

Correction of Extrahepatic Portal Vein Thrombosis by the Mesenteric to Left Portal Vein Bypass

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Objective: The goal of this study was to determine the effectiveness of mesenteric vein to left portal vein bypass operation (MLPVB) in correcting extrahepatic portal vein thrombosis (EHPVT) in children. The treatment of idiopathic EHPVT has been primarily palliative, whereas MLPVB restores hepatic portal flow in patients with EHPVT. **Methods:** Thirty-four children with symptomatic EHPVT underwent surgery with intent to perform MLPVB and were followed for up to 7 years. MLPVB was successful in 31 patients (91%), all of whom maintain patent vein grafts and have symptomatic relief of EHPVT in follow-up. All patients had complete relief from gastrointestinal bleeding. Patients with hypersplenism had significant increases in platelet and leukocyte counts and reduction in spleen size. Superior mesenteric vein flow increased from 119 ± 66 mL/min before bypass to 447 ± 225 mL/min ($P < 0.0001$) after surgery. Postoperative blood flow in the bypass graft expressed as a fraction of calculated ideal portal flow for size correlated inversely with age ($P < 0.001$). Left-portal vein diameter increased from 2.6 ± 1.6 mm to 7.3 ± 2.4 mm 2 years after surgery ($P < 0.002$). Liver volume increased from 703 ± 349 cm³ to 799 ± 351 cm³ 1 week after surgery ($P < 0.001$). Prothrombin time improved to normal in all patients 1 year after surgery.

Conclusions: MLPVB provides excellent relief of symptoms in children with idiopathic EHPVT and results in liver growth and normalization of coagulation parameters. This surgery is corrective and should be done at as early an age as possible.

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Idiopathic extrahepatic portal vein thrombosis (EHPVT) is an important cause of chronic portal hypertension in children.^{1,2} The traditional approach to the treatment of EHPVT has been palliative. It includes sclerotherapy and band ligation of gastroesophageal varices, splenectomy for progressive hypersplenism, and portosystemic shunting in older children when other measures fail to produce sustained resolution of symptoms.^{3–8}

The mesenterico-left-portal vein bypass (MLPVB) was first described as a procedure to correct EHPVT occurring after liver transplantation.⁹ Blood from the superior mesenteric vein (SMV) was redirected to the intrahepatic portal system via the left portal vein within the Rex recessus. We and others have subsequently shown the feasibility of MLPVB in children with idiopathic EHPVT.^{10–15} In this report, we show that the operation can be successfully done in most of children with idiopathic EHPVT and provides for physiologic correction of EHPVT rather than simple palliation.

MATERIALS AND METHODS

Thirty-six children were referred to our service with the diagnosis of idiopathic EHPVT from 1997 through 2003. Two children were found to have complex hepatic vascular anomalies that precluded MLPVB surgery. The remaining 34 patients underwent surgery with intent to perform MLPVB and constitute the study population.

This retrospective review of our results was done with the approval of the hospital Internal Review Board.

Preoperative Assessment

All patients had a preoperative liver biopsy. Blood work included liver function tests, complete blood counts, and coagulation parameters including coagulation factors. Patients were routinely tested for genetically determined hypercoagulable states.

Imaging studies included Doppler ultrasound examinations of the intra-abdominal vasculature, including attempts to image the intrahepatic left portal vein. Digital subtraction angiography of the celiac and superior mesenteric arteries with delayed imaging of the mesenteric veins was done in the first 6 patients. Satisfactory images of the intrahepatic portal veins were difficult to obtain by conventional angiography because of the rapid shunting of contrast away from the liver (Fig. 1A). In subsequent patients, magnetic resonance (MR) and computed tomography (CT) angiograms replaced conventional angiography as noninvasive approaches to obtain accurate images of the intrahepatic portal vasculature (Fig. 1B) and a global assessment of other large veins such as the splenic and left renal. CT or MR angiography also was used to determine liver volumes before and after successful MLPVB surgery.

When the intrahepatic portal vein was detectable by Doppler ultrasound or CT or MR angiography, the diameter of the vein was measured and recorded before and after the surgery.

