

2025 KDIGO Executive Summary: Diagnosis and Management of Pediatric Autosomal Dominant Polycystic Kidney Disease.

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Autosomal dominant polycystic kidney disease (ADPKD) is the most common inherited kidney disorder, affecting 1 in 400 to 1,000 people worldwide [1]. Although ADPKD is largely an adult-onset condition, cyst formation and structural kidney changes can already be present during childhood or even before birth [2]. Recent data from Taiwan highlight this significant pediatric burden of ADPKD, with an incidence of 2.32 to 4.45 per 100,000 individuals and increased risks of hypertension and other systemic complications beginning early in life [3]. These significant care requirements reflect the importance of early diagnosis and prevention for affected patients and the need for evidence-based guidelines for treatment protocols and best practice measures.

The 2025 KDIGO guidelines build on earlier frameworks such as the 2019 international consensus statement on the diagnosis and management of ADPKD in children and young people to improve screening, monitoring, and treatment methodology for pediatric ADPKD patients [4 , 5]. Young and adolescent patients face unique challenges to care from detection of asymptomatic disease progression to early-onset (EO) hypertension, alongside stress faced by caregivers and families. This executive summary synthesizes the updated 2025 KDIGO recommendations and supporting evidence to outline new advancements and provide a practical approach for pediatric nephrologists to support improved treatment and transition into adulthood.

Diagnosis and screening of ADPKD in children requires input from families and coordination with physicians to ensure ethical, individualized patient decisions. The 2025 KDIGO guidelines recommend screening in children after discussion with parents and guardians regarding potential risks and harms stemming from testing, promoting shared decision-making with a comprehensive understanding of testing requirements and procedures [4]. Previous consensus statements emphasize the concept of shared decision-making, with recommendations that parents receive nondirective counseling to help parents consider all implications and prioritize the child's best interests [5]. Furthermore, Gimpel et al. [5] suggest involving teenagers and mature children in conversations to foster understanding and autonomy, as developmentally appropriate discussions and counseling about genetic transmission can also help families prepare emotionally for future treatment needs.

Renal ultrasound is recommended by KDIGO as the preferred imaging modality due to its safety and accessibility. MRI imaging has exceptionally accurate detection capabilities, but the sedation requirements and higher cost compared to ultrasound technology means it is not effective for broad screening applications [4]. A single cyst detected in a child below 15 years of age with a family history of ADPKD is recognized as a significant factor for diagnosis, but a negative ultrasound should not be utilized to exclude the diagnosis due to the age-dependent development of renal cysts [5].

For children with very early-onset (VEO) or atypical presentations, KDIGO recommends offering genetic testing; the expert consensus is to offer testing for cystic kidney disease genes in symptomatic infants and children [5]. The recommendations refine definitions of VEO and EO-ADPKD, supporting risk determination and identification of aggressive disease attributes.

In prenatal settings, enlarged echogenic kidneys with or without discrete cysts may raise suspicion for ADPKD, though differentiation from other cystic diseases such as autosomal recessive polycystic kidney disease can be challenging [4 , 5]. Genetic testing in KDIGO is recommended only in specific contexts, such as children with VEO or atypical presentation or those with cystic kidneys and no family history, allowing for more detailed stratification through clear phenotypic subcategories.

Although direct genetic testing using next-generation or Sanger sequencing is commercially available, its variable diagnostic yield, high cost, and implications for insurance and psychosocial stressors make serial renal ultrasound a more cost-effective and accessible first-line strategy in many pediatric cases [4 , 5]. A major addition in KDIGO 2025 is advising routine ultrasound screening of parents or grandparents when a child presents with cysts and no known family history, which builds upon previous nuclear family screening guidelines. KDIGO further stresses caution in diagnosing isolated cysts, suggesting monitoring and longitudinal assessment to avoid overmedicalization and unnecessary treatments. Ultrasound screening of grandparents when parents are under 40 years of age is also recommended to improve diagnostic certainty in children with renal cysts but no confirmed family history. This suggestion builds on prior family-based approaches and helps distinguish ADPKD from sporadic or benign findings in younger patients. In cases where an isolated renal cyst is identified in a child with no family history and negative imaging in both parents, KDIGO advises considering benign simple cysts in the differential to avoid premature labeling and potential for overmedicalization in uncertain cases [5].

