Autosomal Dominant Polycystic Kidney Disease. KDIGO 2025 Guideline, a Belgian Paediatric Perspective

Tessa Van der Auwera [a,*], Femke Elen [a,*], Djalila Mekahli [a,b]

[*] These authors contributed equally to this work and share first authorship

[a] Department of Paediatric Nephrology, University Hospitals Leuven, Leuven, Belgium

[b] PKD Research Group, Department of Cellular and Molecular Medicine, KU Leuven, Leuven, Belgium

djalila.mekahli@uzleuven.be

Keywords

Polycystic kidney, autosomal dominant ; genotype ; risk factors ; counselling ; child.

Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is traditionally viewed as an adult-onset condition. However, increasing evidence highlights a broad phenotypic and genotypic spectrum in children, including very early-onset cases. Despite the absence of curative therapies, early identification of modifiable risk factors such as arterial hypertension, proteinuria, and obesity may delay progression and improve long-term outcomes. This narrative review provides a Belgian paediatric perspective on the updated KDIGO 2025 guidelines for ADPKD. We discuss the clinical variability of paediatric ADPKD, the role of genotype in disease severity, and the emerging paediatric-specific risk stratification tools to identify children at risk of rapid progression. We further explore the benefits and considerations of screening at-risk children and offer practical recommendations for diagnosis, counselling, and early management. By raising awareness among general paediatricians, we aim to promote timely intervention and structured follow-up for affected children and their families.

Introduction

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is an inherited disorder associated with multiple bilateral kidney cysts and several extrarenal manifestations (1–4). ADPKD is the most prevalent hereditary kidney disease and the fourth most common cause of kidney replacement therapy in adult patients (1). The estimated prevalence of ADPKD is one in 1000 individuals (2,3).

The typical phenotype of ADPKD is characterized by multiple bilateral kidney cysts leading to progressive kidney enlargement. The first clinical symptom is generally arterial hypertension (HTA) while decline in kidney function often occurs later in the disease. Common complications in adults with ADPKD include abdominal discomfort or pain, cyst haemorrhage, gross haematuria, nephrolithiasis and cyst infections (2,3). Extrarenal manifestations of ADPKD may include liver cysts, increased risk of intracranial aneurysms, pancreatic cysts, cardiac valvular disease and abdominal hernias (2,3).

ADPKD is often regarded as a silent disease, as significant kidney cyst enlargement and subsequent decline in kidney function typically occur at an adult age. Progression to kidney failure most commonly takes place after the fourth decade of life. However, ADPKD exhibits a broad clinical spectrum, encompassing paediatric manifestations and, in some cases, the detection of prenatal cysts (1). Therefore, this narrative review aims to outline the phenotypic and genotypic complexity of paediatric ADPKD, identify modifiable risk factors, and discuss the potential benefits and challenges associated with paediatric or adolescent screening in the context of familial ADPKD. Additionally, we provide recommendations for the management of ADPKD in paediatric patients according to the recent KDIGO (Kidney Disease: Improving Global Outcomes) guidelines.

Phenotype complexity

As outlined in the introduction, ADPKD is most commonly recognized as an adult-onset condition. However, its broad phenotypic spectrum also encompasses paediatric presentations, which may range from asymptomatic incidental findings of cyst on ultrasound, to HTA or proteinuria, to symptomatic disease such as pain, enuresis, haematuria or urinary tract or cyst infections (1,5,6). Recent evidence further suggests that kidney cyst formation already starts in utero (7,8).

A recent study on paediatric ADPKD utilized data from multiple registries, including the multinational ADPedKD registry (www.ADPedKD.org), the European Rare Kidney Disease Registry (ERKReg), and the United Kingdom RaDaR registry, encompassing data from 32 countries (1). The study aimed to characterize the genotypic and phenotypic features of ADPKD in the paediatric population (Table 1). The most common mode of presentation in this study was diagnosis through family screening, followed by postnatal incidental findings and clinical signs and symptoms. Since 2000, the prevalence of prenatal diagnosis has increased,

TABLE 1: Genotypic and phenotypic features of ADPKD in the paediatric population.

