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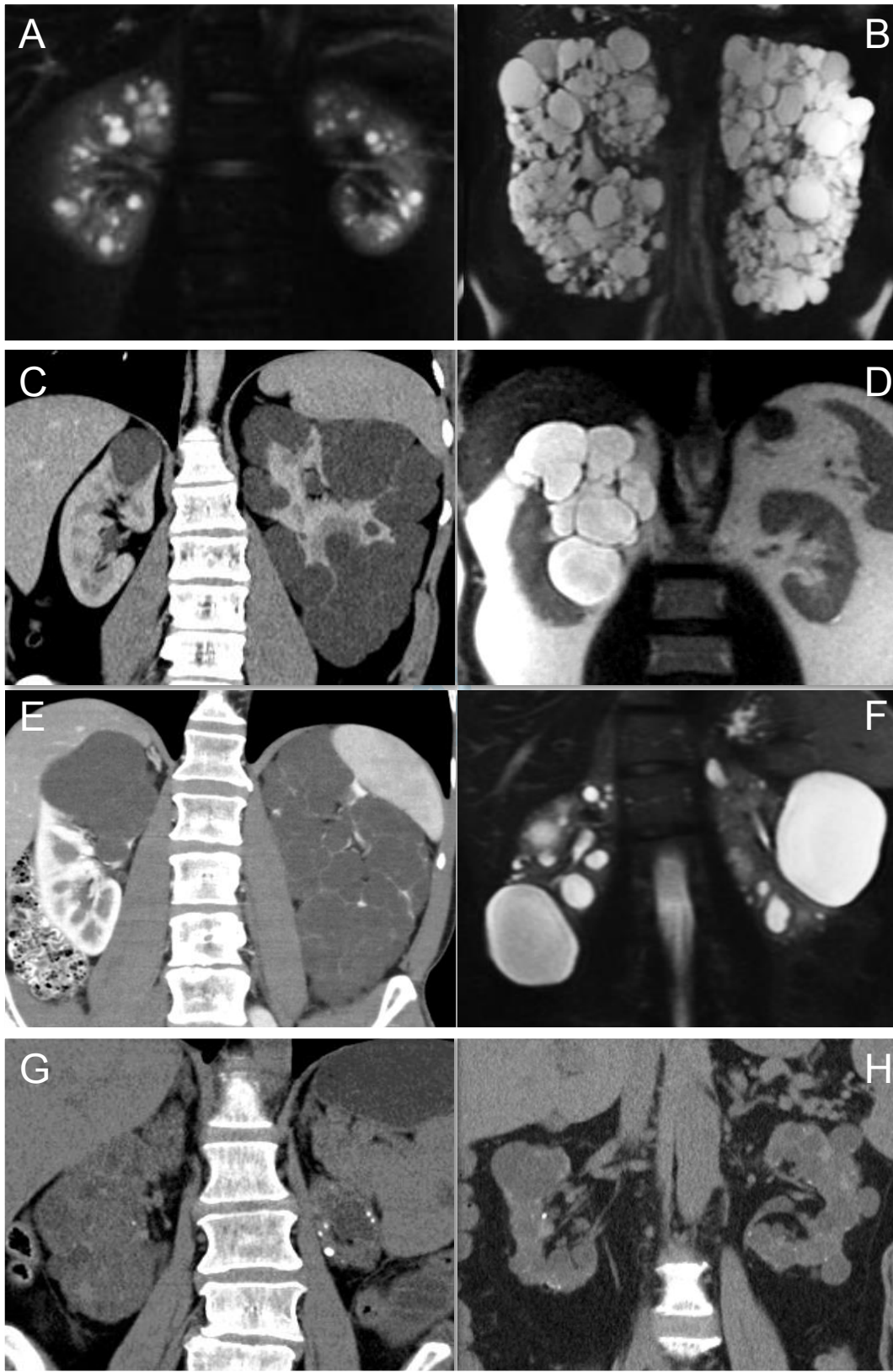### Classification of study participants into typical, Class1, and atypical, Class 2.

The patients were classified into typical (Class 1) and atypical (Class 2) cases based on pre-specified imaging