Hypertension is the most common early complication in pediatric ADPKD, affecting up to 20 percent of children [6]. KDIGO emphasizes that blood pressure should be monitored annually from birth, and ambulatory blood pressure monitoring (ABPM) from age 5 is strongly recommended for improved detection of masked and nocturnal hypertension. This builds on the Network for Early Onset Cystic Kidney Disease (NEOCYST) consensus statement, which emphasized that even without a confirmed diagnosis, children with a family history of ADPKD should still undergo regular monitoring for hypertension and albuminuria [5].

KDIGO sets a more stringent blood pressure goal for pediatric patients, targeting ≤ 50 th percentile for age, sex, and height or $\leq 110/70$ mm Hg in adolescents with ADPKD. Although tighter blood pressure control is supported by adult ADPKD data, pediatric evidence is largely derived from small observational cohorts and extrapolated from general CKD populations, limiting the certainty of these recommendations [5]. The 2019 international consensus statement on pediatric ADPKD emphasized this limitation, with many blood pressure recommendations graded as moderate or weak in strength due to the scarcity of randomized pediatric trials and the reliance on expert consensus [5]. KDIGO acknowledges these limitations, and the recommendation reflects both current evidence and consensus expert opinion aiming to reduce long-term cardiovascular risk. These new guidelines are stricter than NEOCYST's recommendation of <75 th percentile or $<125/72$ mm Hg in older teens, highlighting KDIGO's more aggressive approach to mitigate cardiovascular and renal risk [5]. These thresholds are intended to be applied in the context of standardized blood pressure assessment, with average 24-h ABPM values preferred when available to guide clinical decision-making.

Antihypertensive therapy is indicated when BP exceeds the 95th percentile in adolescents, slightly higher than previous NEOCYST recommendations at the 90th percentile [5]. KDIGO further prioritizes renin-angiotensin system inhibitors (ACE inhibitors or ARBs) as first-line agents due to their dual efficacy in blood pressure and proteinuria control.

Proteinuria, though less frequent than in adults, is a critical early marker of disease progression [7]. Although KDIGO does not emphasize proteinuria as a hallmark of pediatric ADPKD, it recommends managing it by following general pediatric CKD guidelines, supporting early intervention when indicated. KDIGO clarifies that the evaluation and management of proteinuria in children with or at risk for ADPKD should follow the same approach used in other pediatric kidney diseases, without disease-specific modification [5 , 7].

Albumin-to-creatinine ratio monitoring may offer improved sensitivity over dipstick tests, enabling earlier detection and therapeutic response [5]. This proactive monitoring supports early initiation or escalation of ACE inhibitor or ARB therapy to slow progression.

Children with ADPKD and uncontrolled hypertension are at risk of developing early left ventricular hypertrophy (LVH), with pediatric ADPKD cohort studies reporting a higher incidence of hypertension, proteinuria, and LVH [5]. ABPM is more strongly correlated with LVH than clinic-based readings and can help identify masked or nocturnal hypertension, both of which are prevalent in this population. Echocardiographic screening and routine ABPM in high-risk patients with VEO- or EO-ADPKD can guide clinical management and help mitigate long-term cardiovascular risk [5]. In practice, the feasibility of ABPM from age five may be influenced by equipment availability, pediatric patient cooperation, and institutional resources. Broader adoption will require expanded access to validated pediatric ABPM devices and provider training, acknowledging that KDIGO's recommendation reflects an aspirational standard of care. These updates are summarized in Table 1 , which highlights the major changes in KDIGO 2025 compared to prior pediatric consensus guidelines.

2025 KDIGO recommendations for ADPKD: updates, rationale, and comparison to prior guidelines

ABPM, ambulatory blood pressure monitoring; VEO, very early-onset.

KDIGO recommends healthy lifestyle habits for children with or at risk for ADPKD, including physical activity, salt restriction, and weight control, reflecting broader CKD evidence that supports these measures to reduce blood pressure and proteinuria [9]. However, KDIGO offers limited discussion of psychosocial dimensions addressed in previous consensus statements, including proactive counseling on genetic guilt, open family communication, and emotional well-being throughout adolescence [5]. By combining medical management with psychosocial support, clinicians can help families build resilience and foster long-term engagement with disease management.

