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4th Global Surgery and Transplantation Congress

October 03-04, 2016 Atlanta, USA

Successful combined liver-kidney transplantation for methylmalonic acidemia

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To evaluate the emerging role of combined liver-kidney transplantation (CLKT) for methylmalonic acidemia (MMA) with associated renal failure, we report a case and review the literature on outcomes of this treatment. Following successful CLKT, during the four year follow-up, our 16 year old patient demonstrated a marked reduction in serum methylmalonic acid level from a pre-transplant level of 3204.86 μmol/L to 236.46 μmol/L and complete independence from dialysis. Our patient has normal protein intake, growing normally without any further neurologic complication. The patient has graduated from a high school and is ready to study at a college. In our retrospective literature review of 10 patients undergoing CLKT for MMA, all patients were alive with adequate liver function at a mean of 15 months following CLKT. One patient did experience worsening renal function due to non-adherence with medication and restricted diet following CLKT; the other nine patients demonstrated satisfactory renal function. One patient suffered metabolic decompensation with cerebellar infarct 10 months after CLKT. Even though CLKT is not curative for MMA, the patients often gain freedom from dietary protein restriction, normal growth curve, muscular strength and avoidance of metabolic crises with overall improvement in quality of life. In conclusion, CLKT should be considered for those patients with MMA and renal failure who continue to experience episodes of metabolic decompensation in spite of optimal medical therapy. Adequate dietary protein restriction following CLKT should also be considered if serum methylmalonic acid is too high in order to minimize potential late neurologic complications.

Biography

Shi-Feng Li completed his Medical Degree and Surgery Residency from West China University of Medical Sciences. He also completed a Transplant Surgical Fellowship at Integris Health. He has authored numerous peer review articles, as well as medical textbooks. He is serving as a Liver, Kidney and Pancreas Transplant Surgeon at Oklahoma Transplant Center.

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