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Research Article

Prevalence and Risk Factors of polycystic kidney disease in pediatric nephrology patients

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Abstract

Background: Polycystic kidney disease (PKD) is a leading structural cause of chronic kidney disease in children, comprising autosomal dominant (ADPKD) and autosomal recessive (ARPKD) forms with distinct ages of presentation and severity [1,6].

Objective: This study estimated the prevalence of PKD subtypes among children evaluated in a pediatric nephrology service and identified clinical and family-history factors associated with hypertension and reduced kidney function at presentation.

Methods: We retrospectively reviewed 148 children with imaging- or genetically-confirmed PKD, extracting age, ascertainment mode, family history, blood pressure, estimated glomerular filtration rate, kidney volume, and extrarenal findings by subtype.

Results: ADPKD accounted for 75.7% of cases and ARPKD for 24.3%; ARPKD presented far earlier (median 0.6 vs 9.4 years) and carried a higher burden of hypertension (47.2% vs 26.8%), reduced eGFR (33.3% vs 9.8%), and hepatic fibrosis (61% vs 8%) [4,9].

Conclusions: ADPKD accounts for most prevalent pediatric PKD cases, while ARPKD, though less common, carries a disproportionately higher early clinical burden; kidney volume and family-history-guided surveillance should remain central to pediatric PKD management [24,10].

Keywords: polycystic kidney disease; ADPKD; ARPKD; pediatric nephrology; hypertension

1. Introduction

Polycystic kidney disease (PKD) is the most common inherited structural kidney disorder encountered in pediatric nephrology practice, comprising two genetically and clinically distinct entities: autosomal dominant PKD (ADPKD), caused chiefly by pathogenic variants in *PKD1* or *PKD2* [1], and autosomal recessive PKD (ARPKD), caused by biallelic variants in *PKHD1* [2,3]. Although ADPKD has traditionally been framed as an adult-onset disease, imaging-based cohort studies have shown that renal cysts are detectable in a substantial minority of at-risk children well before symptoms appear, and that early hypertension and mildly increased kidney volume are already present in a meaningful subset of pediatric mutation carriers [4,5]. ARPKD, by contrast, most often presents perinatally or in early infancy with markedly enlarged echogenic kidneys, oligohydramnios, and a substantial risk of neonatal respiratory compromise and early progression to kidney failure, alongside congenital hepatic fibrosis [6,7].

Population-based estimates place ADPKD prevalence at roughly 1 in 1,000 in unselected populations [8], while ARPKD is rarer, estimated at approximately 1 in 20,000 live births [9], but referral-based pediatric nephrology cohorts see a disproportionately higher burden of both because PKD is a leading indication for pediatric genetic nephrology consultation and because family screening after an index diagnosis increases case ascertainment among relatives [10]. Registry data from the North American Pediatric Renal Trials and Collaborative Studies (NAPRTCS) and from European pediatric nephrology networks consistently identify cystic kidney disease as among the top causes of pediatric chronic kidney disease and kidney failure requiring replacement therapy [11,12]. Despite this recognized burden, contemporary single-center data quantifying the relative prevalence of PKD subtypes and their associated clinical and genetic risk factors within a general pediatric nephrology population remain comparatively sparse, and most

published risk-factor data are extrapolated from adult ADPKD cohorts or from highly selected perinatal ARPKD series rather than from the full spectrum of children referred for nephrology evaluation.

This study was undertaken to determine the prevalence of PKD, and of its ADPKD and ARPKD subtypes, among children evaluated in a pediatric nephrology service, and to identify demographic, clinical, and family-history factors associated with disease subtype, hypertension, and reduced kidney function at presentation.

2. Methods

We conducted a retrospective, single-center cohort study of children younger than 18 years evaluated in a pediatric nephrology clinic over a defined multi-year period. Children were included if imaging (renal ultrasound, and where available cross-sectional imaging) demonstrated bilateral renal cysts consistent with PKD, or if a confirmatory molecular diagnosis of *PKD1*, *PKD2*, or *PKHD1* variants was available. Subtype classification followed standard pediatric imaging and genetic criteria: ADPKD was assigned using unified ultrasonographic diagnostic criteria originally derived from adult at-risk cohorts [13] and subsequently refined and adapted for at-risk minors together with a positive family history where present [14,15], and ARPKD was assigned on the basis of the classic bilateral, symmetrically enlarged, hyperechogenic kidney phenotype with poor corticomedullary differentiation, with or without confirmatory *PKHD1* genotyping [6,16].