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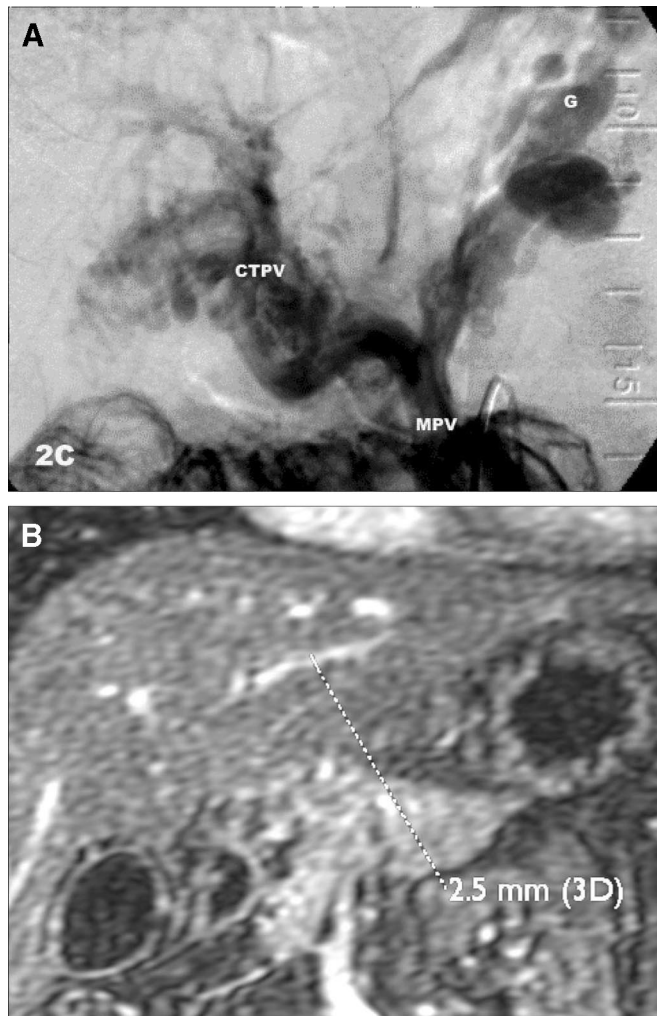


FIGURE 1. A, Conventional angiography usually failed to demonstrate the size or presence of an intact intrahepatic portal vein because the contrast filled the cavernous transformation (CVTP) in the hilum of the liver or flowed into the coronary vein (G). B, Magnetic resonance (MR) or computed tomography (CT) angiography usually demonstrated and allowed precise measurement (2.5 mm) of the portal vein diameter in the left lobe of the liver as indicated here by the dotted line.

Operative Technique

The operative technique has been described previously.¹⁴ Flow of blood in the SMV was measured using an electronic flowmeter before the vascular anastomosis and in the venous bypass graft at the completion of the operation. Pressure in the SMV was measured directly during surgery to confirm portal hypertension. Post shunt pressure was not routinely obtained to minimize the chances of technical complications from bleeding or damaging the vein graft.

Calculated Ideal Portal Venous Flow

Cardiac output was determined by multiplying the cardiac index (3–5 L/min/m²) by the calculated body surface area of the child. Hepatic blood flow was estimated as 25% of

cardiac output, and ideal portal vein flow in normal hemodynamic conditions was calculated as 75% of total hepatic blood flow. The percent of ideal portal flow achieved in the graft immediately after surgery was calculated by dividing the average of 3 blood flow measurements in the graft by the calculated ideal portal blood flow for the child's height and weight.

Statistics

Data are given as mean \pm SD. The values of individual continuous variables measured before and after MLPVB were compared for significance using Student *t* test for paired samples. Repeated-measures analysis of variance was used to determine the significance of changes in continuous variables over time. Linear correlation was used to determine the significance of the relationship of an observed continuous variable to patient age. Comparison of continuous variables between groups was performed using Student *t* test. A *P* value of 0.05 was assigned significance.

