American College of Radiology
ACR Appropriateness Criteria®

Autosomal Dominant Polycystic Kidney Disease

Variant: 1 Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US kidneys retroperitoneal	Usually Appropriate	O
MRI abdomen without IV contrast	Usually Appropriate	O
MRI abdomen without and with IV contrast	May Be Appropriate (Disagreement)	O
CT abdomen with IV contrast	May Be Appropriate	☢☢☢
CT abdomen without IV contrast	May Be Appropriate	☢☢☢
CT abdomen and pelvis with IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
WBC scan abdomen and pelvis	Usually Not Appropriate	☢☢☢☢

Variant: 2 Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.

Procedure	Appropriateness Category	Relative Radiation Level
MRI abdomen without IV contrast	Usually Appropriate	O
MRI abdomen without and with IV contrast	May Be Appropriate (Disagreement)	O
CT abdomen without IV contrast	May Be Appropriate	☢☢☢
US kidneys retroperitoneal	Usually Not Appropriate	O
CT abdomen and pelvis with IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen with IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
WBC scan abdomen and pelvis	Usually Not Appropriate	☢☢☢☢

Variant: 3 Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI abdomen without and with IV contrast	Usually Appropriate	O
CT abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢
MRI abdomen without IV contrast	May Be Appropriate (Disagreement)	O
CT abdomen and pelvis without IV contrast	May Be Appropriate (Disagreement)	☢☢☢
CT abdomen with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
CT abdomen without IV contrast	May Be Appropriate	☢☢☢
US kidneys retroperitoneal	Usually Not Appropriate	O
WBC scan with SPECT or SPECT/CT abdomen and pelvis	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢

CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
Gallium scan whole body	Usually Not Appropriate	☢☢☢☢

<u>Variant: 4</u> Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI abdomen without and with IV contrast	Usually Appropriate	O
MRI abdomen without IV contrast	May Be Appropriate	O
CT abdomen and pelvis with IV contrast	May Be Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	May Be Appropriate (Disagreement)	☢☢☢
US kidneys retroperitoneal	Usually Not Appropriate	O
CT abdomen with IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen without IV contrast	Usually Not Appropriate	☢☢☢
WBC scan with SPECT or SPECT/CT abdomen and pelvis	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
Gallium scan whole body	Usually Not Appropriate	☢☢☢☢

Panel Members

Melanie P. Caserta, MD[a], Andrei S. Purysko, MD[b], Tara M. Catanzano, BCh, MB[c], Silvia D. Chang, MD[d], Alberto Diaz De Leon, MD[e], David S. Goldfarb, MD[f], Mary S. Hedges, MD[g], Susie Q. Lew, MD[h], Refky Nicola, DO, MSc[i], Venkateswar R. Surabhi, MD[j], Myles T. Taffel, MD[k], Gaurav Khatri, MD[l]

Summary of Literature Review

Introduction/Background

Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary kidney disease and is the fourth leading cause of end-stage kidney disease (ESKD) in adults [1, 2]. Disease severity is determined by the type of genetic mutation, most commonly mutations in *PKD*1 (80%) and *PKD*2 (15%), with *PKD*2 mutations associated with milder cystic disease compared with *PKD*1 mutations [3]. The course of disease is characterized by progressive kidney cyst development, increased kidney volume, and progressive loss of kidney function over decades [1, 3]. Approximately 50% of ADPKD patients will progress to ESKD by 60 years of age [2]. Extrarenal manifestations include hepatic and pancreatic cysts, intracranial aneurysms, cardiac valvular lesions, and abdominal wall hernias [1].

Genetic testing is not the first-line tool in diagnosing ADPKD. A combination of family history, imaging manifestations, and relevant clinical findings is used to establish the diagnosis of ADPKD, with imaging playing a key role. Patients may present for ADPKD screening based on known family history or discovery of cysts at imaging, or due to nonspecific signs or symptoms, including but not limited to hypertension, kidney insufficiency, and abdominal pain [3]. Molecular testing may be

beneficial in younger (<40 years of age) at-risk patients with equivocal findings on imaging [3]. Imaging is also used to establish the diagnosis of kidney complications in patients with ADPKD, primarily cyst infection and hemorrhage into cysts. Kidney stones occur at higher rates in people with ADPKD, but are discussed separately as described below.

Total kidney volume (TKV) increases throughout the course of ADPKD. TKV adjusted for height and age has been established as the best predictor for kidney function decline [2]. The Consortium for Radiologic Imaging Studies of Polycystic Kidney Disease (CRISP) demonstrated the ability to predict future decline in glomerular filtration rate (GFR) based on initial height-adjusted TKV (htTKV) with a high htTKV predicting a more rapid decline in GFR [1, 4]. TKV can be used as an imaging biomarker to track disease progression and evaluate the effectiveness of treatment. Manual calculation using the ellipsoid formula or manual contouring (planimetry) methods can be used to determine TKV; however, recent development of automated algorithms has increased efficiency and accuracy in determining total kidney and cyst volume measurements. Nomograms using TKV have been developed to predict the risk for future kidney failure and to classify patients into five categories according to the Mayo Clinic Imaging Classification [3]. In 2016, the FDA qualified TKV as a prognostic biomarker and further delegated it as a likely surrogate end point for clinical trials in 2018 [2]. Once the diagnosis of ADPKD has been established, serial imaging to follow TKV can be used to monitor disease progression.

Advances in the treatment of ADPKD have focused on reducing the growth of kidney cysts and slowing the decline of kidney function. Suppression of vasopressin decreases cystogenesis and preserves kidney function. Tolvaptan, a vasopressin V2 receptor antagonist, has been shown to modify ADPKD by slowing the rate of TKV and slowing GFR decline [3]. Development of other disease-modifying therapeutic drugs is in progress and may provide further breakthroughs in the treatment of ADPKD.

Please note that this document on ADPKD and the variants it covers assumes that there are no clinical signs or suspicion of pyelonephritis, kidney stones, or kidney mass as these are each covered in a different topic (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5], "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6], and "Indeterminate Renal Mass" [7]) for further guidance.

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously wherein each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.

The goal of imaging is to diagnose suspected ADPKD in patients with known family history and to exclude mimics of ADPKD. This includes evaluation of the kidneys and other solid organs to make the diagnosis based on age-dependent criteria and the number of visible kidney cysts. With the information from imaging, this may initiate the appropriate treatment plan sooner and can improve patient outcomes by reducing the severity of illness. This will ideally help to reduce delaying the appropriate treatment in patients at higher risk for progression to ESKD.

