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Pretransplant management of polycystic kidney disease

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Autosomal dominant polycystic kidney disease (ADPKD) is the first genetic cause of end-stage renal disease (ESRD). ADPKD is a systemic disease characterized by the development of cysts in the kidney parenchyma and various extra-renal manifestations involving cysts in other organs and connective tissue abnormalities. Renal function declines slowly in patients with ADPKD, and chronic kidney disease or ESRD occur in >70% of patients and the number of these patients who receive a kidney transplant is continuously increasing over time. Kidney transplantation (KT) is the optimal choice of renal replacement therapy in appropriate patients with ADPKD. Living kidney donation, ideally preemptive, is likely to be associated with best outcomes. Issues related to native nephrectomy and screening for intracranial aneurysm (ICA) are important considerations in patients with ADPKD who are being evaluated for KT. There is no clear guidance or agreement concerning the need, indication, timing and approach for native nephrectomy in patients with ADPKD undergoing KT. In most center, routine nephrectomy before transplantation is no longer recommended as nephrectomy in ADPKD patients is associated with significant morbidity and mortality. Indications for nephrectomy include recurrent infection, symptomatic nephrolithiasis, recurrent and/or severe bleeding, intractable pain, and suspicion of renal cancer. In addition, unilateral nephrectomy in asymptomatic patients might be required at the time of transplantation if massive native kidney size causes space restriction in allograft placement, taking into account that kidney size typically declines after transplantation. Although transcatheter unilateral artery embolization has been suggested as an alternative to nephrectomy to obtain sufficient volume reduction for graft implantation, the studies were limited by the small number of patients and should be validated by adequately powered prospective clinical studies. Although practices vary widely, on average less than one third of patients in published series undergo pretransplant nephrectomy and some centers have preferred for concurrent nephrectomy at the time of KT, but both practices have not been directly compared. Simultaneous nephrectomy has clear advantages for the patient, including a single operation and freedom from pretransplant dialysis if KT is pre-emptive. Finally, the risk-benefit relationship for screening patients for ICA prior to KT remains unknown. General recommendations for the screening of ICA in patients with ADPKD should be used to select patients for pretransplant screening.