Domino liver transplantation (DLT) has been described as a method of solid organ conservation in this current time of liver graft shortage. This process takes advantage of the recipient liver with a metabolic disease to rescue a patient with liver failure from another cause. To date, there have been no published case reports regarding the use of DLT from patients who have required orthotopic liver transplant (OLT) for propionic academia (PA). The use of liver transplantation for patients with PA has been described as only ‘moderately’ successful in correcting the consequences of the enzyme1. The ‘domino’ reuse of livers affected by PA should provide hepatic rescue therapy and not produce a metabolic crisis in the recipient because of enzyme production in other sites in the recipient, specifically the kidneys and gastrointestinal tract2. In the worst case the DLT can provide a bridge to re-transplantation with an enzyme producing liver.

PA is an autosomal recessive disease classified as a branched chain organic aminoacidemia as a result of deficiency in the enzyme propionyl CoA carboxylase, with resultant metabolic acidosis and hyperammonemia3. The manifestations of the disease are systemic and early in onset; and many patients exhibit mental retardation, cardiomyopathy, and immune suppression2.

Our patient was a 4-year-old female diagnosed at birth with PA after development of a metabolic acidosis. The child was on maximal medical therapy with persistent metabolic crises despite this treatment, awaiting OLT. By coincidence, when cadaveric liver became available for the 4 year old with PA, there was another child in the pediatric intensive care unit (PICU) with fulminant hepatic failure of unknown etiology which prompted the initiation of this domino transplant. The second patient in the PICU was a 14-year-old boy carrying a diagnosis of left lower extremity epithelioid sarcoma who was recently admitted with hepatitis of unknown etiology. His course progressed to encephalopathy and fulminant hepatic failure necessitating intubation. The decision to do the domino transplantation was one of life saving necessity; he was likely to expire within the next 24 hours if he did not receive a liver transplant. He received the liver of our 4-year-old child with PA and underwent a left lower extremity below the knee amputation following the DLT.

Both patients had uncomplicated postoperative courses and are doing well. In particular, the recipient of the propionic acidemic liver has recovered astonishingly well; he is only taking amiodipine and magnesium supplementation.

The management of the recipient of the PA liver involved initially restricting protein intake as one would do in a patient with PA. His metabolic profile, particularly his urinary ketones and serum ammonia levels, was followed closely and his protein intake was ultimately liberalized. He was discharged to a rehabilitation facility tolerating a regular diet. His outpatient basis follow-up has been uneventful.

We believe this is one of the first DLT procedures utilizing a liver from a patient with PA and resulted in salvage and recovery of a patient with fulminant hepatic failure.