Renal structure in early autosomal-dominant polycystic kidney disease (ADPKD): The Consortium for Radiologic Imaging Studies of Polycystic Kidney Disease (CRISP) cohort

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Background. Autosomal-dominant polycystic kidney disease (ADPKD) is characterized by gradual renal enlargement and cyst growth prior to loss of renal function. Standard radiographic imaging has not provided the resolution and accuracy necessary to detect small changes in renal volume or to reliably measure renal cyst volumes. The Consortium for Radiologic Imaging Studies in Polycystic Kidney Disease (CRISP) is longitudinally observing ADPKD individuals using high-resolution magnetic resonance (MR) imaging to determine if change in renal and cyst volumes can be detected over a short period of time, and if they correlate with decline in renal function early in disease.

Methods. Standardization studies were conducted in phantoms and four subjects at each participating clinical center. After, in the full-scale protocol, healthy ADPKD individuals 15 to 45 years old with creatinine clearance>70 mL/min underwent standardized MR renal imaging, renal iothalamate clearance, comprehensive clinical evaluation, and determination of 24-hour urinary albumin and electrolyte excretion. Stereology was used from T1-weighted images to quantify renal volume, and region-growing thresholding was used from T2-weighted images to determine cyst volume. Renal structures were evaluated in relation to demographic, clinical, and biochemical variables using means/medians, standard deviations, and Pearson correlations.

Results. Reliability coefficients for MR renal and cyst volume measurements in phantoms were 99.9% and 89.2%, respectively. In
disease severity in autosomal-dominant polycystic kidney disease (ADPKD). Magnetic resonance measurements of renal blood flow as a marker of glomerular filtration rate (GFR). Age-adjusted renal volume ($r = 0.42, P < 0.0001$), cystic ($r = 0.39, P < 0.0001$, and % cyst volume ($r = 0.41, P < 0.0001$) were related with urinary albumin excretion.

**Conclusion.** MR measures of renal and cyst volume are reliable and accurate in patients with ADPKD. ADPKD is characterized by significant cystic involvement that increases with age. Structure (renal and cyst volume) and function (GFR) are inversely related and directly related to glomerular filtration rate (GFR). Cyclic AMP promotes growth and secretion in human polycystic kidney epithelial cells. *Kidney International* 66: 3, 964-973.

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**Key words:** autosomeal-dominant polycystic kidney disease, magnetic resonance imaging, renal volume, cyst volume, renal function

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