RESULTS

Thirty-four patients with idiopathic EHPVT underwent operation with the intent of performing a MLPVB (Table 1). No patient was excluded from MLPVB surgery on the basis of inability to demonstrate a usable intrahepatic left-portal vein by preoperative imaging.

Age and Gender Distribution

Mean age at surgery was 7.7 ± 4.4 years, which was an average of 3.4 years after diagnosis (range, 1 month to 10 years). The youngest patient to undergo a MLPVB was 3 months old and the oldest 14 years. There were 20 boys and 14 girls.

Presenting Symptoms

Twenty-two patients were referred for repeated severe gastrointestinal bleeding, 11 for progressive splenic enlargement and signs of advanced hypersplenism, and 1 for progressive encephalopathy following a central splenorenal shunt. Five of the children with hypersplenism also had a past history of serious gastrointestinal bleeding.

Associated Conditions

Eight patients had previous surgery for the following disorders: congenital heart disease (4), choledochal cyst (2), esophageal atresia with tracheoesophageal fistula (1), and midgut malrotation (1). Eight children had a history of umbilical vein catheterization. These 8 developed symptoms at a significantly younger age (2.0 ± 2.3 vs. 4.8 ± 3.5 years, $P < 0.05$) and underwent MLPVB surgery at a younger age (4.4 ± 3.7 vs. 8.4 ± 4.3 years, $P < 0.05$) than those with no such history. Four children had prior shunts for symptoms of portal hypertension (2 distal splenorenal shunts [DSRSs], 1 central splenorenal shunt, and 1 Clatworthy shunt), and 2 had splenectomy without a shunt.

Operative Results

Thirty-one patients (91%) maintained patent MLPVB grafts with follow-up periods ranging from 1 to 7 years. Three patients failed MLPVB because of inability to sustain flow in the bypass: 1 because of absence of intrahepatic portal vein

TABLE 1. The Characteristics of 34 Consecutive Children Operated on for Extrahepatic Portal Vein Thrombosis With the Intent of Doing the Mesenteric to Left Portal Vein Bypass

Patient ID	Gender	Age (yr)	Symptom	Previous Surgery for Hypertension	Age (yr)	Type of Shunt	Rex Open	Associated Anomalies	FPF
1	Male	1.5	Bleeding	None	2.8	Rex	Yes	Cardiac	
2	Male	10.0	Bleeding	None	10.5	Rex	Yes	None	0.8
5	Male	4.9	Hypersplenism	None	8.0	Rex	Yes	None	
9	Male	5.0	Bleeding	None	7.0	Rex	Yes	None	1.0
10	Male	3.5	Hypersplenism	None	10.0	Rex	Yes	None	0.4
11	Male	2.5	Bleeding	None	6.0	Rex	Yes	None	0.8
12	Male	6.3	Bleeding	None	9.0	Rex	Yes	None	0.9
13	Female	1.0	Bleeding	None	2.0	Rex	Yes	None	0.7
14	Male	0.3	Bleeding	None	1.0	Rex	Yes	Malrotation	0.7
16	Female	6.0	Bleeding	Splenectomy	12.0	Rex	Yes	None	0.7
17	Female	0.6	Hypersplenism	None	8.0	Rex	Yes	None	0.4
18	Male	7.0	Bleeding	None	8.0	Rex	Yes	None	0.5
19	Male	7.0	Hypersplenism	None	7.0	Rex	Yes	None	0.7
20	Female	3.5	Bleeding	Shunt	4.0	Rex	Yes	None	1.6
21	Male	9.5	Hypersplenism	None	10.2	Rex	Yes	Cardiac	0.7
22	Female	2.3	Bleeding	None	2.5	Rex	Yes	Cardiac	1.6
23	Female	0.8	Bleeding	None	2.0	Rex	Yes	None	1.5
24	Male	7.8	Bleeding	None	12.0	Rex	Yes	None	0.4
26	Female	1.5	Hypersplenism	None	2.0	Rex	Yes	TEF	0.9
27	Female	1.0	Bleeding	None	3.1	Rex	Yes	None	1.0
28	Male	2.6	Hypersplenism	None	9.6	Rex	Yes	Down's	0.8
31	Male	3.0	Hypersplenism	None	10.0	Rex	No	None	0.3
32	Female	12.0	Encephalopathy	Shunt	15.0	Rex	Yes	None	0.5
33	Male	0.3	Bleeding	None	0.3	Rex	Yes	Choledochal	1.0
34	Male	3.0	Bleeding	Splenectomy	7.0	Rex	Yes	Choledochal	0.6
35	Male	3.0	Hypersplenism	None	10.0	Rex	Yes	None	1.1
36	Male	8.0	Hypersplenism	None	11.0	Rex	Yes	None	0.5
37	Male	4.0	Bleeding	None	14.0	Rex	Yes	None	0.7
38	Female	2.0	Bleeding	Shunt	8.0	Rex	Yes	None	
39	Female	0.8	Bleeding	None	1.5	Rex	Yes	Cardiac	0.7
40	Female	6.0	Bleeding	Shunt	16.0	Rex	Yes	None	
41	Female	1.3	Bleeding	None	3.5	Rex	Yes	None	1.0
8	Male	2.0	Hypersplenism	None	10.0	Splenorenal	No	None	0.2
29	Female	3.1	Bleeding	None	7.6	Splenorenal	No	None	0.0