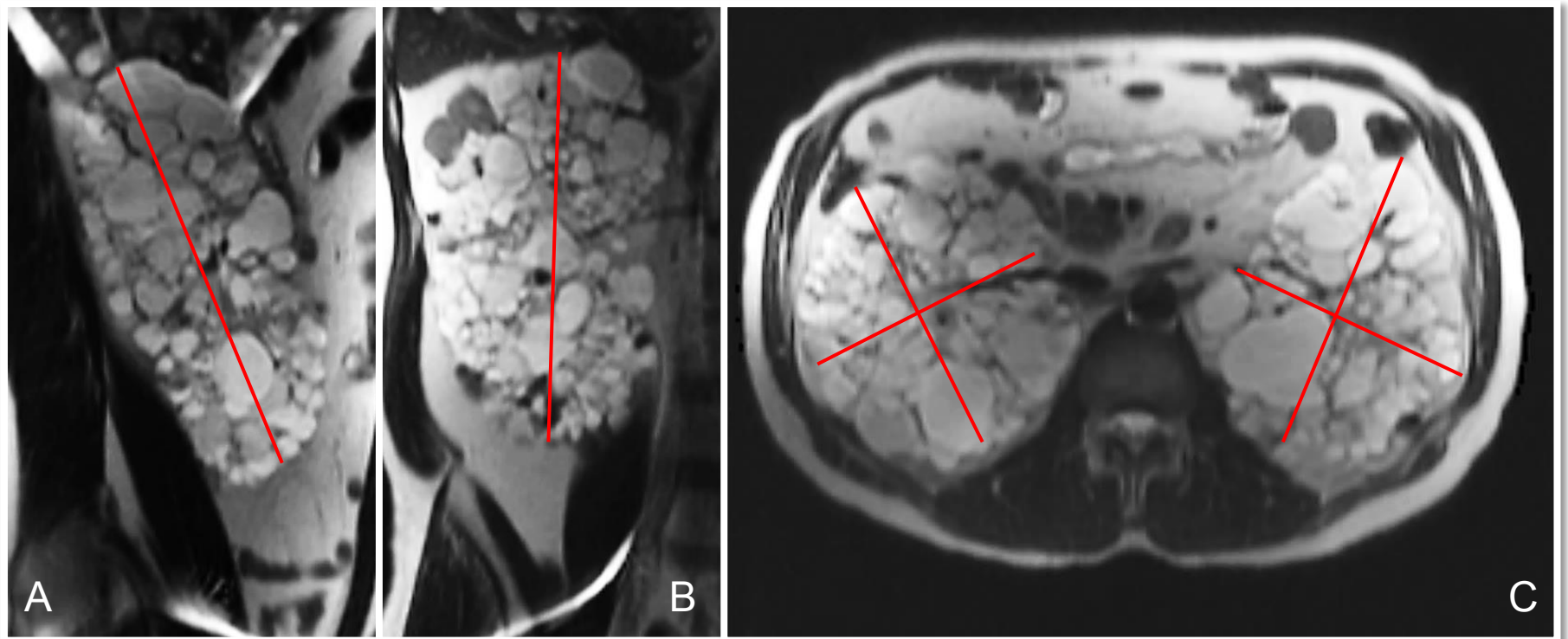findings (Supplementary Figure S1). Image classifications were performed blinded to clinical data.