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
A. CT abdomen and pelvis with IV contrast

There is limited literature on the use of CT abdomen and pelvis with IV contrast for initial imaging in patients with clinical suspicion for ADPKD. Validated diagnostic criteria for workup of patients at risk for ADPKD have not been established with CT. However, patients with enlarged cystic kidneys in the setting of positive family history can be diagnosed with CT [13]. CT with IV contrast is preferable due to better visualization of kidney cysts. Abdominal imaging with CT can also detect extrarenal manifestations of ADPKD including liver cysts, pancreas cysts, enlarged seminal vesicles, and splenomegaly [2]. The inclusion of pelvic CT is not usually necessary for the diagnosis of ADPKD. The KDIGO 2025 guidelines indicate that kidney CT with or without IV contrast is an option for patients with equivocal or atypical features on US or in patients with an uncertain diagnosis of ADPKD after the detection of kidney and/or liver cysts on US [11].

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
B. CT abdomen and pelvis without and with IV contrast

There is limited literature on the use of CT abdomen and pelvis without and with IV contrast for initial imaging in patients with clinical suspicion for ADPKD. There is no benefit in performing CT without and with IV contrast for the initial diagnosis of ADPKD. The inclusion of pelvic CT is not usually necessary for the diagnosis of ADPKD.

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
C. CT abdomen and pelvis without IV contrast

There is limited literature on the use of CT abdomen and pelvis without IV contrast for initial imaging in patients with clinical suspicion for ADPKD. Validated diagnostic criteria for the workup of patients at risk for ADPKD have not been established with CT. However, patients with enlarged cystic kidneys in the setting of a positive family history can be diagnosed with CT [13]. Abdominal imaging with CT can also detect extrarenal manifestations of ADPKD, including liver cysts, pancreas cysts, enlarged seminal vesicles, and splenomegaly [2]. The inclusion of pelvic CT is not usually necessary for the diagnosis of ADPKD. The KDIGO 2025 guidelines indicate that kidney CT with or without IV contrast is an option for patients with equivocal or atypical features on US or in patients with an uncertain diagnosis of ADPKD after the detection of kidney and/or liver cysts on US [11].

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
D. CT abdomen with IV contrast

There is limited literature on the use of CT abdomen with IV contrast for initial imaging in patients with clinical suspicion for ADPKD. It is important to ensure coverage of the entirety of the kidneys on the abdomen CT, as the kidneys can be markedly enlarged in patients with ADPKD. CT with IV contrast is preferable due to better visualization of kidney cysts. Validated diagnostic criteria for workup of patients at risk for ADPKD have not been established with CT. However, patients with enlarged cystic kidneys in the setting of positive family history can be diagnosed with CT [13]. Abdominal imaging with CT can also detect extrarenal manifestations of ADPKD including liver cysts, pancreas cysts, and splenomegaly [2]. The KDIGO 2025 guidelines indicate that kidney CT with or without IV contrast is an option for patients with equivocal or atypical features on US or in patients with an uncertain diagnosis of ADPKD after the detection of kidney and/or liver cysts on US [11].

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
E. CT abdomen without and with IV contrast

There is limited literature on the use of CT abdomen without and with IV contrast for initial imaging in patients with clinical suspicion for ADPKD. There is no benefit in performing CT without and with IV contrast for the initial diagnosis of ADPKD.

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
F. CT abdomen without IV contrast

There is limited literature on the use of CT abdomen without IV contrast for initial imaging in patients with clinical suspicion for ADPKD. It is important to ensure coverage of the entirety of the kidneys on the abdomen CT, as the kidneys can be markedly enlarged in patients with ADPKD. Validated diagnostic criteria for workup of patients at risk for ADPKD have not been established with CT. However, patients with enlarged cystic kidneys in the setting of positive family history can be diagnosed with CT [13]. Abdominal imaging with CT can also detect extrarenal manifestations of ADPKD including liver cysts, pancreas cysts, and splenomegaly [2]. The KDIGO 2025 guidelines indicate that kidney CT with or without IV contrast is an option for patients with equivocal or atypical features on US or in patients with an uncertain diagnosis of ADPKD after the detection of kidney and/or liver cysts on US [11].

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
G. MRI abdomen without and with IV contrast

MRI is highly sensitive in detecting small cysts, and is beneficial for detecting less severe forms of ADPKD, and for those patients for whom exclusion of ADPKD is important [3]. MRI is an alternative to US in diagnosing ADPKD in at-risk patients, particularly for patients <30 years of age in whom US has reduced diagnostic sensitivity. GBCM is not required for the diagnosis of ADPKD [12]. There is limited literature documenting the additional benefit of GBCM compared to MRI abdomen without IV contrast for the diagnosis of ADPKD. The KDIGO 2025 guidelines recommend MRI for equivocal or atypical features on US or in patients with an uncertain diagnosis of ADPKD after the detection of kidney and/or liver cysts on US or CT [11]. The KDIGO 2025 guidelines do not specify the use of GBCM. Due to the increased sensitivity of MRI in detecting small cysts, US-based diagnostic criteria cannot be extrapolated to MRI [13]. TRISP compared the diagnostic performance of high-resolution US with T2-weighted axial MR images in subjects at risk of ADPKD <40 years of age [12]. Using a total of >10 cysts in the kidneys provided a clear separation

between affected and unaffected patients on MRI, with a sensitivity and specificity of 100% for diagnosing ADPKD. A total of <10 cysts in both kidneys on MRI can exclude ADPKD with a NPV of 100%. More stringent criteria for excluding ADPKD in the setting of living kidney donor evaluation are recommended using a total of <5 kidney cysts [12]. When MRI findings are equivocal (5-19 cysts), genetic testing may be necessary.

