# Multi-modal Magnetic Resonance Imaging (MRI) Provides an Accurate Assessment of Kidney Disease Severity in Children and Young Adults with Autosomal Recessive Polycystic Kidney Disease (ARPKD)

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**Introduction:** Currently, there are no clinically available therapies for ARPKD. Several novel therapies have shown promise in ARPKD animal models. Unfortunately, clinical trials of these potential therapies are limited by the lack of safe, sensitive measures of ARPKD kidney disease progression. The goal of this study was to assess the combined ability of $T_1$ and $T_2$ relaxation times to assess cystic burden, and Arterial Spin Labeling (ASL) to assess kidney perfusion to sensitively detect and stage ARPKD kidney disease.

**Methods:** We obtained kidney MRI scans for 12 ARPKD patients (6-22 years of age) and 10 healthy adult volunteers (18-59 years of age). Each subject was scanned on a Siemens 3T MRI scanner with a kidney Magnetic Resonance Fingerprinting (MRF) method to generate co-registered coronal kidney $T_1$ and $T_2$ maps in 15 seconds / imaging slice and a non-contrast Arterial Spin Labeling (ASL) MRI method to generate kidney cortical perfusion maps. To assess repeatability, ten ARPKD patients underwent a second MRI scan the following day. Two-tailed Student's t-tests were used to compare the mean kidney MRI ($T_1$, $T_2$, and perfusion) between the subjects with ARPKD and the healthy volunteers.

**Results:** Significant increases in both kidney $T_1$ and $T_2$, and reduced perfusion were observed between ARPKD patients (n=12) and healthy volunteers (n=10, $p<0.001$) as well as between ARPKD patients with early and mild-to-moderate CKD based on eGFR (above or below 90 ml/min/1.73m$^2$, n=6/group, **Fig. 1**, $p<0.03$). Repeat scans for ARPKD patients (n=10) revealed mean variation of 2.1% for $T_1$, 2.8% for $T_2$, and 16.3% for perfusion.

**Conclusions:** The multi-modal MRI biomarkers in combination evaluated in this study were able to distinctly stratify all three cohorts, including the two ARPKD patient cohorts, demonstrating the utility of a multi-modal MRI approach during future clinical trials aimed at limiting ARPKD kidney disease progression.
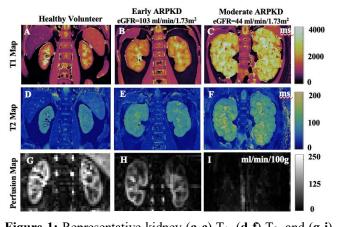
**Figure 1:** Representative kidney (**a-c**) $T_1$, (**d-f**) $T_2$, and (**g-i**) perfusion maps from a healthy volunteer and two pediatric ARPKD patients (eGFR greater than or less than 90 ml/min/1.73m$^2$, n=6/group).