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# Portosystemic Shunt Surgery in Patients with Idiopathic Noncirrhotic Portal Hypertension

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Statistical Analysis C
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**Background:** Idiopathic noncirrhotic portal hypertension (INCPH) is a rare disease characterized by increased portal venous pressure in the absence of cirrhosis and other causes of liver diseases. The aim of the present study was to present our results in using portosystemic shunt surgery in patients with INCPH.

**Material/Methods:** Patients who had been referred to our Liver Transplantation Institute for liver transplantation and who had undergone surgery from January 2010 to December 2015 were retrospectively analyzed. Patients with INCPH who had undergone portosystemic shunt procedure were included in the study. Age, sex, symptoms and findings, type of portosystemic shunt, and postoperative complications were assessed.

**Results:** A total of 1307 patients underwent liver transplantation from January 2010 to December 2015. Eleven patients with INCPH who did not require liver transplantation were successfully operated on with a portosystemic shunt procedure. The mean follow-up was 30.1±19 months (range 7–69 months). There was no mortality in the perioperative period or during the follow-up. Two patients underwent surgery again due to intra-abdominal hemorrhage; one had bleeding from the surgical site except the portacaval anastomosis and the other had bleeding from the h-graft anastomosis. No patient developed encephalopathy and no patient presented with esophageal variceal bleeding after portosystemic shunt surgery. Shunt thrombosis occurred in 1 patient (9.9%). Only 1 patient developed ascites, which was controlled medically.

**Conclusions:** Portosystemic shunt surgery is a safe and effective procedure for the treatment of patients with INCPH.

**MeSH Keywords:** **Ascites • Gastrointestinal Hemorrhage • Hypertension, Portal • Liver Transplantation • Portasystemic Shunt, Surgical • Splenorenal Shunt, Surgical**

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## Background

Portal hypertension is defined as an increase in the portosystemic pressure gradient in any section of the portal venous system. Several factors contribute to this clinical status, resulting in cirrhotic or noncirrhotic diseases, which are classified anatomically based on the side of resistance to blood flow as prehepatic, hepatic, and posthepatic [1]. In the past, the common treatment for portal hypertension was surgical shunting, but this has changed in the last 30 years [2,3]. Patients with portal hypertension and cirrhosis are candidates for liver transplantation (LT) because of definitive therapy and long-term survival. Idiopathic noncirrhotic portal hypertension (INCPH) is a rare disease characterized by increased portal venous pressure in the absence of cirrhosis and other causes of liver diseases and portal vein thrombosis. Treatment of underlying portal hypertensive symptoms is essential in the management of INCPH. Observation, medical control, transjugular intrahepatic portosystemic shunt (TIPS), embolization of varices, and portosystemic shunting are the therapeutic approaches used. Surgical shunts show a transition curve in the management of noncirrhotic portal hypertension and have become the preferred procedure when treating patients who are unable to undergo medical therapy [4,5]. In this article, we present our results using portosystemic shunt surgery in patients with INCPH.

## Material and Methods

Patients who had been referred to our Liver Transplantation Institute for LT and who had undergone surgery from January 2010 to December 2015 were retrospectively analyzed. Cirrhotic and suspicious cases were excluded from the analysis. Patients with INCPH who underwent the portosystemic shunt procedure were included in the study. Cases were screened from the electronic database of the hospital. Age, sex, symptoms and findings, type of portosystemic shunt, and postoperative complications were noted. Diagnostic methods such as abdominal ultrasonography, computed tomography, upper gastrointestinal endoscopy, and liver biopsy were used.

## Results

A total of 1307 patients underwent LT from January 2010 to December 2015. Eleven patients with INCPH, who did not require LT, successfully underwent the portosystemic shunt procedure. Seven were women and 4 were men and the mean age was 31.8 ± 11.4 years; the age range was 10 to 48 years. The preoperative biochemical parameters of the patients are shown in Table 1. The liver biopsies of all of the patients were reported as noncirrhotic. Five patients underwent side-to-side portacaval anastomosis (Figure 1) and 4 patients underwent

**Table 1.** Preoperative biochemical parameters of the patients.

| N=11 | Mean ± standard deviation | |
|---|---|---|
| Aspartate aminotransferase | 32.6±21.0 | U/L |
| Alanine aminotransferase | 35.3±26.8 | U/L |
| Bilirubin total | 1.0±0.5 | mg/dl |
| Alkaline phosphatase | 125.7±72.3 | U/L |
| Gamma-glutamyl transpeptidase | 88.2±105.9 | U/L |
| Albumin | 3.8±0.4 | g/dl |
| Prothrombin time | 78.2±14.6 | % |