Thirty-three had the procedure (all except patient 29 who had no vein present in the Rex recessus), and 31 have patent bypass grafts with follow-up periods ranging from 1 to 7 years. Two MLPVB grafts thrombosed: 1 patient had a successful DSRS, and the other could not have a shunt because of widespread intra abdominal venous thrombosis. FPF indicates fraction of ideal portal flow.

arborization (patient 29, Table 1), 1 because of a thrombus in the SMV that was undetected at the time of surgery (patient 8), and in a third child because of extensive intra-abdominal venous thrombosis that precluded finding a vein with sufficient inflow to maintain graft patency (patient 31, Table 1). The first 2 of these patients underwent successful distal splenorenal shunting, while shunting was not possible in the third.

Changes in Mesenteric and Portal Vein Hemodynamics

SMV pressure before bypass was 33 ± 5 mm Hg (range, 25–44 mm Hg, normal <12 mm Hg). Figure 2 shows that flow within the SMV before bypass was 119 ± 66 mL/min, while flow in the graft just distal to the SMV anastomosis after completing the bypass was 447 ± 225 mL/min ($P < 0.0001$).

Intrahepatic Portal Vein Diameter Measurements

Preoperative portal vein measurement was documented in 23 patients. In 4, the intrahepatic portal vein could not be visualized or measured by any of the imaging techniques that we routinely used. The mean left-portal vein diameter in the remainder was 2.6 ± 1.6 mm. An example of the attenuation of the intrahepatic portal vein branches is seen in Figure 3. Rapid expansion of the left portal vein was uniformly observed immediately after successful bypass. As illustrated in Figure 4, there was a progressive increase in the left-portal vein diameter to 3.7 ± 1.7 mm 1 week after surgery, 5.4 ± 3.4 at 6 months, 6.1 ± 3.0 at 1 year, and 7.3 ± 2.4 at 2 years ($P < 0.002$). Preoperative visualization of the intrahepatic

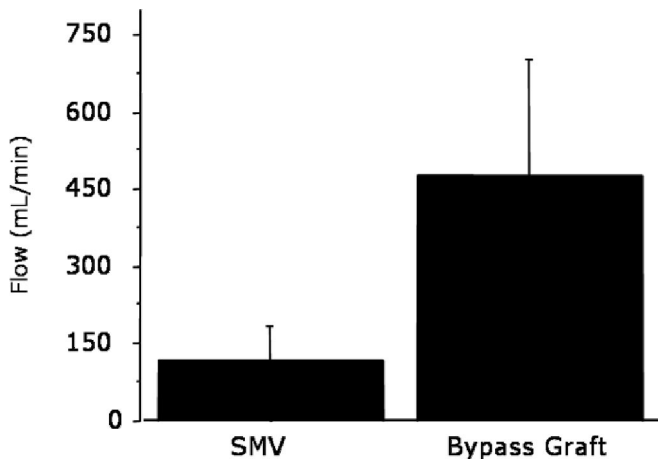


FIGURE 2. Blood flow measured by flow probe superior mesenteric vein (SMV) before mesenteric vein to left portal vein bypass operation (MLPVB) and in the vein graft into the Rex recessus (Rex flow) after completing the operation. Mesenteric flow was significantly increased by MLPVB ($P < 0.0001$).

