IS-27

Living-related intestinal transplantation for a patient with hypoganglionosis

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Purpose: Reporting our first case of the living-related intestinal transplantation for a patient with short-gut syndrome associated with hypoganglionosis.

Case: A 14-year-old boy with TPN-dependent short-gut syndrome associated with hypoganglionosis underwent the living-related intestinal transplantation by using a 150 cm segment of distal ileum taken from a healthy donor. The graft vessels were connected to infrarenal aorta, and inferior vena cava. The immuno-suppressive regimen consisted of daclizumab, tacrolimus, and steroid. The graft surveillance for acute cellular rejection (ACR) was accomplished using zoom endoscopy and mucosal biopsy. The blood trough level of tacrolimus maintained between 20-25 ng/ml for the first 2 months, followed by 15-20 ng/ml thereafter. The 50mg of daclizumab was administered on day of operation, and same dosage has been repeated once for 2 weeks.

Result: The first ACR occurred on POD-9 was progressive, and required a 14-day course of OKT-3 injection. After the treatment of OKT-3, the graft recovered from the ACR, and began to function well enough to discontinue the IV nutrition on POD-55. No infectious complications have occurred. The patient discharged in POD-112, and currently tolerates full oral intake without requiring IV nutritional or fluid supplementation. The donor has been discharged without any complications.

Conclusion: The living-related intestinal transplantation could successfully be performed for a patient with hypoganglionosis, and it can be a treatment of choice for patients with short-gut syndrome associated with pseudo-obstruction syndrome.