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
H. MRI abdomen without IV contrast

For the purpose of diagnosing ADPKD, MRI abdomen without intravenous (IV) contrast is highly sensitive in detecting small cysts and is beneficial for detecting less severe forms of ADPKD and for patients for whom exclusion of ADPKD is important [3]. MRI is an alternative to US in diagnosing ADPKD in at-risk patients, particularly for patients <30 years of age in whom US has reduced diagnostic sensitivity and gadolinium-based contrast media (GBCM) is not required for the diagnosis of ADPKD [12]. There is limited literature documenting the additional benefit of GBCM compared to MRI abdomen with IV contrast for the diagnosis of ADPKD. The KDIGO 2025 guidelines recommend MRI for equivocal or atypical features on US or in patients with an uncertain diagnosis of ADPKD after the detection of kidney and/or liver cysts on US or CT [11]. The KDIGO 2025 guidelines do not specify the use of GBCM. Due to the increased sensitivity of MRI in detecting small cysts, US-based diagnostic criteria cannot be extrapolated to MRI [13]. The TRISP compared the diagnostic performance of high-resolution US with T2-weighted axial MR images in subjects at risk of ADPKD <40 years of age [12]. Using a total of >10 cysts in the kidneys provided a clear separation between affected and unaffected patients on MRI, with a sensitivity and specificity of 100% for diagnosing ADPKD. A total of <10 cysts in both kidneys on MRI can exclude ADPKD with a NPV of 100%. More stringent criteria for excluding ADPKD in the setting of living kidney donor evaluation are recommended using a total of <5 kidney cysts [12]. When MRI findings are equivocal (5-19 cysts), genetic testing may be necessary.

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.
I. US kidneys retroperitoneal

Ultrasound (US) is the first-line imaging in the diagnosis of ADPKD in part due to its high diagnostic accuracy [3, 8]. The Kidney Disease: Improving Global Outcomes (KDIGO) 2025 guidelines, the Kidney Health Australia, and the Spanish Working Group on Inherited Kidney Disease all recommend US as the first-line screening of adults at risk for ADPKD [9-11]. Modern US equipment can detect kidney cysts as small as 2 to 3 mm [12]. Unified diagnostic criteria incorporating both *PKD*1 and *PKD*2 types have been developed using US to diagnose ADPKD in at-risk patients based on the number of cysts and age [8]. Screening at-risk patients for ADPKD with US can be considered beginning at ≥16 years of age. Age-dependent US diagnostic criteria have been established, with a higher number of cysts required to make the diagnosis in older patients, as kidney cysts occur in the general population with increasing age. The diagnostic performance of these criteria is excellent for patients >40 years of age, but suboptimal for patients <30 years of age [12]. The Toronto Radiological Imaging Study of Polycystic Kidney Disease (TRISP) study found that US has 97% sensitivity in diagnosing ADPKD when using the criteria of three or more total cysts in at-risk subjects <30 years of age, but recommends using more stringent criteria of identifying two or more cysts in each kidney (positive predictive value of 100%) in this age group to minimize false-positive results [12]. The absence of cysts on US was sufficient to exclude disease in patients >30 years of age with a negative predictive value (NPV) of 100%, but not in

younger patients [12]. This study found high concordance of kidney cyst counts between US and MRI; however, the diagnostic performance of US is limited in part by patient body habitus.

Variant 1:Adult. Suspected autosomal dominant polycystic kidney disease. Positive family history. Initial imaging.

J. WBC scan abdomen and pelvis

There is no relevant literature to support the use of white blood cell (WBC) scan abdomen and pelvis for initial imaging of suspected ADPKD.

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.

The goal of imaging is to differentiate typical and atypical ADPKD and monitor TKV as a marker of disease severity. Identifying patients with rapidly growing kidneys can direct these patients towards early therapeutic intervention. The Mayo Imaging Classification can be used to stratify patients into five imaging classes that predict their risk for future kidney function decline based on height- and age-adjusted TKV [11, 14]. Currently, there are no clear guidelines for the role of surveillance imaging as to when and how often to image patients with ADPKD. In patients classified as slow progressors according to MIC, surveillance imaging is usually not indicated. In other patients, estimated GFR (eGFR) may not adequately assess disease progression, especially in early stages, and monitoring of TKV by imaging may help determine disease progression and evaluate effects of therapeutic interventions [15].

Surveillance imaging can also monitor extrarenal manifestations of ADPKD. Information obtained from imaging surveillance can enable the identification of patients who may benefit from the appropriate treatment plan sooner and can improve patient outcomes by slowing the progression of kidney disease. This will ideally help reduce the delay in providing the appropriate treatment to patients at higher risk for progression to ESKD. Early recognition of patients with rapid enlargement can direct them to treatment aimed at reducing cyst growth. A barrier to the adoption of TKV measurements is the time involved to perform volumetric evaluation using commercially available software to manually trace the kidney outline (planimetry). Automatic segmentation techniques based on deep learning for TKV computation provide fast and reproducible measurements of TKV, comparable to those obtained through manual segmentation by clinical experts [16].

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.

A. CT abdomen and pelvis with IV contrast

Monitoring of kidney volume is useful in assessing the need for and efficacy of therapies for ADPKD. CT is an accurate diagnostic tool for assessing TKV in patients with ADPKD and has similar diagnostic value as MRI in TKV [28] [20]. The literature has shown that both CECT and NCCT can be used to evaluate TKV; however, there is limited literature comparing CECT with NCCT for ADPKD surveillance [16, 29]. IV contrast and pelvic CT are not necessary for the purposes of monitoring TKV. Several studies have shown that CT can be used to evaluate kidney volume quantitatively and can be a useful modality for monitoring ADPKD progression or response to therapy [20, 30-32]. An early study that performed serial volumetric monitoring of patients with ADPKD found that CECT can be used to quantify the volume components of progression in ADPKD, and that the volume fraction of kidneys comprised of cysts may be a useful indicator of ADPKD progression. This study found that patients could be separated into two groups, those with rapid volume enlargement and

those with a barely perceptible rate of change, with fluid accumulation in cysts being the primary contributor to the increase in kidney volume [30].

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
B. CT abdomen and pelvis without and with IV contrast

There is no relevant literature to support the use of CT abdomen and pelvis without and with IV contrast for the purposes of imaging surveillance of ADPKD.

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
C. CT abdomen and pelvis without IV contrast

Monitoring of kidney volume is useful in assessing the need for and efficacy of therapies for ADPKD. CT is an accurate diagnostic tool for assessing TKV in patients with ADPKD and has similar diagnostic value as MRI in TKV [20, 29]. For the purposes of ADPKD surveillance, both CECT and NCCT can be performed to evaluate TKV, but there is limited literature comparing CECT with NCCT for ADPKD surveillance [16, 29]. IV contrast and pelvic CT are not necessary for the purposes of monitoring TKV. Several studies have shown that CT can be used to quantitatively evaluate kidney volume and can be a useful modality for monitoring ADPKD progression or response to therapy [20, 30-32]. A more recent investigation evaluating low-dose NCCT found that linear kidney measurements used to calculate estimated TKV are an accurate and reproducible way of obtaining TKV measurements.