**Figure 1.** Intraoperative view of a side-to-side portacaval anastomosis.

H-graft portacaval shunt with allogeneic venous graft. Two patients without ascites underwent distal splenorenal shunt procedure. The mean hospitalization time was 12±8 days and ranged from 5 to 32 days. The mean follow-up was 30.1±19 months (range 7–69 months). There was no mortality in the perioperative period or during the follow-up. Two patients underwent surgery again due to intra-abdominal hemorrhage. One had bleeding from the surgical site but not from the portacaval anastomosis and the other had bleeding from the H-graft anastomosis. No patient developed encephalopathy and no patient presented with esophageal variceal bleeding after the portosystemic shunt surgery. Shunt thrombosis occurred in 1 (9.9%) patient with H-graft portacaval shunt. Only 1 patient who underwent the distal splenorenal shunt procedure developed ascites, which was controlled medically (Table 2).

## Discussion

Portosystemic shunt surgery is a treatment option in patients with portal hypertension who do not require LT. INCPH is a rare disease that accounts for 3–5% of all portal hypertension



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**Table 2.** Characteristics and clinical features of patients with INCPH.

| Patients | *n* |
|---|---|
| **Total** | 11 |
| Female | 7 |
| Male | 4 |
| Mean age | 31.8 |
| **Clinical presentation** | |
| Upper gastrointestinal variceal bleeding | 7 |
| Chronic ascites | 3 |
| Hypersplenism | 1 |
| **Signs** | |
| Esophageal varices | 11 |
| Splenomegali | 10 |
| Ascites | 9 |
| **Procudures** | |
| Side-to-side portocaval shunt | 5 |
| H-graft portocaval shunt | 4 |
| Distal splenorenal shunt | 2 |
| **Outcomes** | |
| Mortality | 0 |
| Postoperative encephalopathy | 0 |
| Ascites | 1 |

cases [6]. The management of patients with noncirrhotic portal hypertension involves the control of variceal bleeding and symptoms due to hypersplenism. If liver function is well-preserved and pharmacological and endoscopic treatment is not successful in preventing variceal hemorrhage, portosystemic shunting is the best treatment option [4,7,8].

In the present report, 11 patients with INCPH were treated with a portosystemic shunt procedure. In all patients, the liver biopsy results were negative for cirrhosis. There were 9 cases with ascites who underwent portacaval shunt surgery. Five patients underwent side-to-side portacaval anastomosis. Four patients underwent H-graft portacaval shunting because of anatomical restrictions. The remaining 2 patients without ascites underwent the distal splenorenal shunt procedure. In the early postoperative period, 2 patients underwent surgery again due to intra-abdominal hemorrhage from the surgical sites. All patients were discharged with complete recovery. No variceal bleeding was observed after the surgical shunt procedures.

Nicolai Eck performed the first portosystemic shunt in a dog in 1877 [9]. In 1945, Whipple [10] and Blakemore [11] presented their portosystemic shunting cases with portal hypertension. To date, a few procedures have been demonstrated to accomplish these operations, and animal studies have been recommended for studying its complications [12], but there is no single treatment approach for all patients. Clinical condition and status of hepatic hemodynamics are important in the selection of the appropriate surgery. When ascites is intractable to medical management, one of the varieties of side-to-side portacaval shunts should be performed, as these are the only procedures that decompress both the splanchnic viscera and the hepatic sinusoids on the side of the ascites. When anatomically possible, a direct side-to-side portacaval shunt is preferable because of its high long-term patency rate [13]. Distal splenorenal shunts are projected to continue the perfusion of portal flow through the portal vein and are aimed at decreasing the risk of portosystemic encephalopathy. Postoperative ascites is more common after distal splenorenal shunt procedures [14]. To date, only 1 of our patients who underwent distal splenorenal shunt surgery developed ascites. We medically controlled the ascites in this patient.

TIPS has been in use for more than 30 years to treat the complications of portal hypertension [15–17]. However, the use of TIPS fell out of favor as the complication rates were reported. Boyer and Haskal [18] noted that the incidence of stenosis varies from 18% to 78%. Rosemurgy et al. [8] reported the superiority of the portacaval shunt relative to TIPS, both in terms of time to shunt failure and of survival for patients with better hepatic function.

In our study, portosystemic shunt surgery was successful in preventing esophageal variceal bleeding. No complication was seen due to surgical management after discharge. We believe that portosystemic shunting is preferable in patients with portal hypertension when LT is not envisioned.

## Conclusions

Portosystemic shunt surgery is a safe and effective procedure for the treatment of patients with INCPH.

### Conflict of interest statement

The authors declare that there is no conflict of interest.



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