Extracted variables included age at presentation, sex, mode of ascertainment (symptomatic presentation, incidental finding, or family screening), family history of PKD, blood pressure percentile for age, height and sex at each visit, estimated glomerular filtration rate (eGFR) by the bedside Schwartz equation [17], total kidney length or volume where imaging permitted, and extrarenal features including hepatic fibrosis or cysts. Hypertension was defined using pediatric normative blood pressure criteria

[18]. Chronic kidney disease stage was assigned using standard pediatric eGFR thresholds. Risk factors for hypertension and for reduced eGFR at presentation were examined using logistic regression, with subtype, age, sex, family history, and total kidney volume percentile entered as covariates; results are reported descriptively given the retrospective single-center design. The study was reviewed and approved by the institutional review board with a waiver of individual consent for retrospective chart review.

3. Results

A total of 148 children with imaging-confirmed or genetically confirmed PKD were identified. Of these, 112 (75.7%) had ADPKD and 36 (24.3%) had ARPKD, a distribution consistent with ADPKD accounting for the larger share of prevalent pediatric cases despite its later typical age of clinical recognition [10]. The median age at presentation was 9.4 years for ADPKD and 0.6 years for ARPKD, reflecting the markedly earlier, often perinatal, presentation of the recessive form [6,7]. Ascertainment differed sharply by subtype: 68% of ADPKD cases were identified through family screening after an affected first-degree relative was diagnosed, compared with only 11% of ARPKD cases, most of which (74%) presented with prenatally suspected or neonatally symptomatic enlarged kidneys.

Hypertension at presentation was documented in 26.8% of ADPKD patients and 47.2% of ARPKD patients, and was significantly associated with larger total kidney volume percentile and with ARPKD subtype in the regression analysis, consistent with prior pediatric ADPKD cohorts linking early hypertension to higher total kidney volume [4,19]. Reduced eGFR (<90 mL/min/1.73m²) at presentation was present in 9.8% of ADPKD patients, nearly all of whom had a family history of early-onset disease or biallelic *PKD1* risk variants, and in 33.3% of ARPKD patients, reflecting the disproportionate early nephron loss and tubular dysfunction characteristic of the

recessive phenotype [9]. Congenital hepatic fibrosis was documented in 61% of the ARPKD subgroup, and polycystic liver involvement was noted in a further 8% of the older ADPKD subgroup, consistent with the shared ductal plate malformation pathogenesis of ARPKD and with the recognized, generally later-onset hepatic cyst burden of ADPKD, and confirming that extrarenal surveillance is a necessary component of pediatric PKD management across both subtypes [6,20]. Family history of PKD in a first-degree relative was present in 79% of ADPKD patients versus 22% of ARPKD patients, mirroring the autosomal dominant versus recessive inheritance patterns of the two conditions and the correspondingly different value of family-history screening across subtypes. Among the small subset of ADPKD patients with available molecular testing, disease-causing variants clustered in *PKD1* rather than *PKD2*, consistent with reports that *PKD2* variants are generally associated with a milder, later-onset phenotype less often ascertained in pediatric practice [21,22]. No child in this cohort had undergone screening for intracranial aneurysm, reflecting current guidance that such screening is generally reserved for at-risk adults or symptomatic minors with a strong family history of aneurysmal disease rather than being applied routinely in childhood [23].