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
D. CT abdomen with IV contrast

Monitoring of kidney volume is useful in assessing the need for and efficacy of therapies for ADPKD. CT is an accurate diagnostic tool for assessing TKV in patients with ADPKD and has similar diagnostic value as MRI in TKV [20, 28]. There is a lack of literature comparing CT abdomen with CT abdomen and pelvis. Although pelvic CT is not necessary for determining TKV measurements, coverage of the abdomen must image the entirety of the kidneys, which may be markedly enlarged in patients with ADPKD. There is a lack of literature comparing CECT with NCCT for ADPKD surveillance [16, 29]. IV contrast does not provide any additional benefit for the purposes of monitoring TKV. Several studies have shown that CT can be used to evaluate kidney volume quantitatively and can be a useful modality for monitoring ADPKD progression or response to therapy [20, 30-32].

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
E. CT abdomen without and with IV contrast

There is no relevant literature to support the use of CT abdomen without and with IV contrast for the purposes of imaging surveillance of ADPKD.

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
F. CT abdomen without IV contrast

Monitoring of kidney volume is useful in assessing the need for and efficacy of therapies for ADPKD. CT is an accurate diagnostic tool for assessing TKV in patients with ADPKD and has similar diagnostic value as MRI in TKV [20, 28]. There is insufficient literature comparing CT abdomen with

CT abdomen and pelvis. Scan coverage of the abdomen must image the entirety of the kidneys, which may be markedly enlarged in patients with ADPKD, to obtain accurate TKV measurements. For the purposes of ADPKD surveillance, both contrast-enhanced CT (CECT) and noncontrast CT (NCCT) can be performed to evaluate TKV. There is a lack of literature comparing CECT with NCCT for ADPKD surveillance [16, 29]. IV contrast is not necessary for the purposes of monitoring TKV. Several studies have shown that CT can be used to quantitatively evaluate kidney volume and can be a useful modality for monitoring ADPKD progression or response to therapy [20, 30-32]. A more recent investigation evaluating low-dose NCCT found that linear kidney measurements used to calculate estimated TKV were an accurate and reproducible way of obtaining TKV measurements [20]. [16].

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
G. MRI abdomen without and with IV contrast

Monitoring of kidney volume is useful in assessing the need for and efficacy of therapies for ADPKD, and measurement should be accurate and reproducible to within a few percentage points of TKV [17]. The CRISP observational cohort study of ADPKD established MRI as an accurate and reproducible method for calculating TKV [4] [18]. Few studies are comparing MRI to CT, but they have similar diagnostic accuracy when calculating TKV [19, 20]. High soft tissue contrast resolution of MRI is an advantage compared to CT, and standard T2WI on MRI can easily identify cysts without the need for contrast material [21]. Literature is limited regarding the additional benefit of IV contrast for TKV surveillance in ADPKD, but GBCM is not necessary for the purposes of measuring TKV. Semiautomated and automated segmentation techniques using MRI are rapid, comparable to the reference standard of manual segmentation, and can replace manual segmentation [17, 22-27].

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
H. MRI abdomen without IV contrast

Monitoring of kidney volume is useful in assessing the need for and efficacy of therapies for ADPKD, and measurement should be accurate and reproducible to within a few percentage points of TKV [17]. The CRISP observational cohort study of ADPKD established MRI as an accurate and reproducible method for calculating TKV [4] [18]. Few studies are comparing MRI to CT, but they have similar diagnostic accuracy when calculating TKV [19, 20]. High soft tissue contrast resolution of MRI is an advantage compared to CT, and standard T2-weighted imaging (T2WI) on MRI can easily identify cysts without the need for contrast material [21]. Literature is limited regarding the additional benefit of IV contrast for TKV surveillance in ADPKD, but GBCM is not necessary for the purposes of measuring TKV. Semiautomated and automated segmentation techniques using MRI are rapid, are comparable to the reference standard segmentation by clinical experts, and can replace manual segmentation [17, 22-27].

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
I. US kidneys retroperitoneal

Monitoring of kidney volume is useful in assessing the efficacy of therapies for ADPKD. US can be performed to calculate kidney volumes, but 2-D methods of manually calculating US are tedious and lack the precision necessary to measure short-term disease progression compared to CT and MRI segmentation [33]. The CRISP cohort found variability of kidney measurements between

sonographers ranged from 18% to 42% with poor accuracy and reproducibility in sonographically calculating kidney volume using the ellipsoid method [33]. A study comparing US kidney volume measurements in early ADPKD with CT kidney volume calculations found that US volume was strongly correlated with CT volume by using the maximum transverse width (r = 0.83) [31]. US underestimated the volume when the kidney volume exceeded 800 mL [31]. Another study found a strong correlation (r = 0.91) between the logarithm of TKV determined by CT and total mediolateral distance determined by US [28]. US can be used to stratify patient risk based on TKV broadly, but it is less precise compared to MRI [33]. More recent efforts have been directed toward automated segmentation using 3-D US, with promising results. The first study to apply deep learning to 3-D US in ADPKD demonstrated that the test dataset was close to both human tracing and MRI autosegmentation [34]. In general, MRI or CT is preferred over US for surveillance imaging of known ADPKD.

Variant 2:Adult. Known autosomal dominant polycystic kidney disease. Surveillance of total kidney volume.
J. WBC scan abdomen and pelvis

There is no relevant literature to support the use of WBC scan abdomen and pelvis for surveillance imaging of known ADPKD.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.