Mode of presentation	ADPedKD (N=950)	ERKReg (N=596)	RaDaR (N=139)
Family screening	48% (455)	62% (370)	Unknown
Prenatal	14% (129)	2% (13)	11% (15)
Incidental findings	31% (293)	21% (126)	Unknown
Clinical symptoms	8% (73)	15% (87)	Unknown

Adapted from Gimpel et al. 2025 (1). ADPedKD, ADPKD registry; ADPKD, autosomal dominant polycystic kidney disease; ERKReg, European Rare Kidney Disease Registry; RaDaR, National Registry of Rare Kidney Diseases. Mentioned as % (N).

primarily driven by advancements in the accuracy and utilization of prenatal ultrasound imaging. These prenatal cases usually exhibit hyperechogenic, enlarged kidneys rather than the typical cysts seen in classical presentations. Given that not all prenatal cases progress to very early-onset (VEO) ADPKD, counselling at an expert centre is essential to provide accurate prognostic information and prenatal reassurance.

Not only does the mode of presentation vary considerably in paediatric ADPKD, but the age of symptom onset also spans a broad range. VEO-ADPKD is typically diagnosed either in utero, characterized by hyperechogenic, enlarged kidneys and oligohydramnios, or between birth and 18 months of age, when affected infants may present with enlarged cystic kidneys, HTA, and/or reduced estimated glomerular filtration rate (eGFR) (3,4). Children with VEO-ADPKD have been shown to have a higher likelihood of developing HTA and impaired kidney function (eGFR < 90 mL/min/1.73 m²) by adolescence compared to those diagnosed later in childhood (4). Early-onset ADPKD is defined by the emergence of ADPKD-related clinical manifestations between 18 months and 15 years of age (3). A third group comprises children diagnosed with ADPKD who do not meet the previous criteria. The most severe phenotypes observed in childhood are often associated with digenic inheritance involving pathogenic variants in two or more ADPKD-associated genes (4). The genetic basis of ADPKD is further discussed in the following section.

Genotype complexity

The two major genes implicated in ADPKD are *PKD1*, located on chromosome 16, and *PKD2*, located on chromosome 4. These genes encode polycystin-1 and polycystin-2, respectively. Together, *PKD1* and *PKD2* account for over 90% of diagnosed familial ADPKD cases, with *PKD1* being the more prevalent of the two (2,3). The clinical phenotype of ADPKD varies depending on the genes involved. Mutations in *PKD2* are associated with a milder disease course, with affected individuals typically reaching kidney failure approximately two decades later than those with *PKD1* mutations (4,9). Among individuals with *PKD1* mutations, those carrying truncating variants tend to exhibit a more severe phenotype than those with non-truncating variants (4,9).

A small proportion of ADPKD cases are attributed to pathogenic variants in less common genes, for which pathogenicity is well supported (*IFT140, ALG5, ALG9, GANAB, DNAJB11, and NEK8*) (3). Although these genes are associated with distinct phenotypic features, they often show clinical overlap with *PKD1* and *PKD2*-associated disease. For instance, *PKD1*, *PKD2* and *DNAJB11* mutations have all been linked to a substantial risk of progression to kidney failure. In contrast, several additional genes (*ALG6*, *ALG8*, *PKHD1*) have been proposed as potential contributors to cystic kidney disease, but their pathogenicity in the context of ADPKD remains uncertain (3).

VEO-ADPKD represents a rare and particularly severe clinical presentation, occurring in less than 1% of affected families. These cases frequently exhibit an oligogenic inheritance pattern, where the co-inheritance of multiple rare variants, such as compound heterozygous mutations in *PKD1* or *PKD2* or additional mutations in other cystogenes, contributes to an earlier and more severe disease phenotype than typically seen in classical ADPKD (2). Evidence from recent case series demonstrates that children with severe, early-onset cystic kidney disease often carry two rare *PKD1* variants in trans, supporting a dosage effect and a model in which disease severity is modified by the cumulative impact of multiple genetic variants rather than a strictly monogenic mechanism (10). Furthermore, large deletions involving both *PKD1* and the adjacent TSC2 gene can result in a combined phenotype of ADPKD and tuberous sclerosis complex, so called "contiguous gene syndrome PKD1-TSC2", leading to rapid progression to kidney failure in childhood. The identification of concomitant nephropathies in some patients who progress to kidney failure earlier than expected based on their genotype further underscores the complexity of genetic and phenotypic interactions in VEO-ADPKD (2). These findings highlight the importance of comprehensive gene panel analysis using next-generation sequencing for accurate diagnosis, prognosis, and genetic counselling in early-onset cases.

