

Paediatrics

Marginal parental donors for pediatric living donor liver transplantation

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Living donor liver transplantation (LDLT) has been increasingly recognized as an alternative to deceased donor liver transplantation to mitigate waiting list mortality, especially in the pediatric population, and this procedure has flourished in the Asian countries. The current outcomes of pediatric LDLT show excellent patient survival.

A wide variety of liver diseases is indicated for LT in children, and the number of LT performed for familial hereditary liver disease cases increased over the past three decades. When living donors are selected from blood relatives in the family including the patient with an inherited liver disease, they are meticulously evaluated to prevent morbidity in both the donor and recipient.

In our experience, there was no observed mortality or morbidity related to recurrence of inherited liver diseases with the use of heterozygotes, except for relevant cases such as heterozygous mothers with ornithine transcarbamylase deficiency (OTCD), heterozygous protein C deficiency, heterozygous hypercholesterolemia, heterozygous protoporphyria, and asymptomatic parental donors with paucity of intrahepatic bile duct in Alagille syndrome (Table). Using a graft from a human leukocyte antigen-homozygous parental donor can cause graft-versus-host disease after LDLT.

The procedure of LDLT has a risk of morbidity in two family members at a time. Therefore, marginal indications or contraindications of living donors should be recognized for optimal donor-to-recipient matching.

Table: Relevant risks of using grafts from parental donors in pediatric LDLT for inherited liver diseases

Original liver disease	Inheritance pattern	Risk associated with LDLT
OTCD	X-linked	Hyperammonemia
Protein C deficiency	Autosomal recessive	Thromboembolic events
Familial hypercholesterolemia	Autosomal dominant	Hypercholesterolemia
Acute intermittent porphyria	Autosomal dominant	Porphobilinogen deaminase activity ↓
Erythropoietic porphyria	Autosomal recessive	Ferrochelatase activity ↓
Alagille syndrome	Autosomal dominant	Cholestasis