portal vein was not essential for successful performance of MLPVB; 3 of 4 patients with no vein visible on preoperative imaging had good outcomes. In the fourth, no usable vein could be found in the liver at the time of surgery, and a MLPVB could not be done. This patient underwent an uneventful DSRS (patient 29, Table 1).

Mesenteric Flow to the Liver After Surgery

Flow in the bypass graft was measured in each patient and expressed as a percent of ideal portal blood flow for age as calculated above. The 3 patients with failed bypasses were excluded from this analysis. The average venous bypass flow in the 31 patients with successful MLPVB was 457 ± 212 mL/min, significantly less than the mean calculated ideal portal flow of 702 ± 308 ($P < 0.002$). Bypass flow expressed as a percent of ideal portal blood flow for age inversely correlated with age at surgery (Fig. 5, $P < 0.001$). Younger patients had flow that approximated or exceeded calculated ideal portal vein flow, whereas patients above age 10 infrequently obtained flows that approximated normal and often had shunt flows that were less than half of ideal calculated portal vein flow.

Two patients required a repeat abdominal exploration at 2 and 7 days after the primary operation, respectively. One child was reoperated as part of a planned delayed abdominal wall closure because visceral edema had not allowed primary closure at the first operation. A second child required a cholecystectomy 7 days after the first surgery. Flows in the grafts were remeasured on each occasion. After 7 days, flows in 1 patient had almost doubled from 450 to 750 mL/min (41%–72% of calculated ideal portal vein flow). In the second, flow increased minimally from 690 to 700 mL/min, but this child had already achieved more than 100% of ideal portal flow immediately after surgery.

Changes in Hepatic Volume

Liver volume was calculated from MR and CT images obtained before surgery and 1 week, 1 year, and 2 years after

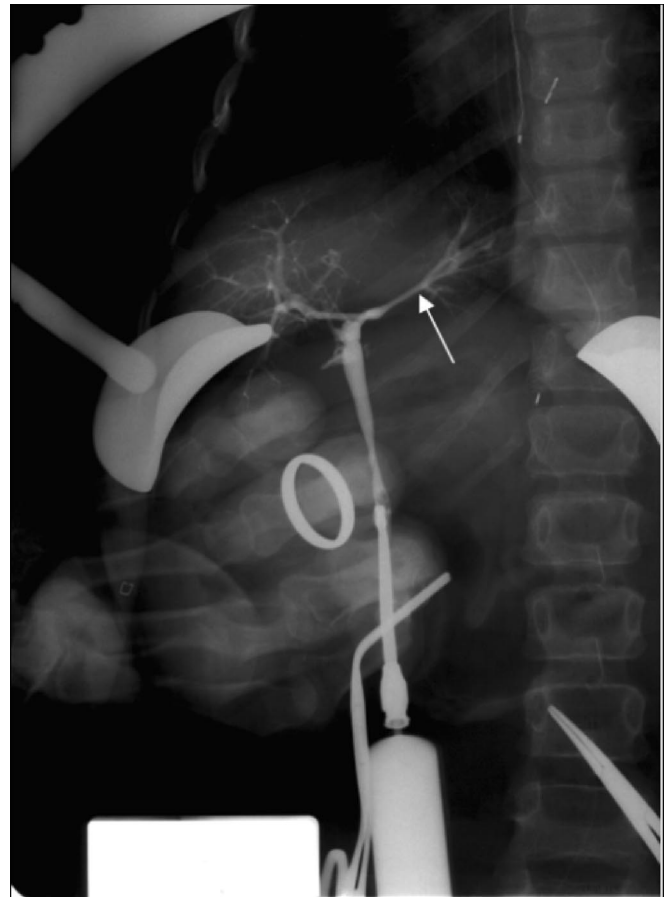


FIGURE 3. An example of the attenuation of the intrahepatic portal venous tree as observed in most of the patients in this series. This intraoperative portal venogram was obtained by injecting contrast into the patent umbilical vein. The left portal vein (arrow) measures 2 mm in diameter where the vein will be opened within the Rex recessus to perform the MLPVB anastomosis.