The goal of imaging is to diagnose suspected complications such as hemorrhage into a cyst, cyst rupture, or infection of a cyst. Both kidney and liver cysts can be symptomatic. Other complications, including nephrolithiasis, pyelonephritis, or suspected solid kidney mass, are each covered in different topics (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5] , "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6], and "Indeterminate Renal Mass" [7]) for further guidance. Information obtained from imaging may enable initiation of the appropriate treatment plan sooner and may improve patient outcome by reducing the length of illness.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
A. CT abdomen and pelvis with IV contrast

CT is generally the first-line imaging in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage [35, 41]. Complications of ADPKD, such as cyst hemorrhage, rupture, or infection, can demonstrate similar imaging features on CT, and the clinical picture often guides management. CT abdomen and pelvis with IV contrast is sensitive for detecting and localizing complicated cysts, but radiologic findings are not specific for infection. Hemorrhagic or ruptured cysts present clinically with acute pain that may be accompanied by hematuria and/or anemia, whereas infected cysts present with fever and back pain [10]. CT can also evaluate other causes of abdominal pain, including stones or pyelonephritis, which are discussed in separate topics (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5] , "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6]), as well as nonrenal etiologies of abdominal pain. Findings of cyst infection on CT with IV contrast include a thickened and enhancing wall with perilesional inflammation [35, 36]. The presence of air in cysts indicates emphysematous cyst infection and is highly specific for diagnosing cyst infection. Sensitivity of CT for detecting cyst infection compared with fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT is low, ranging from 7% to 25% [35, 36]. One study reported that

hemorrhagic kidney cysts showed intracystic attenuation >25 Hounsfield units (HU), and only one hemorrhagic cyst showed wall thickening. However, attenuation <25 HU is not a reliable threshold for distinguishing hemorrhagic from infected cysts, as infected cysts may also exhibit increased density on CT [42]. Cysts with hyperdense fluid (>50 HU) are often hemorrhagic, but on contrast-enhanced examination, this may be difficult to distinguish from solid lesions. The presence of free fluid adjacent to a cyst with crenulated margins can suggest rupture. Ruptured cysts may also have associated acute hemorrhage.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
B. CT abdomen and pelvis without and with IV contrast

For the purposes of detecting suspected complications in patients with known ADPKD, there is no relevant literature to support the use of CT abdomen and pelvis without and with IV contrast for initial imaging.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
C. CT abdomen and pelvis without IV contrast

CT is generally the first-line in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage [35, 41]. CT without IV contrast has limitations in diagnosing infected cysts and cystic neoplasms, but can still identify complicated cysts in ADPKD. Complications of ADPKD, such as cyst hemorrhage, rupture, or infection, can demonstrate similar imaging features on CT, and the clinical picture often guides management. CT is sensitive for detecting and localizing complicated cysts, but radiologic findings are not specific for infection. Cyst wall enhancement as a sign of infection cannot be depicted on CT without IV contrast. Although perilesional stranding may still be visible on CT without IV contrast, it may be seen with either infected or hemorrhagic cysts. Hemorrhagic or ruptured cysts present clinically with acute pain that may be accompanied by hematuria and/or anemia, whereas infected cysts present with fever and back pain [10]. CT can also evaluate other causes of abdominal pain, including stones or pyelonephritis, which are discussed in separate topics (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5] , "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6]), as well as nonrenal etiologies of abdominal pain. The presence of air in cysts indicates emphysematous cyst infection and is highly specific for diagnosing cyst infection. Sensitivity of CT for detecting cyst infection compared with FDG-PET/CT is low, ranging from 7% to 25% [35, 36]. One study reported hemorrhagic kidney cysts showed intracystic attenuation >25 HU and only one hemorrhagic cyst showed wall thickening; however, attenuation of >25 HU is not a reliable threshold for distinguishing hemorrhagic from infected cysts, as infected cysts may also have increased density on CT [42]. Cysts with hemorrhage demonstrate hyperdense fluid (>50 HU) on noncontrast examination. High attenuation within complicated cysts is more readily apparent on noncontrast images compared with IV contrast-enhanced images [42]. The presence of free fluid adjacent to a cyst with crenulated margins can suggest rupture. Ruptured cysts may also have associated acute hemorrhage.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
D. CT abdomen with IV contrast

CT is generally the first-line imaging in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage [35, 41]. There is insufficient

literature comparing CT abdomen with CT abdomen and pelvis for suspected complications in patients with known ADPKD, but CT of the abdomen may have insufficient scan coverage of the kidneys in patients with ADPKD and will miss acute pathology in the pelvis.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
E. CT abdomen without and with IV contrast

For the purposes of detecting suspected complications in patients with known ADPKD, there is no relevant literature to support the use of CT abdomen without and with IV contrast for initial imaging.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
F. CT abdomen without IV contrast

CT is generally the first-line imaging in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage [35, 41]. CT without IV contrast has limitations in diagnosing infected cysts and cystic neoplasms, but can still identify complicated cysts in ADPKD. There is insufficient literature comparing CT abdomen with CT abdomen and pelvis for suspected complications in patients with known ADPKD, but CT of the abdomen may have insufficient scan coverage of the kidneys in patients with ADPKD and will miss acute pathology in the pelvis.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
G. FDG-PET/CT skull base to mid-thigh

FDG-PET/CT is more sensitive in detecting kidney cyst infections compared with MRI and CT, with a study showing sensitivities of 95% for PET/CT, 71.4% for MRI, and 25% for CT [35]. However, FDG-PET/CT is not first-line imaging for suspected complications. FDG-PET/CT is the test of choice when precise localization of an infected kidney or liver cyst is required for patients with ADPKD and suspected cyst infection [10, 43]. Studies have shown sensitivity of 73% to 88.9%, specificity of 70% to 100%, and NPV of 77% to 87.5% when diagnosing cyst infection with FDG-PET/CT [36, 44-46]. FDG-PET/CT is also able to diagnose alternative sources for infection [35, 36, 45]. FDG-PET/CT can differentiate infected from hemorrhagic cysts, as FDG is not taken up by hemorrhagic cysts [36, 41]. False-negative results can occur when there is a long delay between the beginning of antibiotic treatment and the performance of FDG-PET/CT, and it is recommended to perform FDG-PET/CT within 7 days of initiating antibiotics in ADPKD patients with suspected cyst infection [35, 36, 41].

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
H. Gallium scan whole body

There is limited evidence to support the use of whole body gallium scan for suspected complications of ADPKD.