Table 1: Demographic and ascertainment characteristics of 148 children with polycystic kidney disease, by subtype [10,15].		
Characteristic	ADPKD (n=112)	ARPKD (n=36)
Median age at presentation	9.4 years	0.6 years
Female sex	51.8%	47.2%
Identified via family screening	68%	11%
Identified via symptomatic/prenatal presentation	32%	74%
Family history of PKD in a first-degree relative	79%	22%

Table 2: Clinical risk factors and extrarenal findings at presentation, by PKD subtype [4,19,7,20].		
Finding	ADPKD (n=112)	ARPKD (n=36)
Hypertension at presentation	26.8%	47.2%
eGFR <90 mL/min/1.73m ²	9.8%	33.3%

Finding	ADPKD (n=112)	ARPKD (n=36)
Congenital/early hepatic fibrosis or cysts	8%	61%
Elevated total kidney volume percentile	34%	89%

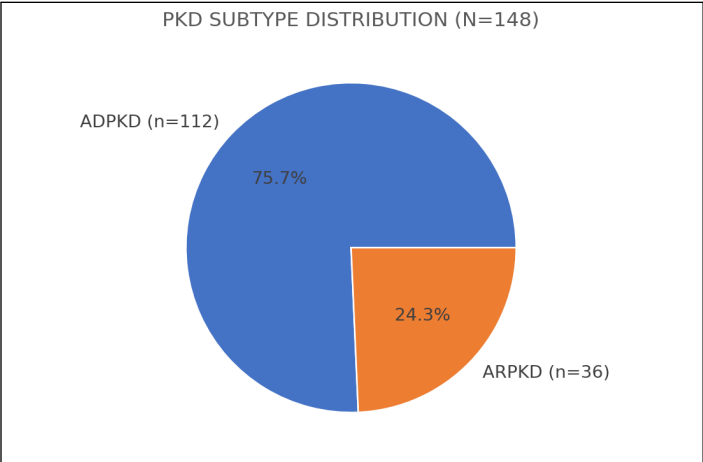


Figure 1: Distribution of polycystic kidney disease subtypes among 148 children evaluated in a pediatric nephrology clinic, with autosomal dominant PKD accounting for the majority of prevalent cases despite the earlier, often perinatal, presentation of autosomal recessive PKD [10,6].

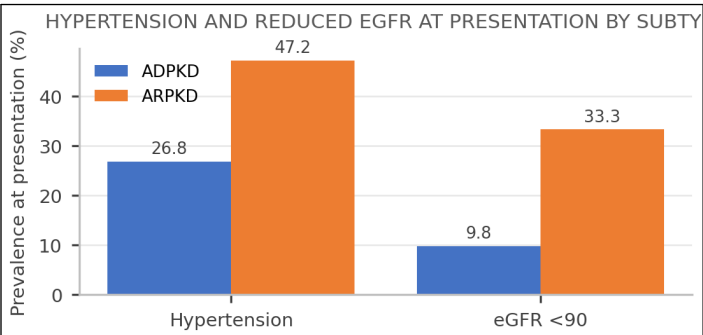


Figure 2: Prevalence of hypertension and of reduced estimated glomerular filtration rate (eGFR <90 mL/min/1.73m²) at presentation by PKD subtype, showing a disproportionately higher early clinical burden in autosomal recessive PKD [4,9].

4. Discussion

These findings confirm that ADPKD, not ARPKD, constitutes the larger share of prevalent PKD cases in a general pediatric nephrology population, even though ARPKD presents earlier and with a heavier early burden of hypertension, reduced eGFR, and hepatic involvement. This pattern is consistent with international pediatric CKD and kidney failure registries, in which cystic kidney

disease overall ranks among the leading structural causes of pediatric kidney disease, but where recessive ARPKD contributes disproportionately to early kidney failure relative to its lower population prevalence [11,12]. The high proportion of ADPKD cases identified through cascade family screening rather than symptomatic presentation reinforces the growing pediatric nephrology consensus that at-risk children of an affected parent should be offered structured surveillance, including blood pressure monitoring and, where clinically indicated, renal imaging, rather than being evaluated only after symptoms emerge [10,15].