surgery. Mean liver volume increased significantly 1 week after surgery from 703 ± 349 cm³ to 799 ± 351 cm³ ($P < 0.001$, range of increase, 1.3–1.7 times baseline). Liver volume increased additionally but not significantly during the next 2 years ($P =$ not significant).

Resolution of Symptoms of Portal Hypertension

All 22 patients referred primarily for bleeding had patent MLPVB grafts at follow-up and complete resolution of symptoms. Of the 11 patients shunted for hypersplenism, 9 maintained patent grafts on follow-up. In this group, platelet counts rose from 48.7 ± 21.4 thou/ μ L (range, 21–86 thou/ μ L) before the shunt to 101 ± 32 thou/ μ L at 6 months ($P < 0.005$), 109 ± 41 thou/ μ L at 1 year and 112 ± 53 thou/ μ L at 2 years after bypass. Taking the entire group with successful MLPVB, excluding 2 patients with prior splenectomies and 1 with a patent proximal splenorenal shunt before surgery, platelet counts increased progressively from 54 ± 18 thou/ μ L to 148 ± 53 thou/ μ L at 6 months and to 160 ± 74 thou/ μ L at 2 years after surgery ($P < 0.0005$). Spleen size also

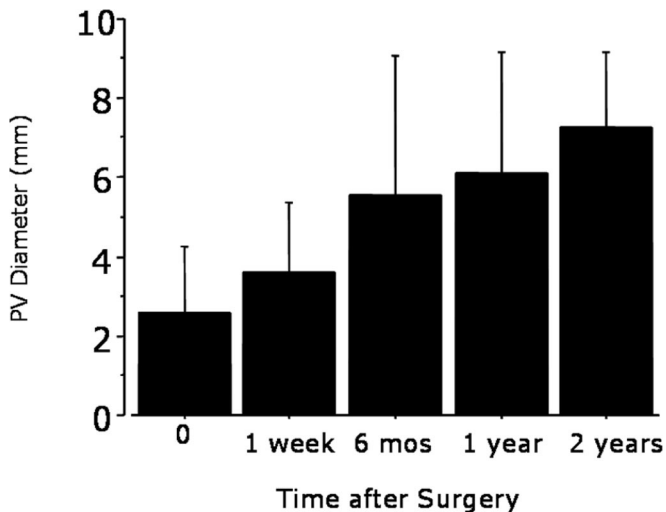


FIGURE 4. The diameter of the left portal vein diameter in the Rex recessus was measured by Doppler ultrasound. The diameter increased progressively over time after MLPVB ($P < 0.005$, ANOVA).

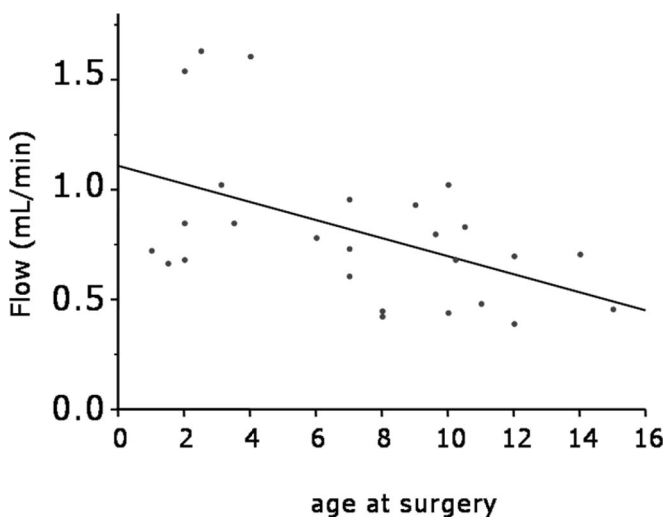


FIGURE 5. Flow in the bypass expressed as a proportion of calculated ideal portal vein flow is plotted against the age of the child at surgery. A significant inverse correlation between flow achieved and the age of the patient suggests increasingly poor compliance of the intrahepatic venous system over time ($P < 0.001$).

