Measurement and Reporting of Total Kidney Volume in Autosomal Dominant Polycystic Kidney Disease (ADPKD)

Step 1: Measurement of Total Kidney Volume (TKV)

Renal volume is calculated by using the ellipsoid formula:
Volume = length × width × thickness × π/6.

Length is obtained from longitudinal images at the maximum longitudinal diameter, in both coronal (See figures 1a and 2a) and sagittal (1b and 2b) planes. **The length used in the equation is the mathematical average of these two values.**

The width and thickness are measured in two orthogonal directions on an axial transverse image at the level of the hila. The hilar level is determined via a combination of appearances on the axial dataset (i.e. location of renal vessels and collecting system) as well as reference to the longitudinal images. (Note the solid white reference line on the coronal and sagittal images helping to indicate the correct axial image to measure).

Width should represent a measure from medial hila to lateral interpolar region of the kidney, and the depth measure to be perpendicular to that measure (figure 1c and 2c). These measurements should include all renal parenchyma and cysts, including exophytic cysts.
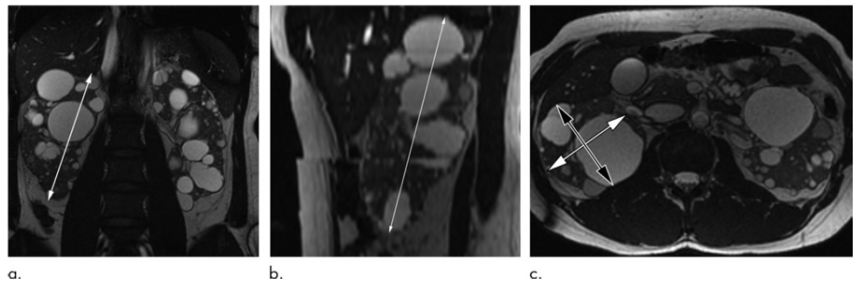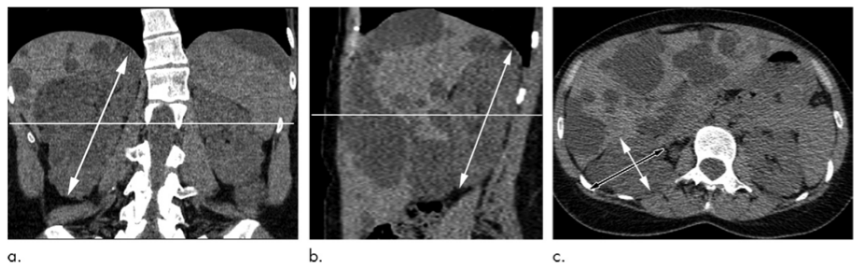
Figure 1: Example measurements on MRI

a.   b.   c.

Figure 2: Example measurements on CT

a.   b.   c.

Step 2: Reporting of imaging in ADPKD

These are the components of a report that are helpful for clinicians who manage patients with ADPKD:

1. **Confirm that diagnostic criteria for ADPKD are present based on unified diagnostic criteria (table 1).**
   - These criteria are for patients with a known family history; you may not have that information, but you can report that in the proper clinical setting the findings would be consistent with a diagnosis of ADPKD.
   - The key morphologic points are bilateral cystic involvement, number of cysts on each kidney (do not need to count if innumerable), and if present, renal enlargement.
   - These criteria are to rule in ADPKD. Exclusion criteria are also age-based but the clinician can make that determination; if there are fewer than 5 cysts on each side please report the exact number.
   - If you do not have enough information to definitively make the diagnosis, you can report the number of cysts and suggest that this may fit in the right clinical context.
   - If the diagnosis is uncertain, reporting the presence of hepatic cysts (if present) is helpful.

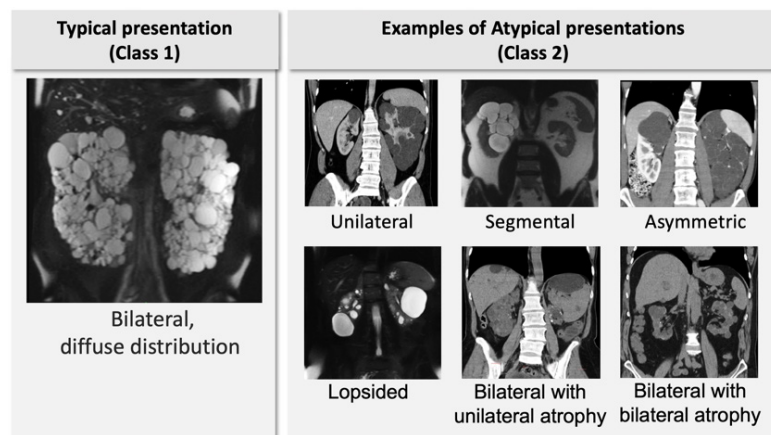   **Table 1: Criteria for Diagnosis of ADPKD in the Setting of Known Family History**

Age (years)	Number of cysts	Sensitivity
15-29	Total of ≥ 3 cysts (unilateral or bilateral)	81.7%
30-39	Total of ≥ 3 cysts (unilateral or bilateral)	95.5%
40-59	≥ 2 cysts in each kidney	90%

   Adapted from: Pei Y et al. *J Am Soc Nephrol.* 2009; 20(1):205-12

2. **Classify as 'typical' or 'atypical' morphology as described in the Mayo Imaging Classification (Figure 3)**
   - If atypical, it is helpful to provide the distribution (eg, 'unilateral', 'lopsided', etc)

   Figure 3: Mayo Imaging Classification Typical and Atypical presentations of ADPKD

   Adapted from Irazabal MV, et al. *J Am Soc Nephrol.* 2015; 26(1):160-72

3. **Report volumes of each kidney as determined by the ellipsoid equation described above.**
   - Please report the volumes of each kidney separately, as well as the method used to calculate this (e.g.; right kidney 760 ml as calculated by ellipsoid equation)
   - You do not necessarily need to provide measurements/volumes of individual cysts. This can however be helpful for example in lopsided distributions or where a single cyst or several cysts can account for a large portion of the overall volume.
   - If applicable, comparison to previous kidney volumes (best expressed as absolute change in volume).

4. **If seen, presence or absence of hepatic cysts.**
5. **Any other relevant findings as per any other assessment.**