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1: [J Pak Med Assoc.](#) 2008 Jun;58(6):305-9.

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Clinical presentation and outcome of autosomal dominant polycystic kidney disease in Pakistan: a single center experience.

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OBJECTIVE: To delineate clinical presentation and outcome of Autosomal Dominant Polycystic Kidney Disease (ADPKD) in terms of need for renal replacement therapy of kidney transplantation in Pakistani patients. **METHODS:** Patients with ADPKD were identified using strict clinical criteria. Medical charts were evaluated retrospectively for initial presenting complaints, co-morbidities, family history of ADPKD, any palpable mass on abdominal examination, cardiac examination for any abnormal finding and use of any anti-hypertensive drugs. Laboratory parameters were assessed. Chi square and Logistic regression analyses at 95% CI were used for statistical significance. A p value of less 0.05 was considered statistically significant. **RESULTS:** A total of 56 patients fulfilled our criteria of ADPKD. There were 40 (71.4%) males and 16 (28.6%) females in our study. The mean age at the time of diagnosis of ADPKD was 47.0 +/- 14.5 years. Mean follow up period for all patients was 7.6 +/- 4.2 years. Most common form of presentation was hypertension in 38 (67.9%) patients. Kidneys were palpable in 33 (58.9%), liver in 16 (28.6%) and spleen in 6 patients (10.7%). Microscopic haematuria was observed in 38 (67.8%) patients while gross haematuria was present in 10 (17.9%) patients. The murmur of mitral valve prolapse was found in 10 patients on clinical examination which was later confirmed on transthoracic echocardiography (TEE). On MRI/MRA 2 (3.6%) patients each had berry aneurysm and AV malformations. Three patients (5.4%) received renal transplant and 19 (33.9%) patients were dialysis dependent at the end of study. A total 11 (20%) were lost to follow up. Two patients (3.5%) died during six years follow up. Male sex and uncontrolled hypertension were most important predictors of poor prognosis (p < 0.03 and < 0.048 respectively). **CONCLUSION:** Pre-symptomatic patients with ADPKD should be monitored with blood pressure measurements and assessment of their renal function. The advantages of such monitoring include the ability to prevent or control infection and hypertension, to identify potential kidney donors from among the family, to offer advice on marriage and childbearing, and to provide prenatal diagnosis.

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