decreased on physical examination after bypass from 11 ± 4 cm below the left costal margin to 3 ± 1 cm by 6 months after surgery. Similarly, white blood cell counts in patients with hypersplenism rose from 2.6 ± 1.1 thou/ μL before surgery to 4.6 ± 1.1 thou/ μL at 1 year ($P < 0.05$).

Improvement in Liver Function

The prothrombin time (PT) improved in most patients following successful bypass surgery. Before MLPVB, the mean PT was 16.6 ± 1.6 seconds (range, 13.9–19.1 seconds). One year after bypass, the mean PT decreased to 14.2 ± 1.3

seconds and at 2 years had further improved to 13.7 ± 0.2 seconds ($P < 0.005$).

In 1 patient (patient 32, Table 1), successful MLPVB resulted in reversal of symptoms of advanced encephalopathy manifested by inability to concentrate and deterioration of school grades. Her blood ammonia levels also decreased from a high of 147 before surgery to normal values after the operation.

DISCUSSION

The management paradigm for children with EHPVT has evolved little over the past 2 decades. Therapeutic intervention has been palliative, aimed at controlling the symptoms of what was considered a noncorrectable lesion, and surgery has been reserved as a last resort. Even though children with EHPVT have suffered through repeated hospital admissions, transfusions, and endoscopies, the prevailing philosophy has often been that, given enough time, symptoms would disappear or become more easily managed. Major procedures such as splenectomy have also been done to alleviate some of the symptoms of portal hypertension, but not to solve the troublesome and life-threatening complications of the underlying disease. Even portosystemic shunting, selective or nonselective, converts multiple high resistance shunts ideally into a single low resistance conduit that reduces the risks of bleeding from varices, may alleviate hypersplenism, and hopefully does not reduce existing mesenteric blood flow to the liver.

This series of patients is the largest group so far treated with the MLPBV, and this experience has allowed us to glean insights into the importance of the portal circulation to the function of the liver beyond the simple resolution of the symptoms of portal hypertension. Our data show that EHPVT can now be thought of as a correctable condition. The obstruction in the extrahepatic portal vein can be bypassed in the majority of patients, and the normal flow of mesenteric blood to the liver restored.

Long-term resolution of symptoms has been excellent. Thirty-one of 34 grafts showed excellent immediate flow and have remained patent with follow-up periods as long as 7 years. All 22 patients with bleeding from the gastrointestinal tract as the principal symptom have experienced complete resolution of gastrointestinal bleeding. In all patients, blood counts have returned to normal or near-normal levels, and spleen size has substantially decreased. It is apparent from this extended series that MLPVB provides excellent control of the symptoms and complications of portal hypertension in patients with idiopathic EHPVT.

Our initial experience with this procedure in 5 patients¹⁴ led us to conclude that our inability to image a patent intrahepatic left portal vein did not necessarily mean that a suitable vein was not present. This observation has been reinforced in our more extended series. Satisfactory imaging of a patent intrahepatic portal tree before surgery has been considered an important factor in determining a successful outcome of MLPVB surgery.^{10,12} In our series, no patient was excluded from MLPVB surgery because of adverse findings on preoperative imaging. Three of 4 patients in our series had successful venous bypass surgery despite the inability of imag-

ing techniques to detect a vein in the Rex recessus. The single child in our series who was suspected by imaging and confirmed at surgery to have an absent intrahepatic left portal vein underwent an immediate DSRS. Many patients were reported to have rudimentary or very small veins measuring only 1 or 2 mm in diameter in whom very usable vessels were found at exploration. It is probable that, in the future, imaging techniques will improve to the point where small venous structures with low flow states will be more accurately imaged and assessed. However, for the present, the absence of a detectable intrahepatic left portal vein does not constitute a contraindication for MLPVB surgery at our institution. This is an important deviation from the practice described in the literature, in which a significant proportion of patients were not offered MLPVB because of unfavorable portal vein imaging.^{10,12,13} Preoperative imaging of the portal vein is valuable for surgical planning, but its findings should not exclude children from MLPVB surgery.

