystemic collaterals between the portoenterostomy stoma and the abdominal wall is also frequent, serious, and preventable. The purpose of this study is to evaluate a group of children with advanced liver disease following portoenterostomy to determine the incidence of stoma variceal hemorrhage, the optimal management of this complication, and the outcome of treatment.

## MATERIALS AND METHODS

The medical records of 52 children with biliary atresia treated by portoenterostomy and evaluated for liver transplantation from March 1981 to July 1985 had adequate information for this review. Twelve patients without stomas were eliminated from the study. The

records of the remaining 40 children were analyzed for the following: status of the stoma (open or closed), type of stoma closure, number of bleeding episodes, methods of diagnosis, type of treatment, and outcome of treatment. A significant bleeding episode was defined as blood loss requiring transfusion of at least 10 mL/kg of packed red blood cells. The average age at portoenterostomy was 2.7 months.

## RESULTS

### Open Stomas

Twenty-two patients had a biliary stoma present at the time of transplant evaluation. All had at least one severe bleeding episode. The stoma was nonfunctioning in 15 of 22 (68%) cases. The average time from portoenterostomy to onset of bleeding was 18.7 months (range 1 month to 9 years). Treatment (Table 1) consisted of transfusion and local pressure to the bleeding site in nine (41%) patients, suture ligation of a bleeding varix in five (23%) patients, and stoma takedown and closure in eight (36%) patients. Local treatment failed to control bleeding in eight of 14 (57%) patients. Bleeding continued until the patient either underwent transplantation or died. Stoma closure and takedown successfully controlled the stoma hemorrhage in all cases. There was one death directly related to a bleeding stoma. The patient had four episodes of stoma hemorrhage treated by local pressure and transfusion during the first two bleeds and suture ligation of the bleeding varix during the third. The final bleeding episode led to consumptive coagulopathy and death at age 2 years, 8 months.

### Closed Stomas

Eighteen patients underwent stoma closure prior to transplant evaluation. Four had a preperitoneal closure without takedown from the abdominal wall. Occult intestinal variceal bleeding from the stoma closure site occurred in three of these four patients. The bleeding site was identified by arteriogram in two cases (Fig 1) and by radionuclide bleeding scan in one case. One

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Table 1. Treatment of Bleeding Open Stomas

Type of Treatment		Recurrence of Bleeding
Transfusion and local pressure	9 (41)	8 (62)
Suture ligation of varix	5 (23)	
Stoma closure and takedown	8 (36)	
Total	22	0

child had intermittent bleeding episodes over a 7-year period attributed to esophageal variceal hemorrhage. A mesenteric angiogram ultimately demonstrated the preperitoneal stoma closure site as the source of the occult bleeding. Bleeding was successfully controlled in all three patients by detaching the closed stoma from the abdominal wall, dividing the portosystemic connections, and placing the intestinal segment intraperitoneally.

#### DISCUSSION

It has become increasingly evident that portal hypertension is a major late complication in both functioning and nonfunctioning portoenterostomy patients. Lilly initially reported that variceal hemorrhage was unusual following successful operation for biliary atresia.<sup>6</sup> More recently, he revised this opinion and reported a 23% incidence of variceal hemorrhage

in patients with sustained bile drainage.<sup>7</sup> Gautier et al<sup>8</sup> reported the 5-year evaluation of 20 cases of extrahepatic biliary atresia with functioning portoenterostomies. Definite biliary cirrhosis was noted by serial liver biopsy in 18 of 20 patients. Portal hypertension was confirmed by the presence of esophageal varices in 16 of 20 patients. Four of the 20 children (20%) bled profusely from esophageal varices.

Stoma variceal hemorrhage is a problem unique to patients who have had portoenterostomy for biliary atresia. Andrews et al reported significant stoma bleeding that necessitated stoma closure before 1 year of age in 12 of 40 (30%) patients.<sup>9</sup> In this report, all patients with open stomas developed significant bleeding and one patient died.

Of the 22 stomas present at evaluation, 15 were nonfunctional. Since bleeding from this site is almost inevitable and the nonfunctional stoma serves no physiologic function, closure and takedown is recommended.

The management of a functioning stoma in a patient with advanced liver disease deserves reevaluation. Burnweit and Coln<sup>10</sup> reported no difference in incidence of cholangitis between 12 children with diverting stomas and portoenterostomies and 19 children with nondiverting portoenterostomies. Four of the 12 chil-



Fig 1. Angiogram demonstrating varices at preperitoneal stoma closure site.

dren with stomas developed stoma hemorrhage and two required stoma closure to control the bleeding. In our series seven of 22 stomas were functional and all bled.

We recommend takedown and stoma closure in patients with advanced liver disease following portoenterostomy for biliary atresia whether the stoma is functioning or not. An aggressive approach to the prevention and treatment of stoma varices is warranted as more patients with advanced liver disease following

portoenterostomy proceed to successful liver transplantation. Preperitoneal closure as recommended by Lilly and Altman<sup>11</sup> should not be used in these patients. The persistent connection of the closed intestine to the abdominal wall allows for continued portosystemic connection. A high risk of subsequent variceal hemorrhage at the stoma site results. This source of bleeding is particularly difficult to diagnose because of its preperitoneal location.

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