Long-term follow-up in pediatric ADPKD requires symptom-driven intervention and individualized treatment methodology to provide the best patient care and results. KDIGO recommends monitoring based on blood pressure, renal function, and new symptoms, reflecting a shift from routine interval-based imaging toward symptom-driven assessment [5].

Routine screening for extrarenal manifestations is not advised in asymptomatic children, with a recommended application of adult screening protocols when symptoms arise. This approach aligns with current adult ADPKD context, which discourages early screening for liver cysts or intracranial aneurysms unless risk factors or family history are present [10 , 11].

By emphasizing clinical context over routine surveillance, KDIGO minimizes unnecessary anxiety and avoids interventions unlikely to alter pediatric management. Furthermore, the updated guidelines recommend that urinary tract infections and nephrolithiasis should be managed according to general pediatric standards.

However, KDIGO additionally notes that atypical UTIs or persistent abdominal pain warrant diagnostic imaging to rule out cyst infection, expanding upon prior pediatric nephrology cases by offering ADPKD-specific context for treatment [12]. Uncomplicated urinary tract infections in

children with ADPKD should still be managed according to general pediatric standards, without need for ADPKD-specific interventions.

Nonsteroidal anti-inflammatory drugs are discouraged due to underlying kidney disease, a precaution supported by observational data showing increased cyst growth and nephrotoxicity risk with nonsteroidal anti-inflammatory drug exposure [13]. Vasopressin analogs are also not recommended for nocturnal enuresis in ADPKD children, countering older pediatric enuresis protocols that did not account for cystic disease mechanisms [5]. KDIGO recommends that nephrolithiasis in children with ADPKD be treated according to standard pediatric nephrology protocols, as there is no evidence for disease-specific differences in management.

KDIGO strongly emphasizes the importance of a structured transition to adult care for pediatric patients, with formal transition planning beginning in adolescence to improve self-management and family communication. KDIGO builds on existing frameworks in recommending open conversations between providers, patients, and their families regarding the transition process, reflecting increased awareness of transition-related morbidity and care gaps in chronic pediatric kidney diseases.

Failure to implement structured transition pathways has been associated with medication nonadherence and increased risk of disease progression during early adulthood [8]. In renal transplant cohorts, structured transition programs have been associated with fewer acute rejection episodes and improved adherence and patient satisfaction [8]. KDIGO emphasizes initiating a structured transition plan as children with or at risk for ADPKD move into young adulthood, evaluating kidney health and potential extrarenal complications. Incorporating education and shared decision-making can help prepare adolescents for independent disease management alongside maintaining family involvement. KDIGO further recommends nephrologists to empower parents and grandparents to openly discuss ADPKD with affected or at-risk children, highlighting the importance of communication in fostering resilience and self-management during the transition period. Future iterations of the guidelines may consider more specific implementation strategies drawing from structured models such as joint pediatric and adult transition clinics or multidisciplinary HCT programs to define staged competencies and integrated care teams to promote a successful handover from pediatric to adult nephrology services [14 , 15]. Figure 1 provides a schematic overview of the care pathway, integrating recommendations across diagnosis, monitoring, management, and structured transition to adult nephrology.

Flow diagram illustrating the clinical pathway for pediatric autosomal dominant polycystic kidney disease (ADPKD), outlining steps for diagnosis, monitoring, management, and structured transition to adult care.

The 2025 KDIGO pediatric ADPKD guidelines provide comprehensive, evidence-based recommendations addressing diagnosis and disease management to improve longitudinal care pathways. The guidelines introduce more precise definitions of disease subtypes and tighter blood pressure targets and provide expanded guidance on genetic testing and psychosocial support. Furthermore, the recommendations support evolving insights from pediatric nephrology research while revising practices such as routine imaging and indiscriminate pharmacologic use.

However, future iterations should look to incorporate structured approaches to assessing patient-reported outcomes and long-term quality of life, which are key metrics for evaluating the broader impact of early diagnosis. Additional focus on transition of care and support for both providers and patients through standardized tools and multidisciplinary coordination can help strengthen long-term outcomes and support patient continuity of care.

KDIGO offers a practical, family-centered framework for pediatric nephrologists to individualize care and support children and adolescents in building self-management skills essential for adulthood. By pairing medical recommendations with principles of shared decision-making and structured transition protocols, the 2025 guidelines lay the groundwork for a more holistic and forward-thinking

approach to pediatric ADPKD care.

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