The ability of the portal venous system to adapt to restored flow was age-dependent in our series. Younger patients were more likely to achieve bypass flow that approximated ideal portal venous flow, and the fraction of ideal flow achieved was inversely proportional to age. It can be postulated that deprivation of blood flow results in atrophy of the intrahepatic portal veins that is progressive over time. After successful MLPVB, recruitment of blood from high-resistance collaterals to the lower-resistance hepatic vascular bed occurs immediately as demonstrated by the abrupt increase in mesenteric flow after opening the bypass. A second phase of adaptation occurs more gradually as demonstrated by the increase in graft flow in our patients in whom flow was remeasured and by the progressive expansion in portal vein diameter documented by imaging after surgery. Irreversible changes in older children may decrease the compliance of the portal venous bed and limit both immediate expansion as well as long-term adaptation of the intrahepatic portal veins. This observation argues for performing MLPVB as early after diagnosis as possible.

DSRSs have become the mainstay of surgical treatment in children with refractory symptoms of EHPVT,^{17,18} although the need for surgery has decreased since endoscopic control of bleeding esophageal varices has become so successful.¹⁹ Even though the DSRS has excellent results in controlling bleeding from upper gastrointestinal varices and reversing the symptoms of advanced hypersplenism,¹⁸ it has been long suspected that even selective shunting in children with otherwise normal liver function may result in subclinical manifestations of encephalopathy such as behavioral or learning difficulties.²⁰

Prior portosystemic shunt surgery is not a contraindication to MLPVB surgery. Four patients in our series had prior shunts, all but 1 of which had clotted. All 4 patients underwent successful MLPVB. Failure of portosystemic shunts in this disorder may reflect their inability to adapt to the increased mesenteric blood flow imposed by growth of relatively normal children or their failure to correct coagulation defects, in particular, the anticoagulants proteins C and S.^{4,7,15}

Our results suggest that restoration of portal flow to the liver, in addition to reversing the symptoms of portal hyper-

tension, results in growth of the liver and enhanced synthetic function. The deficiency of portal vein flow to the liver in children with EHPVT may have significant consequences on liver function that have not been previously appreciated and are yet to be fully characterized. The highly prevalent coagulation abnormalities are one manifestation of liver dysfunction in these children. This study demonstrated a consistent correction of abnormal PT after successful MLPVB that validates the results previously reported in a smaller cohort of patients in whom correction of deficient liver-dependent procoagulants and anti-coagulants was observed after restoration of mesenteric blood flow to the liver.¹⁵ The data suggest that these abnormalities are due to deprivation of the liver from portal blood because they are corrected by restoring portal flow to the liver with the MLPVB. Our findings allude to other physiologic consequences of having EHPVT from early in life. Failure to achieve hepatic mass is one consequence,²¹ as indicated by the rapid increase in size of the livers in these children after restoring portal blood flow. Liver size increased over 30% within a week of successful MLPVB and maintained the increased size at 1 year. Finally, overt encephalopathy associated with very high blood ammonia level was reversed in 1 patient. It is possible that the detoxifying functions of the liver may be compromised in others with EHPVT because the majority of mesenteric blood flow inevitably finds its way back to the heart through shunts in the retroperitoneum and around the gastroesophageal area.

CONCLUSION

MLPVB is a very effective treatment of children with idiopathic EHPVT. Surgical success and resolution of symptoms exceeded 90% in an unselected group of patients. Portal blood flow can be restored to near normal levels, particularly in younger children. Finally, our results suggest that portal vein flow may be important in regulating hepatic growth and optimizing the metabolic function of the liver, including the synthesis of liver-dependent coagulation factors. The MLPVB is the treatment of choice in all children with idiopathic EHPVT and should be performed as soon as the diagnosis is made.

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