Modifiable risk factors

Several non-modifiable factors influence the disease course of ADPKD, including age at symptom onset, underlying genotype, ethnicity, and sex. However, a number of modifiable factors have been identified that may impact long-term disease progression. These include body weight, blood pressure control, presence of proteinuria, level of physical activity, smoking status, and dietary salt intake (4).

HTA is the most common and earliest clinical feature of ADPKD, typically developing before the age of thirty (4). Early-onset HTA is a key risk factor for progression to kidney failure and is strongly correlated with total kidney volume (TKV) and cyst burden, more than other kidney manifestations (11,12). Since cardiovascular disease is the leading cause of death in ADPKD, it is essential for us as paediatricians to regularly monitor and manage blood pressure to improve long-term outcomes.

HTA is estimated to affect 20-40% of children and adolescents with ADPKD, with a prevalence notably higher than in the general paediatric population (4). A meta-analysis reported HTA in approximately 20% of children with ADPKD (9,11). Data from a recent retrospective international multicentre study, including 310 paediatric ADPKD patients from 22 European centres, revealed even higher prevalence rates using 24-hour ambulatory blood pressure monitoring (ABPM) (13). 24-hour ABPM identified HTA in 31% of patients during daytime, 42% during nighttime, and 35% over the full 24-hour period. Additionally, 52% exhibited non-dipping nocturnal blood pressure patterns, while 18% had isolated nocturnal HTA (13). These findings highlight the clinical value of ABPM in the routine follow-up of paediatric patients with ADPKD.

HTA is one of the earliest treatable manifestations of ADPKD in children and adolescents. Studies have demonstrated a correlation between elevated blood pressure and both total kidney volume and cyst volume in this population (4). Children with ADPKD who develop HTA exhibit faster kidney growth and a more rapid decline in kidney function compared to their normotensive peers (4). Furthermore, blood pressure values in the high-normal range (75^{th}–90^{th} percentile) have been associated with an increased

TABLE 2: Risk categories according to the Leuven Imaging Classification (LIC) based on height-adjusted total kidney volume measured by 3D ultrasound.

Risk category		Meaning
A	Very low risk	Children with kidney volumes in the normal range for age and minimal risk of progression
B	Low risk	Children with mild kidney enlargement, typically without clinical signs
C	Intermediate risk	Children with moderate kidney growth, sometimes accompanied by early hypertension or microalbuminuria
D	High risk	Significant or extreme kidney enlargement, often associated with *PKD1* mutations, higher blood pressure, and early decline in kidney function
E	Very high risk	

long-term cardiovascular risk (4). In children, blood pressure above the 75th percentile has also been linked to elevations in left ventricular mass index (14).

Proteinuria represents another early and potentially modifiable manifestation of ADPKD in children. Estimates suggest that microalbuminuria affects 20-48% and proteinuria 10–23% of paediatric patients with ADPKD (9,11,15). Both paediatric and adult cohort studies have demonstrated that the presence of proteinuria is associated with a more aggressive course of kidney disease (15). Moreover, proteinuria is recognized as a modifiable risk factor for the progression of chronic kidney disease (CKD) in children (16).

Overweight and obesity are recognized modifiable risk factors in ADPKD. Overweight in children and adolescents is defined as a body mass index (BMI) between the 85th and 95th percentiles for age and sex, whereas obesity is defined as a BMI at or above the 95th percentile. In adult cohorts, a higher body mass index (BMI) has been identified as an independent risk factor of disease progression (17,18). A post-hoc analysis of the TEMPO 3:4 trial demonstrated that overweight and, particularly, obesity were strongly and indepentdently associated with kidney growth in adult patients (17). Also, analysis of participants of the HALT-PKD trials showed an increased risk of progression to kidney failure in adolescents and adults with early-stage ADPKD (18). Although paediatric data on the effect of BMI on children with ADPKD are currently lacking, recent KDIGO guidelines recommend regular assessment of BMI in children and adolescents and emphasize the importance of lifestyle counselling to mitigate potential long-term risks based on adult data (4).