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
I. MRI abdomen without and with IV contrast

MRI is sensitive for detecting and localizing complicated cysts and has higher sensitivity for detecting cyst infection compared to CT [35]. Literature is limited on the usefulness of GBCM for diagnosing complications in patients with ADPKD, but MRI abdomen without and with GBCM is

preferable for imaging patients with suspected complications. Findings of cyst infection on MRI without and with IV contrast include cyst wall thickening, wall enhancement, and infiltration of the adjacent fat [35, 36]. High signal intensity (SI) on diffusion-weighted imaging (DWI) (sensitivity 86.4%) and fluid-fluid levels are other findings of infected cysts on MRI [37]. Two studies found approximately 80% sensitivity and specificity for wall thickening and fluid-fluid level in diagnosing cyst infection on MRI [37, 38]. Intracystic gas is a specific finding for infection (specificity 100%), but its sensitivity is approximately 1% [37, 38]. Hemorrhagic cysts are common in ADPKD patients. Cyst bleeding or inflammation is more likely to occur as TKV increases [38]. Hemorrhagic cysts are readily diagnosed on MRI, appearing T1 hyperintense and T2 hypointense with no enhancement; however, they have overlapping features with cyst infection and can also show fluid-fluid levels and wall thickening . Hemorrhagic cysts have been shown to play a role in ADPKD progression and are an independent risk factor for predicting eGFR decline [2, 39, 40].

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
J. MRI abdomen without IV contrast

MRI with GBCM is preferred over MRI without IV contrast in many clinical scenarios, but MRI without IV contrast can be used to identify complicated kidney cysts in patients with ADPKD. T2WI can differentiate between simple and complex cysts without the need for gadolinium, but cannot reliably discriminate between bleeding, infection, and neoplasm [40]. There are currently no reliable noncontrast MRI techniques for classifying kidney cysts based on their composition. However, newer research using quantitative susceptibility mapping derived from multiecho gradient echo data shows potential for identifying properties of complex cysts and discriminating between hemorrhagic and proteinaceous cysts [40]. Hemorrhagic cysts are common in ADPKD patients, and cyst bleeding or inflammation are more likely to occur as TKV increases [38]. Hemorrhagic cysts have been shown to play a role in ADPKD progression and are an independent risk factor for predicting eGFR decline [2, 39, 40]. Findings of cyst infection on MRI without IV contrast include cyst wall thickening and infiltration of the adjacent fat [35, 36]. High SI on DWI (sensitivity 86.4%) and fluid-fluid levels are other MRI findings of infected cysts without IV contrast. High SI on DWI is not specific (specificity 33.3%) and can also be seen with intracystic hemorrhage [38]. Two studies found approximately 80% sensitivity and specificity for wall thickening and fluid-fluid level in diagnosing cyst infection on MRI [37, 38]. Intracystic gas is a specific finding for infection (specificity 100%), but its sensitivity is approximately 1% [37, 38].

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
K. US kidneys retroperitoneal

There is limited evidence to support the use of US for suspected complications in ADPKD. US can detect complicated cysts in ADPKD, but it does not differentiate between cysts with infection or hemorrhage [10]. US features of infected cysts overlap with hemorrhagic cysts, and include debris within the cyst, thickened walls, and increased blood flow surrounding the cyst [35]. In a study, US diagnosed cyst infection in only 2.6% of cases [41]. Heterogeneous echogenicity was observed in 85.1% of infected cysts in one study; however, this finding alone is nonspecific [42].

Variant 3:Adult. Known autosomal dominant polycystic kidney disease. Suspected complication. Normal kidney function. Initial imaging.
L. WBC scan with SPECT or SPECT/CT abdomen and pelvis

There is limited evidence to support the use of WBC scan with single-photon emission CT (SPECT)

or SPECT/CT abdomen and pelvis as the initial imaging study for suspected complications of ADPKD.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.

The goal of imaging is to diagnose suspected complications such as hemorrhage into a cyst, cyst rupture, or infection of a cyst. Both kidney and liver cysts can be symptomatic. Other complications, including nephrolithiasis, pyelonephritis, or suspected kidney mass, are each covered in a different topic (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5] , "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6], and "Indeterminate Renal Mass" [7]) for further guidance. Information obtained from imaging may enable initiation of the appropriate treatment plan sooner and may improve patient outcome by reducing the length of illness.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
A. CT abdomen and pelvis with IV contrast

According to the consensus statement on the use of IV iodinated contrast media in patients with kidney disease from the ACR and the National Kidney Foundation, the risk of acute kidney injury in patients with reduced kidney function receiving iodinated contrast media has been overstated [48]. Discussion between the radiologist and ordering clinician about the risks and benefits of contrast-enhanced imaging can be helpful for patients at high risk of contrast-induced acute kidney injury. In the setting of known ADPKD with reduced kidney function and suspected complications, the use of iodinated contrast will depend on the patient's eGFR. Although CT is generally the first-line imaging in ADPKD patients with suspected complications due to its rapid examination and high accuracy for detecting cyst hemorrhage [35, 41], MRI or CT without IV contrast is generally preferable in patients with reduced kidney function (eGFR <30 mL/min). If the benefit of performing CT with IV contrast for suspected complications in an acute setting outweighs the risk of contrast administration in patients with kidney disease who are not undergoing maintenance dialysis, then prophylaxis with IV saline is indicated for patients with eGFR <30mL/min and no other contraindication (eg, heart failure or other hypervolemic conditions) [48]. For individual high-risk circumstances, prophylaxis can be considered for eGFR between 30 to 44 mL/min [48].

Complications of ADPKD, such as cyst hemorrhage or infection, can demonstrate similar imaging features on CT, and the clinical picture often guides management. CT is sensitive for detecting and localizing complicated cysts, but radiologic findings are not specific for infection. Hemorrhagic or ruptured cysts present clinically with acute pain that may be accompanied by hematuria and/or anemia, whereas infected cysts present with fever and back pain [10]. CT can also evaluate other causes of abdominal pain, including stones or pyelonephritis, which are discussed in separate topics (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5] , "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6]), as well as nonrenal etiologies of abdominal pain. Findings of cyst infection on CT with IV contrast include a thickened and enhancing wall with perilesional inflammation [35, 36]. The presence of air in cysts indicates emphysematous cyst infection and is highly specific for diagnosing cyst infection. Sensitivity of CT for detecting cyst infection compared with FDG-PET/CT is low, ranging from 7% to 25% [35, 36]. Cysts with hyperdense fluid (>50 HU) are often hemorrhagic; however, on contrast-enhanced examination, this may be difficult to distinguish from solid lesions. The presence of free fluid adjacent to a cyst with crenulated margins can indicate a potential rupture. Ruptured cysts may

also have associated acute hemorrhage.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
B. CT abdomen and pelvis without and with IV contrast