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Suppl. Figure S1. Imaging characteristics of typical and atypical ADPKD patients
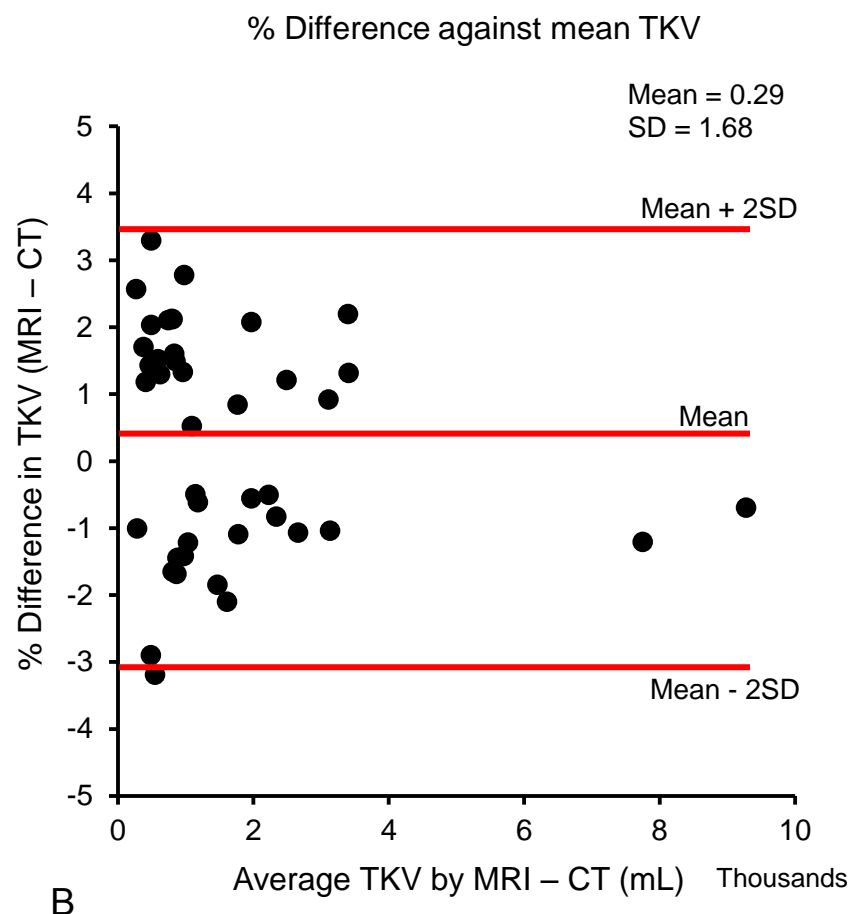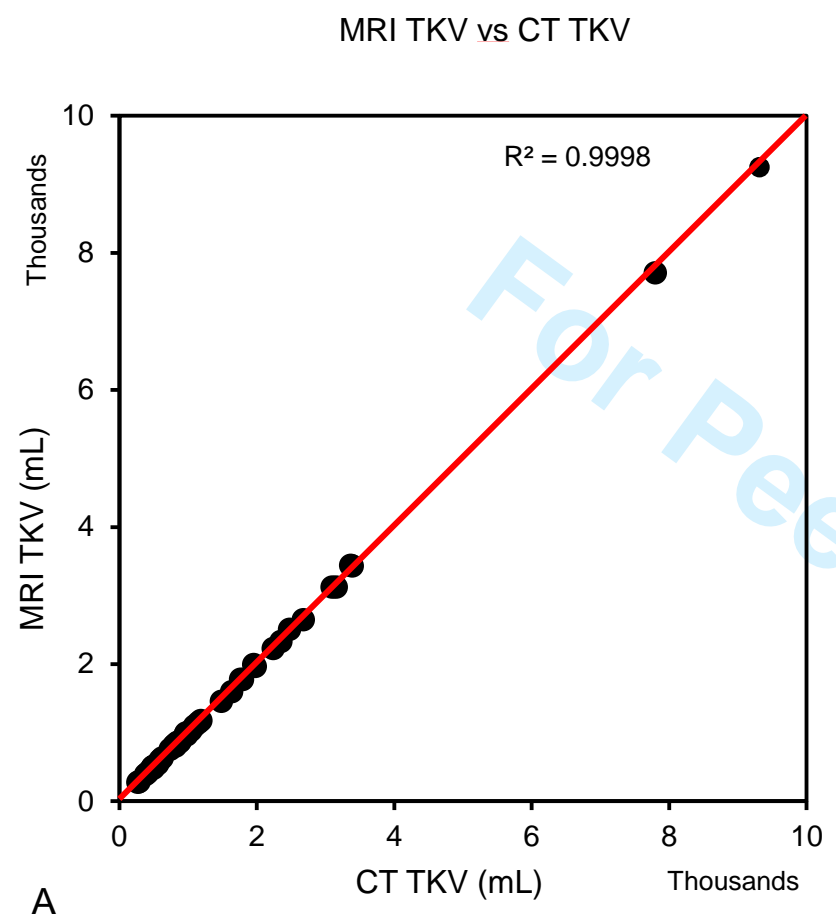

Typical (or Class 1) ADPKD patients present with bilateral and diffuse distribution, from mild (A) to severe (B) replacement of kidney tissue by cysts, where all cysts contribute similarly to the patient's TKV. Atypical (or Class 2) ADPKD includes ADPKD patients that present with unilateral (C), segmental (D), asymmetric (E), or bilateral atypical presentation (F); subclass 2A, but also includes patients that present with bilateral distribution with acquired unilateral atrophy (G) or bilateral kidney atrophy (H); subclass 2B.

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Suppl. Figure S2. Estimation of TKV from MR or CT images using the ellipsoid equation

Length was obtained from longitudinal images at the maximum longitudinal diameter, in both sagittal (A) and coronal (B) planes. Width and depth were obtained from transverse images at the maximum transverse diameter, acquired in the plane perpendicular to the longitudinal plane (C).

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Suppl. Figure S3. Comparison of TKV stereology (TKVs) measurements with MR or CT images

MRI TKV vs CT TKV

$R^2 = 0.9998$

Y-axis: MRI TKV (mL) — Thousands, scale 0 to 10
X-axis: CT TKV (mL) — Thousands, scale 0 to 10

A

% Difference against mean TKV

Mean = 0.29
SD = 1.68

Mean + 2SD

Mean

Mean - 2SD

Y-axis: % Difference in TKV (MRI – CT), scale -5 to 5
X-axis: Average TKV by MRI – CT (mL) — Thousands, scale 0 to 10

B

TKVs measurements from MR and CT images were strongly correlated (A). A plot of the % difference between the measurements against their mean (B) is more informative to assess between-measurements differences. The mean % difference in TKVs=0.29±1.68, and 42/43 measurements lay within the mean ± 2SD. The combined % measurement error was 1.53 (0.79 MR; 0.83 CT; p=NS).