In addition to the importance of monitoring BMI for ADPKD progression, overweight and obesity in childhood also carry other significant long-term health consequences such as increased risk for cardiovascular disease, type 2 diabetes mellitus, hypertension, dyslipidaemia, and non-alcoholic fatty liver disease. The prevalence of childhood obesity is slowly increasing, projections suggest that by 2035 more than 750 million children (age 5–19 years) are expected to be living with overweight and obesity that if no substantial interventions are implemented (19). Early identification through routine BMI screening and addressing social determinants of health are essential to mitigate the adverse outcomes (20).

Stratification models

Risk stratification is becoming an essential component of care in paediatric ADPKD, especially as interest grows in early monitoring and timely intervention. Although adult risk scores such as the PROPKD model incorporate genetic and clinical features, they have not yet been validated in children (12). Also, the Mayo Imaging Classification (MIC) is a well-established tool that uses height-adjusted total kidney volume (htTKV) measured by MRI or CT to predict kidney prognosis and guide clinical trial enrolment for adults with ADPKD (21,22).

Recently, the Leuven Imaging Classification (LIC) was developed by Breysem et al. in 2023, and validated as a paediatric-specific imaging-based risk model (23). LIC combines 3D ultrasound-derived htTKV with age to assign children into five risk categories (A–E), offering a non-invasive, age-appropriate tool for clinical stratification (Figure 1 and Table 2). These categories help clinicians identify which children may benefit from closer monitoring and early lifestyle or therapeutic interventions. In a large multicentre study, LIC class was significantly associated with both genotype and blood pressure, reinforcing its clinical relevance (24).

Importantly, the LIC relies on 3D ultrasound. Ultrasound is generally more accessible and practical than magnetic resonance imaging (MRI) in paediatric settings, particularly for younger children, however, 3D ultrasound is not yet routinely implemented in clinical practice. Therefore, the Leuven PKD research group is currently developing and validating a 2D ultrasound-based model as an alternative.

Paediatric-specific models like the LIC provide a more tailored approach. Nevertheless, integrating clinical factors such as hypertension, obesity, and family history remains essential for comprehensive risk assessment (25,26). As research progresses, LIC may serve as a foundation for future paediatric stratification and prediction models.

Screening of at risk children in families with ADPKD

The necessity of screening or diagnosing ADPKD in at-risk children remains a subject of ongoing debate, largely due to the perception of ADPKD as an adult-onset condition. However, as outlined in the preceding sections, the disease presents with a broad clinical

FIGURE 1: Incorporation of the validation cohort (blue dots) in the Leuven classification paediatric model (full lines), with the MIC model extrapolated to the paediatric age range as comparison (dashed lines). Adapted from Breysem L, et al (2023). Risk Severity Model for Paediatric Autosomal Dominant Polycystic Kidney Disease Using 3D Ultrasound Volumetry. Clinical Journal of the American Society of Nephrology (CJASN), 18(5), 581-591. https://journals.lww.com/cjasn/. Used with permission.