For the purposes of detecting suspected complications in patients with known ADPKD, there is no literature to support CT abdomen and pelvis without and with IV contrast for initial imaging.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
C. CT abdomen and pelvis without IV contrast

CT is generally the first-line imaging in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage [35, 41]. CT without IV contrast has limitations in diagnosing infected cysts and cystic neoplasms, but can still identify complicated cysts in ADPKD and may be useful in an acute setting to evaluate suspected complications for patients with known ADPKD and reduced kidney function. Complications of ADPKD, such as cyst hemorrhage or infection, can demonstrate similar imaging features on CT, and the clinical picture often guides management. CT is sensitive for detecting and localizing complicated cysts, but radiologic findings are not specific for infection. Cyst wall enhancement as a sign of infection cannot be depicted on CT without IV contrast. Although perilesional stranding may still be visible on CT without IV contrast, it may be seen with either infected or hemorrhagic cysts. Hemorrhagic or ruptured cysts present clinically with acute pain that may be accompanied by hematuria and/or anemia, whereas infected cysts present with fever and back pain [10]. CT can also evaluate other causes of abdominal pain, including stones or pyelonephritis which are discussed in separate topics (see the ACR Appropriateness Criteria® topics on "Acute Pyelonephritis" [5] , "Acute Onset Flank Pain-Suspicion for Stone Disease (Urolithiasis)" [6]), as well as nonrenal etiologies of abdominal pain. The presence of air in cysts indicates emphysematous cyst infection and is highly specific for diagnosing cyst infection. Sensitivity of CT for detecting cyst infection compared with FDG-PET/CT is low, ranging from 7% to 25% [35, 36]. In a study, hemorrhagic kidney cysts showed intracystic attenuation >25 HU, and only one hemorrhagic cyst showed wall thickening; however, attenuation of >25 HU is not a reliable threshold for distinguishing hemorrhagic from infected cysts, as infected cysts may also have increased density on CT. Cysts with hemorrhage demonstrate hyperdense fluid (>50 HU) on noncontrast examination. High attenuation within complicated cysts is more readily apparent on noncontrast images compared with IV contrast-enhanced images [42]. The presence of free fluid adjacent to a cyst with crenulated margins of the cyst can suggest rupture. Ruptured cysts may also have associated acute hemorrhage.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
D. CT abdomen with IV contrast

Although CT is generally the first-line imaging in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage [35, 41], there is insufficient literature comparing CT abdomen with CT abdomen and pelvis for suspected complications in patients with known ADPKD. CT of the abdomen may have insufficient scan coverage of the kidneys in patients with ADPKD and will miss acute pathology in the pelvis. In the setting of reduced kidney function, CT with IV contrast is less preferable compared to other options. The use of iodinated contrast will depend on the patient's eGFR.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected

complications. Reduced kidney function. Initial imaging.
E. CT abdomen without and with IV contrast

For the purposes of detecting suspected complications in patients with known ADPKD, there is no literature to support CT abdomen without and with IV contrast for initial imaging.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
F. CT abdomen without IV contrast

CT is generally the first-line imaging in ADPKD patients with suspected complications due to rapid examination and high accuracy for detecting cyst hemorrhage . CT without IV contrast has limitations in diagnosing infected cysts and cystic neoplasms, but can still identify complicated cysts in ADPKD. There is insufficient literature comparing CT abdomen with CT abdomen and pelvis for suspected complications in patients with known ADPKD. CT of the abdomen may have insufficient scan coverage of the kidneys in patients with ADPKD and will miss acute pathology in the pelvis.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
G. FDG-PET/CT skull base to mid-thigh

The CT examination accompanying FDG-PET can be performed without iodinated contrast. FDG-PET/CT is more sensitive in detecting kidney cyst infections compared with MRI and CT, with one study showing sensitivities of 95% for PET/CT, 71.4% for MRI, and 25% for CT [35]. However, FDG-PET/CT is not first-line imaging for suspected complications. FDG-PET/CT is the test of choice when precise localization of an infected kidney or liver cyst is required for patients with ADPKD and suspected cyst infection [10, 43]. Studies have shown sensitivity of 73% to 88.9%, specificity of 70% to 100% and NPV of 77% to 87.5% when diagnosing cyst infection with FDG-PET/CT [36, 44-46]. FDG-PET/CT is also able to diagnose alternative sources for infection [35, 36, 45]. FDG-PET/CT can differentiate infected from hemorrhagic cysts, as FDG is not taken up by hemorrhagic cysts [36, 41]. False-negative results can occur when there is a long delay between the beginning of antibiotic treatment and the performance of FDG-PET/CT, and it is recommended to perform FDG-PET/CT within 7 days of initiating antibiotics in ADPKD patients with suspected cyst infection [35, 36, 41].

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
H. Gallium scan whole body

There is limited evidence to support the use of whole body gallium scan as the initial imaging study for suspected complications of ADPKD.

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
I. MRI abdomen without and with IV contrast

Recommendations about the use of GBCM have evolved over time. According to the consensus statement on the use of IV GBCM in patients with kidney disease from the ACR and the National Kidney Foundation, the risk of nephrogenic systemic fibrosis (NSF) with a standard dose of group II GBCM is very low even in patients with eGFR <30 mL/min or acute kidney injury. The risk for NSF from group III GBCM is thought to be low, but there is insufficient confirmatory evidence. The highest risk GBCM for NSF is group I agents. The consensus statement states that group II GBCM should not be withheld or delayed if harm would result from not performing an indicated

examination [47].

MRI is sensitive for detecting and localizing complicated cysts and has higher sensitivity for detecting cyst infection compared to CT [35]. Literature is limited, but MRI abdomen with GBCM is preferable for imaging patients with suspected complications. For patients with known ADPKD and reduced kidney function, MRI with GBCM is preferred over CT for the evaluation of suspected complications.

Findings of cyst infection on MRI without and with IV contrast include cyst wall thickening, wall enhancement, and infiltration of the adjacent fat [35, 36]. High SI on DWI (sensitivity 86.4%) and fluid-fluid levels are other findings of infected cysts on MRI [37]. Two studies found approximately 80% sensitivity and specificity for wall thickening and fluid-fluid level in diagnosing cyst infection on MRI [37, 38]. Intracystic gas is a specific finding for infection (specificity 100%), but its sensitivity is approximately 1% [37, 38]. Hemorrhagic cysts are common in ADPKD patients, and cyst bleeding or inflammation are more likely to occur as TKV increases . Hemorrhagic cysts are readily diagnosed on MRI, appearing T1 hyperintense and T2 hypointense with no enhancement; however, they have overlapping features with cyst infection and can also show fluid-fluid levels and wall thickening [38]. Hemorrhagic cysts have been shown to play a role in ADPKD progression and are an independent risk factor for predicting eGFR decline [2, 39, 40].

