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2 Current management of autosomal dominant polycystic kidney disease

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD), the most frequent cause of genetic renal disease affecting approximately 4 to 7 million individuals worldwide and accounting for 7%-15% of patients on renal replacement therapy, is a systemic disorder mainly involving the kidney but cysts can also occur in other organs such as the liver, pancreas, arachnoid membrane and seminal vesicles. Though CT and MRI were similar in evaluating 81% of cystic lesions of the kidney, MRI may depict septa, wall thickening or enhancement leading to upgrade in cyst classification that can affect management. A screening strategy for intracranial aneurysms would provide 1.0 additional year of life without neurological disability to a 20-year-old patient with ADPKD and reduce the financial impact on society of the disease. Current treatment strategies include reducing: cyclic adenosine monophosphate levels, cell proliferation and fluid secretion. Several randomised clinical trials (RCT) including mammalian target of rapamycin inhibitors, somatostatin analogues and a vasopressin V2 receptor antagonist have been performed to study the effect of diverse drugs on growth of renal and hepatic cysts, and on deterioration of renal function. Prophylactic native nephrectomy is indicated in patients with a history of cyst infection or recurrent

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