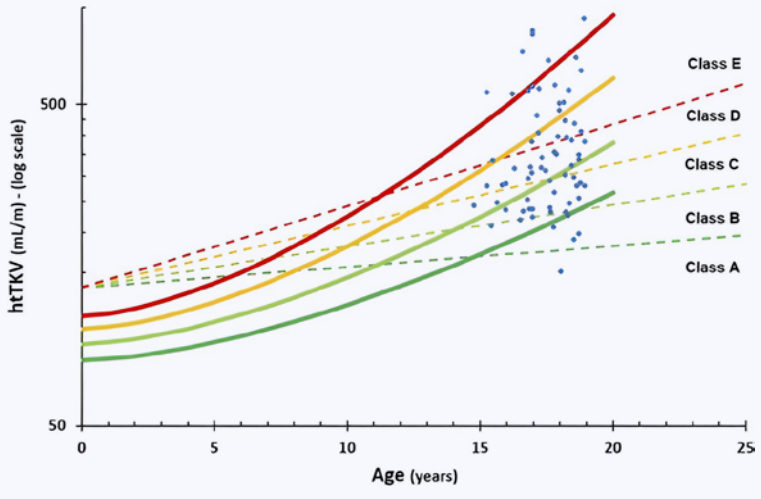
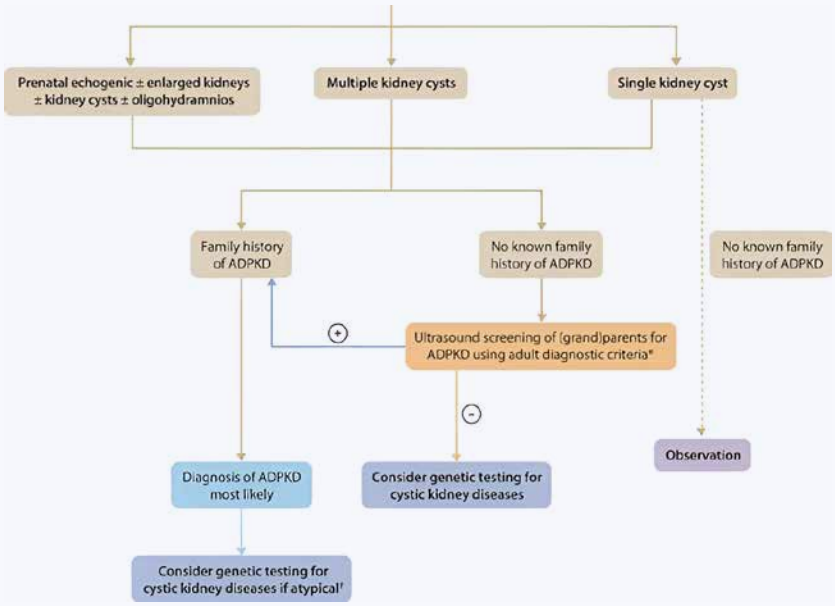


FIGURE 2: Diagnosis of children with clinical consideration of autosomal dominant polycystic kidney disease (ADPKD). Dash lines denote other pathway for consideration. *Consider screening grandparents if parent screening is negative or parents are aged<40 years. †For example, very early onset ADPKD; severe kidney involvement relative to age. From Kidney Disease: Improving Global Outcomes (KDIGO) ADPKD Work Group. KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease (ADPKD). Kidney Int. February 2025;107(2S):S1-239. Used with permission.



spectrum in childhood. Hypertension and proteinuria can occur in children even in the absence of overt symptoms. So early screening provides an opportunity to identify and address modifiable risk factors and timely intervention may confer significant long-term benefits, as previously discussed (9).

One of the main arguments against screening or diagnosing ADPKD during childhood is the absence of a definitive cure (7). However, despite the lack of curative treatment, early lifestyle interventions and the management of modifiable risk factors can yield significant long-term benefits for affected individuals. Another frequently cited concern is the potential impact on insurance and future employment opportunities (1,5). While presymptomatic genetic testing may be legally protected from disclosure to insurance companies, no such protections currently apply to radiological findings (9).

The psychological burden of diagnosis and follow-up on both children and their families is also acknowledged (4). However, current guidelines recommend monitoring for hypertension, proteinuria, and other early manifestations in at-risk children, regardless of a confirmed diagnosis. On the other hand, since each child of an affected parent has only a 50% chance of inheriting the disease, children without the mutation could potentially be spared from unnecessary testing and repeated hospital visits.

A final argument often raised is the concern that screening during childhood removes the individual's autonomy to make an informed decision about knowing their disease status later in life (2). While this perspective is valid in conditions without therapeutic consequences during childhood, in the case of ADPKD, early intervention targeting modifiable risk factors may positively influence disease progression.

