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Isolated liver and multivisceral transplantation for total parenteral nutrition–related end-stage liver disease

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Abstract

Purpose: Total parenteral nutrition (TPN) has prolonged survival in children with intestinal failure; however, end-stage liver disease owing to TPN-induced cholestasis (ESLD-TPN) may preclude its use. ESLD-TPN is an indication for isolated liver transplantation (ILT) or multivisceral transplantation (MVT). Isolated liver transplantation for ESLD-TPN should only be considered in patients who have the potential for enteral autonomy.

Methods: We retrospectively reviewed the records of patients with ESLD-TPN who underwent ILT (n = 7) or MVT (n = 5) between 1994 and 2005. The median age at the time of transplantation was 10.0 months. Intestinal failure followed necrotizing enterocolitis (n = 3), gastroschisis (n = 3), gastroschisis with volvulus (n = 3), gastroschisis with atresia (n = 1), malrotation (n = 1), and megacystis microcolon intestinal hypoperistalsis syndrome (n = 1).

Results: Isolated liver transplant patients had a median length of small bowel of 70 cm and tolerated a median of 50% of enteral calories. The median length of small bowel in patients who underwent MVT was 29 cm, and none tolerated more than 30% of goal enteral feeds. Reduced-size (n = 5) and whole-liver (n = 2) allografts were used for patients undergoing ILT. Patients undergoing MVT received liver-small bowel-pancreas (n = 4) or liver-small bowel-pancreas-colon (n = 1). Overall patient survival was 57% in ILT (median follow-up = 25.1 months); 3 survivors are TPN independent, and the fourth patient requires TPN 3 days/wk. Patient survival was 40% after MVT (median follow-up = 13.0 months); 1 MVT patient died of abuse 16.9 months after transplant and was TPN independent at the time of death. Both survivors are TPN independent. Bilirubin levels are within normal range in all survivors.

Conclusion: Isolated liver transplantation for ESLD-TPN in the setting of intestinal failure is a viable option in patients who have the potential for enteral autonomy. Multivisceral transplantation is the only alternative in patients without the potential for intestinal recovery. Survival can be achieved in patients with ESLD-TPN, but mortality remains high for both procedures.

Intestinal failure is characterized by reduced intestinal absorption such that the maintenance of growth requires macronutrient, water, and electrolyte supplementation [1]. A

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variety of conditions can result in intestinal failure, including motility and absorptive disorders (eg, megacystis microcolon intestinal hypoperistalsis syndrome [MMIHS]), as well as surgical conditions that require extensive intestinal resections causing short bowel syndrome (eg, necrotizing enterocolitis [NEC], gastroschisis, intestinal atresia, midgut volvulus). Survival in patients with intestinal failure is dependent on the long-term use of total parenteral nutrition (TPN) [2].

The use of TPN has prolonged survival and improved the prognosis in children with intestinal failure by fulfilling nutritional requirements while the remaining intestine undergoes adaptation or compensation [3,4]. The capacity of the intestine to adapt determines the ability of the patient to achieve enteral autonomy. Although many infants experience early intestinal adaptation, chronic TPN dependence is inevitable in a large cohort of patients. These patients continue to develop and grow while on parenteral nutrition; however, TPN-associated complications may eventually preclude further use. These complications include hepatic dysfunction, recurrent intravenous catheter–related sepsis, thrombosis of vascular access sites, and recurrent episodes of severe dehydration. TPN-induced hepatic dysfunction in infants with intestinal failure ranges from asymptomatic biochemical abnormalities to steatosis, cholestasis, fibrosis, and cirrhosis with progressive liver failure (ESLD-TPN) [5]. Unfortunately, even some patients who attain enteral autonomy over time may have already sustained irreversible liver disease requiring liver transplantation for survival.

In patients with intestinal failure and chronic TPN dependence, small bowel transplantation may be considered to avoid the complications of long-term parenteral nutrition [6]. Unfortunately, because of a shortage of size-matched small intestinal allografts, the mortality rate for infants awaiting small bowel transplantation remains high [7]. Once cholestatic, fibrotic liver disease and portal hypertension develop in patients with intestinal failure, the ability to achieve enteral autonomy is further compromised because of malabsorption, portal hypertensive enteropathy, and gastrointestinal bleeding [8,9].

The only alternative to salvage patients with intestinal failure who have progressed to ESLD-TPN is liver replacement therapy, often in the context of combined liver-small bowel transplantation or multivisceral transplantation (MVT). In a subgroup of these patients, isolated liver transplantation (ILT) is a therapeutic option if there is still reasonable potential for enteral autonomy. In the setting of normalization of liver function and resolution of portal hypertension, intestinal adaptation and compensation may continue and full enteral tolerance may be achieved before cholestasis and liver dysfunction recur. Combined liver-small bowel transplantation or MVT is necessary in patients in whom adaptation is unlikely and intestinal failure is irreversible. Because experience with transplantation for the management of ESLD-TPN in intestinal failure remains limited, and, in particular, very few reports exist describing ILT as a therapeutic modality in this cohort [10-12], we examined our experience with ILT and MVT in intestinal failure patients with ESLD-TPN.

1. Methods

We performed a retrospective analysis of patients with ESLD-TPN who underwent either ILT or MVT (liver-small bowel-pancreas allograft or liver-small bowel-pancreas-colon allograft) from 1994 to 2005 at Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio. Patients with ESLD-TPN who underwent transplantation were identified by review of the transplant database. Data extracted from medical records included demographics, etiology of intestinal failure, pretransplant small bowel length and presence or absence of the ileocecal valve, pretransplant liver function studies, pretransplant ESLD-TPN–related complications, percent of calories tolerated enterally, type of graft, patient and graft survival, postoperative complications, and posttransplant feeding tolerance and liver function studies. Institutional review board approval of the study was obtained (Studies in Pediatric Liver Transplant, May 15, 1996). Statistical comparisons between ILT and MVT groups were made using the Student t test, and differences were considered significant at P less than .05. Mean data are expressed as mean ± SEM.

2. Results

2.1. Pretransplant status

We identified 12 children with end-stage liver disease secondary to TPN-induced cholestasis who underwent either

Table 1 Pretransplant clinical data

	Isolated liver transplant	Multivisceral transplant
Total no. of patients	7	5
Median age at transplant (mo)	8.4	12.9
Mean INR	4.7 ± 1.7	2.2 ± 0.6
Mean serum bilirubin (mg/dL)	19.4 ± 3.7	22.6 ± 5.9
Mean plasma albumin (g/dL)	2.9 ± 0.3	2.4 ± 0.2
Mean platelet count	31400 ± 5500	48400 ± 10600
Mean residual small bowel length (cm)	77.3 ± 9.2	36.4 ± 11.8*
Mean enteral tolerance (% calories)	62.6 ± 9.8	6.0 ± 6.0**

INR, International Normalized Ratio.
* $P < .05$.
** $P < .01$.