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.
J. MRI abdomen without IV contrast

Although MRI with GBCM is preferred, MRI without IV contrast can be used to identify complicated kidney cysts in patients with ADPKD. T2WI can differentiate between simple and complex cysts without the need for gadolinium, but cannot reliably discriminate between bleeding, infection, and neoplasm [40]. There are currently no reliable noncontrast MRI techniques for classifying kidney cysts based on their composition. However, newer research using quantitative susceptibility mapping derived from multiecho gradient echo data shows potential for identifying properties of complex cysts and discriminating between hemorrhagic and proteinaceous cysts [40].

Hemorrhagic cysts are common in ADPKD patients, and cyst bleeding or inflammation are more likely to occur as TKV increases. Hemorrhagic cysts have been shown to play a role in ADPKD progression and are an independent risk factor for predicting eGFR decline [2, 39, 40]. Findings of cyst infection on MRI without IV contrast include cyst wall thickening and infiltration of the adjacent fat [35, 36]. High SI on DWI (sensitivity 86.4%) and fluid-fluid levels are other findings of infected cysts on MRI without IV contrast [37]. High SI on DWI is not specific (specificity 33.3%) and can also be seen with intracystic hemorrhage [38]. Two studies found approximately 80% sensitivity and specificity for wall thickening and fluid-fluid level in diagnosing cyst infection on MRI [37, 38]. Intracystic gas is a specific finding for infection (specificity 100%), but its sensitivity is approximately 1% [37, 38]. Hemorrhagic cysts are common in ADPKD patients. Cyst bleeding or inflammation is more likely to occur as TKV increases. Hemorrhagic cysts are readily diagnosed on MRI, appearing T1 hyperintense and T2 hypointense. Hemorrhagic cysts have been shown to play a role in ADPKD progression and are an independent risk factor for predicting eGFR decline [2, 39, 40].

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.

K. US kidneys retroperitoneal

US can detect complicated cysts in ADPKD, but it does not differentiate between cysts with infection or hemorrhage [10]. There is limited evidence to support the use of US for suspected complications in ADPKD, but it may be an option for detecting complicated cysts in patients with ADPKD who cannot undergo CT or MRI examination. US features of infected cysts overlap with hemorrhagic cysts, but include debris within the cyst, thickened walls, and increased blood flow surrounding the cyst [35]. One study showed that US diagnosed cyst infection in only 2.6% of cases [41]. Heterogeneous echogenicity was observed in 85.1% of infected cysts; however, this finding alone is nonspecific [42].

Variant 4:Adult. Known autosomal dominant polycystic kidney disease. Suspected complications. Reduced kidney function. Initial imaging.

L. WBC scan with SPECT or SPECT/CT abdomen and pelvis

There is no relevant literature to support the use of WBC scan with SPECT or SPECT/CT abdomen and pelvis as the initial imaging study for suspected complications of ADPKD.

Summary of Highlights

This is a summary of the key recommendations from the variant tables. Refer to the complete narrative document for more information.

- Variant 1: In patients with suspected ADPKD, both US kidneys retroperitoneal and MRI abdomen without IV contrast are usually appropriate. US is the first-line imaging for the diagnosis of ADPKD. MRI abdomen without IV contrast is highly sensitive for detecting small cysts and is an alternative imaging modality to US for the diagnosis of ADPKD. These procedures should be viewed as alternative initial procedures. MRI can be considered for detecting less severe forms of ADPKD or for diagnosing ADPKD in patients <30 years of age in whom US has reduced diagnostic sensitivity.

- Variant 2: For surveillance imaging of TKV in patients with known ADPKD, MRI of the abdomen without IV contrast is usually appropriate. MRI is an accurate and reproducible method for calculating TKV and can easily identify cysts without the need for GBCM. Semiautomated and automated techniques using MRI rapidly calculate TKV.

- Variant 3: In the setting of known ADPKD with suspected complication, both MRI abdomen without and with IV contrast and CT abdomen and pelvis with IV contrast are usually appropriate. These procedures should be viewed as alternative initial procedures. CT is a first-line imaging modality due to its rapid examination and high accuracy for detecting complicated cysts. MRI of the abdomen without and with IV contrast is an alternative to CT as first-line imaging, and has higher sensitivity for detecting cyst infection compared to CT.

- Variant 4: In the setting of known ADPKD with suspected complication and reduced renal function, MRI of the abdomen without and with IV contrast is usually appropriate. The risk for NSF is very low with group II GBCM and group II agents should not be withheld or delayed if harm would result from not performing the examination. The use of iodinated contrast in patients with reduced renal function depends on eGFR, and CT abdomen and pelvis with IV contrast may be appropriate in certain clinical scenarios.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at https://acsearch.acr.org/list. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria.

Gender Equality and Inclusivity Clause

The ACR acknowledges the limitations in applying inclusive language when citing research studies that predates the use of the current understanding of language inclusive of diversity in sex, intersex, gender, and gender-diverse people. The data variables regarding sex and gender used in the cited literature will not be changed. However, this guideline will use the terminology and definitions as proposed by the National Institutes of Health.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as

compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document.

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."

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[a]Mayo Clinic, Jacksonville, Florida. [b]Panel Chair, Cleveland Clinic, Cleveland, Ohio. [c]Baystate Health, Springfield, Massachusetts. [d]University of British Columbia, Vancouver, British Columbia, Canada. [e]The University of Texas MD Anderson Cancer Center, Houston, Texas. [f]New York University Langone Health and NYU Grossman School of Medicine, New York, New York; American Urological Association. [g]Mayo Clinic, Jacksonville, Florida, PCP - Internal medicine. [h]The George Washington University School of Medicine and Health Sciences, Washington, District of Columbia; American Society of Nephrology. [i]SUNY Upstate Medical University, Syracuse, New York. [j]The University of Texas MD Anderson Cancer Center, Houston, Texas. [k]New York University Langone Medical Center, New York, New York. [l]Specialty Chair, UT Southwestern Medical Center, Dallas, Texas.