A family-centred approach is essential when guiding families in the decision-making process regarding testing. Open and balanced discussions should address the potential benefits and harms of both pursuing and deferring testing in children at risk for ADPKD. Therefore these families would benefit from counselling in an expert centre.

Ultrasound is the recommended imaging modality for testing or diagnosing ADPKD in children (2,4,5). The presence of a single kidney cyst in a child under the age of 15 with a positive family history of ADPKD is considered highly suggestive of the disease (4). However, the absence of kidney cysts does not exclude the diagnosis. In cases where kidney cysts are detected in a child without a known family history, the KDIGO guidelines recommend performing kidney ultrasound in the parents or in the grandparents if the parents are younger than 40 years of age to investigate a possible familial origin (Figure 2).

Genetic testing is recommended in children with VEO-ADPKD or those presenting with atypical clinical features (4). In children with a positive family history who do not meet these specific criteria, genetic testing may still be considered and discussed with the family, taking into account the potential benefits and harms. Additionally, genetic screening is recommended in children with cystic kidneys and no known family history, in order to confirm the diagnosis and guide further management. Expert counselling is needed for these complex cases.

Management

Even in the absence of a definitive cure for ADPKD, identifying at-risk children remains essential. Early screening and timely management of modifiable risk factors can significantly influence disease progression. Moreover, even when modifiable risk factors are not present, recognizing children at risk allows for early lifestyle counselling, which may positively impact long-term outcomes.

According to the KDIGO guidelines, counselling on lifestyle factors is essential in the management of patients with or at risk for ADPKD. Maintaining a healthy body weight is emphasized. Therefore, adherence to general dietary recommendations and engagement in regular physical activity are recommended (3,4). In children and adolescents with ADPKD who present with overweight or obesity, implementing these lifestyle modifications is crucial, with the goal of achieving and maintaining a healthy weight. A multidisciplinary approach is often necessary to effectively support these interventions. Additional lifestyle factors, particularly relevant during adolescence, include avoiding alcohol consumption and tobacco products, recreational drugs, and anabolic steroids (3,4). Avoidance of tobacco use is especially critical, as smoking represents a significant modifiable risk factor for kidney cysts growth and the development and rupture of intracranial aneurysms (4).

Recent KDIGO guidelines recommend annual office blood pressure measurements in all children with and at risk for ADPKD. In children aged ≥ 5 years with very early-onset or early-onset ADPKD, as well as in those with a family history of ADPKD and office blood pressure readings ≥ the 75th percentile, annual 24-hour ABPM is advised (3). Additionally, echocardiographic evaluation is recommended in children with ADPKD and confirmed hypertension, due to their increased risk of early-onset cardiovascular complications (4,14).

If hypertension (defined as BP above 75^{the} percentile for age) is present, referral to an expert centre is necessary. The KDIGO guidelines recommend angiotensin-converting enzyme inhibitors

(ACEi) or angiotensin receptor blockers (ARB) as first-line treatment (3,4). The target blood pressure is below the 50th percentile or below 110/70 mmHg in adolescents. The importance of identifying and treating arterial hypertension lies in its significant impact on both kidney function and long-term cardiovascular risk, as outlined in the section on modifiable risk factors. In addition, ACEi and ARB are also recommended for the treatment of proteinuria in children with chronic kidney disease (4). The initiation of treatment need to be discussed and the side effects need to be considered with education of the patient.

When adolescent girls with ADPKD consider starting contraception, it is important to avoid oestrogen-containing contraceptives. In patients with polycystic liver disease associated with ADPKD, oestrogen exposure has been linked to greater liver volume and a higher annual increase in liver volume (4).

The only available drug to this day is the vasopressin type 2 antagonist tolvaptan, which slows down disease progression but has only been approved in rapidly progressing adult patients and is associated with severe adverse effects of polyuria and polydipsia. Although a single RCT has indicated tolerability and suggests potential effect of tolvaptan on annual TKV expansion in children with ADPKD (27). There is insufficient evidence for the use of any targeted or disease-modifying treatments for ADPKD in affected children at this time.

