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Roy Michael Culpepper

University of South Alabama College of Medicine, USA

Advances in management of autosomal dominant polycystic kidney disease (ADPKD)

ADPKD accounts for 5-7% of patients requiring dialysis or kidney transplant worldwide. The natural course of the disease was first characterized in the 1950's. The basis of the disorder is now known to involve 2 genes, PKD1 and PKD2 that account for about 85% and 15%, respectively, of disease in affected individuals. PKD1 mutations are associated with the more rapid decline in kidney function with an average age to reach end-stage disease of 50-55 year in males and slightly longer in females. Recent data demonstrate a clear relation between the rate of cyst growth, measured as total kidney volume (TKV), and the rate of decline in kidney function, measured as eGFE. Further research implicates AVP as a key factor in the stimulation of cyst growth and increase in TKV. A number of intracellular targets to slow cyst growth have been identified and clinical studies with long-acting somatostatin analogs and with the renal AVP V2 receptor antagonist tolvaptan have been completed. Two seminal studies with tolvaptan have shown sufficiently robust effects to slow increases in TKV and declines in GFR as to warrant approval for use in ADPKD patients for prolongation of kidney function. ERA-EDTA has devised a concise outline, based on TKV and patient characteristics, to guide clinicians in the choice of patients most likely to benefit from tolvaptan.

Biography

Roy Michael Culpepper earned his MD from the University of Alabama in Birmingham (UAB) and completed post-doctoral training at Loma Linda University, California, and UAB, AL. He serves as Professor of Medicine and Nephrology at the University of South Alabama and has been Director of Nephrology and served on Board of Directors of the National Kidney Foundation and the regional dialysis Network 8.

mculpepp@health.southalabama.edu

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