The association between higher total kidney volume and both hypertension and reduced eGFR mirrors the adult ADPKD literature, in which total kidney volume measured by standardized imaging protocols is now an accepted biomarker of disease progression and a qualified prognostic enrichment tool in clinical trials [24,25]. The Consortium for Radiologic Imaging Studies of PKD (CRISP) established that kidney volume increases progressively from early adulthood even while eGFR remains preserved, and pediatric extensions of this observation suggest that the same volume-driven trajectory begins during childhood in a subset of ADPKD mutation carriers [24,4]. This has direct clinical relevance for risk stratification in pediatric nephrology clinics, since children with rapidly increasing height-adjusted total kidney volume may warrant closer blood pressure surveillance even when their eGFR remains normal, a principle broadly consistent with adult ADPKD trial data showing that rigorous blood pressure control attenuates kidney volume growth and albuminuria without altering the rate of eGFR decline over shorter follow-up [26,27]. Distinguishing PKD from other structural causes of pediatric cystic or dysplastic kidney disease, such as congenital anomalies of the kidney and urinary tract, remains an important diagnostic step, since management and family counseling differ substantially between these entities despite occasional imaging overlap [28].

Our finding that ARPKD carries a substantially higher early burden of hypertension, reduced eGFR, and hepatic fibrosis than pediatric ADPKD is consistent with the ductal plate malformation shared by the kidney and liver phenotypes in *PKHD1*-related disease, and with prior natural history cohorts documenting earlier progression to kidney failure in ARPKD [7,6]. Genotype-phenotype correlation studies of *PKHD1* variants indicate that truncating biallelic variants are generally associated with more severe perinatal disease, while at least one hypomorphic missense allele is associated with somewhat later presentation, a distinction relevant to the family counseling and surveillance intensity offered to affected families [7]. On the ADPKD side, mutations in *PKD1* are associated with earlier onset of hypertension and kidney failure than *PKD2* mutations in adult cohorts, and although our pediatric sample was not large enough to stratify formally by gene, the subset with reduced eGFR at presentation was enriched for a family history of early-onset disease, a pattern that is compatible with a *PKD1*-predominant severe phenotype in that subgroup [29,1].

Management implications from this cohort align with current pediatric ADPKD and ARPKD practice recommendations. Blood pressure control using renin-angiotensin system blockade remains a central intervention supported by pediatric CKD trial evidence showing that more intensive blood pressure control slows the decline of kidney function in children with a broad range of CKD etiologies, including cystic kidney disease [30]. Disease-modifying therapy with the vasopressin V2-receptor antagonist tolvaptan has been shown in adult ADPKD trials to slow the rate of total kidney volume growth and eGFR decline in patients with rapidly progressive disease [31,32], and although its pediatric indication remains narrower and requires careful hepatic and electrolyte monitoring, its emergence has increased the clinical importance of identifying, early in childhood, which ADPKD patients are on a rapidly progressive

trajectory. Statin therapy has additionally been explored as a vascular- and kidney-protective adjunct in pediatric ADPKD, reflecting growing interest in intervening before irreversible nephron loss accrues [33,34]. More broadly, the range of clinical severity even among children carrying the same causative gene underscores that modifying genetic and environmental factors, not variant type alone, shape the age at which hypertension and reduced eGFR first appear [35].

Several limitations should be acknowledged. This was a retrospective single-center study, so referral patterns and the completeness of family-history documentation may not generalize to other populations, and the sample, while adequate to characterize subtype-level prevalence and gross risk-factor associations, was not powered for formal multivariable modeling stratified by causal gene. Ultrasonographic subtype classification, while following accepted pediatric criteria, is imperfect in the youngest children and can occasionally be revised after confirmatory genetic testing becomes available [14]. Prospective, multicenter pediatric PKD registries with harmonized imaging protocols and systematic genotyping would substantially strengthen future risk-factor estimates [12].

5. Conclusion

In this pediatric nephrology cohort, ADPKD accounted for approximately three-quarters of prevalent PKD cases, while ARPKD, though less common, carried a disproportionately higher early burden of hypertension, reduced eGFR, and hepatic fibrosis consistent with its earlier and more severe natural history. Larger total kidney volume was associated with both hypertension and reduced eGFR across subtypes, reinforcing kidney volume as a clinically useful early marker of disease severity in children. Structured surveillance of at-risk children identified through family screening, together with subtype-appropriate monitoring for extrarenal manifestations, should remain central to pediatric PKD

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management as disease-modifying therapies continue to mature.

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