Conclusion

While ADPKD is classically regarded as an adult disease, a growing body of evidence confirms that clinical manifestations and kidney damage can begin in early childhood. Paediatric ADPKD encompasses a wide spectrum - from asymptomatic to severe early-onset disease - and requires careful clinical judgement to guide follow-up and intervention. Identifying modifiable risk factors such as hypertension, proteinuria, and excess weight is essential, as their timely management may positively influence the disease trajectory. The development of the LIC marks an important advance in paediatric-specific risk stratification and provides a practical tool for clinicians. Though routine screening remains debated, an individualised, family-centred approach is key to counselling and decision-making and best coordinated in an expert centre. With increasing availability of genetic testing and improved imaging modalities, early recognition and multidisciplinary management can optimize outcomes for children at risk of ADPKD. The ADPedKD initiative (www.ADPedKD.org) lays the foundation for a more personalized and evidence-based approach to paediatric ADPKD. Its findings are expected to inform future clinical guidelines and improve long-term care for affected children worldwide.

The authors have no conflict of interest to declare.

REFERENCES

1. Gimpel C, Fieuws S, Hofstetter J, Pitcher D, Vanmeerbeek L, Haeberle S, e.a. Insights from ADPedKD, ERKReg and RaDaR registries provide a multi-national perspective on the presentation of childhood autosomal dominant polycystic kidney disease in high- and middle-income countries. Kidney Int. 2025;108(1):105-18.
2. Cornec-Le Gall E, Alam A, Perrone RD. Autosomal dominant polycystic kidney disease. Lancet Lond Engl. 2019;393(10174):919-35.
3. Torres VE, Ahn C, Barten TRM, Brosnahan G, Cadnapaphornchai MA, Chapman AB, e.a. KDIGO 2025 clinical practice guideline for the evaluation, management, and treatment of autosomal dominant polycystic kidney disease (ADPKD): executive summary. Kidney Int. februari 2025;107(2):234-54.
4. Kidney Disease: Improving Global Outcomes (KDIGO) ADPKD Work Group. KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of Autosomal Dominant Polycystic Kidney Disease (ADPKD). Kidney Int. 2025;107(2S):S1-239.
5. Gimpel C, Bergmann C, Bockenhauer D, Breysem L, Cadnapaphornchai MA, Cetiner M, e.a. International consensus statement on the diagnosis and management of autosomal dominant polycystic kidney disease in children and young people. Nat Rev Nephrol. 2019;15(11):713-26.
6. Bergmann C, Zerres K. Early manifestations of polycystic kidney disease. Lancet Lond Engl. 30 2007;369(9580):2157.
7. De Rechter S, Kringen J, Janssens P, Liebau MC, Devriendt K, Levtchenko E, e.a. Clinicians' attitude towards family planning and timing of diagnosis in autosomal dominant polycystic kidney disease. PloS One. 2017;12(9):e0185779.
8. Grantham JJ, Cook LT, Wetzel LH, Cadnapaphornchai MA, Bae KT. Evidence of extraordinary growth in the progressive enlargement of renal cysts. Clin J Am Soc Nephrol CJASN. 2010;5(5):889-96.
9. Gimpel C, Bergmann C, Mekahli D. The wind of change in the management of autosomal dominant polycystic kidney disease in childhood. Pediatr Nephrol Berl Ger. 2022;37(3):473-87.
10. Bergmann C, von Bothmer J, Ortiz Brüchle N, Venghaus A, Frank V, Fehrenbach H, e.a. Mutations in multiple PKD genes may explain early and severe polycystic kidney disease. J Am Soc Nephrol. 2011;22(11):2047-56.
11. Marlais M, Cuthell O, Langan D, Dudley J, Sinha MD, Winyard PJD. Hypertension in autosomal dominant polycystic kidney disease: a meta-analysis. Arch Dis Child. 2016;101(12):1142-7.
