Intestinal Transplantation for Patients With Short Gut Syndrome and Hypercoagulable States


INTESTINAL transplantation has become a life-saving procedure for patients with irreversible intestinal failure who can no longer be maintained on total parenteral nutrition (TPN). This is the first report to address the management policy and efficacy of intestinal transplantation as a rescue therapy for patients with intestinal failure and visceral vascular thrombosis.

MATERIALS AND METHODS
Between May 2, 1990 and August 18, 1999, a total of 130 patients received primary intestinal transplantation at our center. Fifty-five were adults and splanchic vascular thrombosis was the cause of loss of the native intestine in 17 (31%). Of these 17 patients, 8 (47%) received isolated intestine, and 9 (53%) received composite visceral grafts that contained liver (5 liver/intestine, 4 multivisceral). Twelve (71%) recipients were male and five (29%) were female with a mean age of 37 ± 13 years. The preoperative work-up included a full hypercoagulable laboratory profile and visceral angiography for all patients. The thrombosis was arterial in 12 (70.5%), venous (portomesenteric) in 4 (23.5%), and combined in the remaining case (6%). Concomitant extensive central venous thrombosis was documented preoperatively in 8 (47%) patients. The etiology of vascular thrombosis was hypercoagulable state in 9 (53%), TTP in 1 (5.8%), myeloproliferative disorder in 1 (5.8%), and undetermined in the remaining 6 (35%) patients.

The hypercoagulable syndrome was due to isolated/combined deficiency of proteins C and S and antithrombin III in all 9 patients, with concomitant factor V mutation in 2 cases. Composite visceral grafts were given only to patients with combined liver and intestinal failure and/or total vascular occlusion of the splanchic circulation. Anticoagulant therapy was continued postoperatively as a lifelong treatment for all patients with superior vena cava syndrome and those who were hypercoagulable and received isolated intestinal transplantation. The donor characteristics, full details of donor/recipient operations, and perioperative management, including immunosuppressive protocol, have been described elsewhere.

RESULTS
With a mean follow-up of 27 months (range 5 to 106), five of the composite allograft recipients died and two of the isolated intestinal recipients required graft enterectomy, with an overall patient and graft survival of 71% and 59%, respectively. The causes of death were bacterial/fungal sepsis (n = 3), cytomegalovirus (CMV) pneumonia (n = 1), and postoperative lymphoproliferative disorder (PTLD) (n = 1). The two enterectomies were performed 9 and 20 days after transplantation because of postoperative bacterial pneumonia with subsequent withdrawal of immunosuppression (n = 1) and intractable rejection (n = 1). The first patient died 21 months after graft enterectomy due to TPN-induced liver failure and the second is currently alive on TPN therapy. None of the isolated or composite visceral allografts were lost due to vascular thrombosis. All current survivors with graft in place (n = 10) are completely off TPN with full enteric nutritional autonomy.

DISCUSSION
Visceral vascular thrombosis was the common cause of irreversible intestinal failure in adult patients who received intestinal transplantation at our center. The diagnosis and etiology of the hypercoagulable state was achieved with certainty in more than half of the cases because of the recent development and availability of new hematologic, molecular diagnostic, and genetic tests.

Despite replacement of the native liver and subsequent correction of the underlying inborn metabolic errors, two of our composite visceral recipients continued to develop recurrent thrombosis of their native vessels. One possible explanation is the coexistence of extrahepatic hereditary defect at the endothelial level. Therefore, it has been our current policy to anticoagulate all of these patients for a minimum of 6 months postoperatively.

In this study, five hypercoagulable patients with chronic intestinal failure and reversible TPN-induced liver damage...
received intestinal transplantation without liver replacement. None of these patients developed recurrent vascular thrombosis with postoperative anticoagulant therapy. In conclusion, visceral vascular thrombosis is a common indication for intestinal transplantation among adult patients. The diagnosis of a hypercoagulable state does not indicate replacement of an adequately functioning native liver but mandates lifelong anticoagulant therapy, particularly after isolated intestinal transplantation.

REFERENCES