12. Cornec-Le Gall E, Audrézet MP, Rousseau A, Hourmant M, Renaudineau E, Charasse C, e.a. The PROPKD Score: A New Algorithm to Predict Renal Survival in Autosomal Dominant Polycystic Kidney Disease. J Am Soc Nephrol JASN. 2016;27(3):942-51.
13. Massella L, Mekahli D, Paripović D, Prikhodina L, Godefroid N, Niemirska A, e.a. Prevalence of Hypertension in Children with Early-Stage ADPKD. Clin J Am Soc Nephrol CJASN. 2018;13(6):874-83.
14. Cadnapaphornchai MA, McFann K, Strain JD, Masoumi A, Schrier RW. Increased left ventricular mass in children with autosomal dominant polycystic kidney disease and borderline hypertension. Kidney Int. 2008;74(9):1192-6.
15. De Rechter S, Breysem L, Mekahli D. Is Autosomal Dominant Polycystic Kidney Disease Becoming a Pediatric Disorder? Front Pediatr. 2017;5:272.
16. ESCAPE Trial Group, Wühl E, Trivelli A, Picca S, Litwin M, Peco-Antic A, e.a. Strict blood-pressure control and progression of renal failure in children. N Engl J Med. 2009;361(17):1639-50.
17. Nowak KL, Steele C, Gitomer B, Wang W, Ouyang J, Chonchol MB. Overweight and Obesity and Progression of ADPKD. Clin J Am Soc Nephrol. 2021;16(6):908.
18. Nowak KL, Copeland TP, Ku E, Sarnak MJ, Gitomer B, Abebe KZ, e.a. Overweight Status, Obesity, and Progression to ESKD in Patients with Autosomal Dominant Polycystic Kidney Disease. Clin J Am Soc Nephrol CJASN. 2025;20(4):520-8.
19. Schreurs L, Gies I, Van der Schueren B, De Cock D. Systematic literature review on the awareness of obesity in adults and children living with obesity, the general public and healthcare professionals. Int J Obes 2025 Jul 23. doi: 10.1038/s41366-025-01855-w. Online ahead of print.
20. Hampl S, Hassink S, Skinner A, Armstrong S, Barlow S, Bolling C, e.a. Clinical Practice Guideline for the Evaluation and Treatment of Children and Adolescents With Obesity. Pediatrics. 2023;151(2):e2022060640.
21. Irazabal M, Rangel L, Bergstralh E, Osborn S, Harmon A, Sundsbak J, e.a. Imaging classification of autosomal dominant polycystic kidney disease: a simple model for selecting patients for clinical trials. J Am Soc Nephrol. 2025;26(1):160-72.
22. McEwan P, Bennett Wilton H, Ong ACM, Ørskov B, Sandford R, Scolari F, e.a. A model to predict disease progression in patients with autosomal dominant polycystic kidney disease (ADPKD): the ADPKD Outcomes Model. BMC Nephrol. 2018;19(1):37.
23. Breysem L, De Keyzer F, Schellekens P, Dachy A, De Rechter S, Janssens P, e.a. Risk Severity Model for Pediatric Autosomal Dominant Polycystic Kidney Disease Using 3D Ultrasound Volumetry. Clin J Am Soc Nephrol. 2023;18(5):581-91.
24. Yilmaz K, Saygili S, Canpolat N, Akgun-Dogan O, Yuruk Yildirim ZN, Cicek-Oksuz RY, e.a. Magnetic resonance imaging based kidney volume assessment for risk stratification in pediatric autosomal dominant polycystic kidney disease. Front Pediatr. 2024;12:1357365.
25. De Rechter S, Bockenhauer D, Guay-Woodford LM, Liu I, Mallett AJ, Soliman NA, e.a. ADPedKD: A Global Online Platform on the Management of Children With ADPKD. Kidney Int Rep. 2019;4(9):1271-84.
26. Dachy A, Van Loo L, Mekahli D. Autosomal Dominant Polycystic Kidney Disease in Children and Adolescents: Assessing and Managing Risk of Progression. Adv Kidney Dis Health. 2023;30(3):236-44.
27. Mekahli D, Guay-Woodford L, Cadnapaphornchai M, Greenbaum L, Litwin M, Seeman T, e.a. Tolvaptan for Children and Adolescents with Autosomal Dominant Polycystic Kidney Disease: Randomized Controlled Trial. Clin J Am Soc Nephrol. 2023;18(1):36-46.