Peter C Harris

List of Publications by Year
in descending order

230 papers	23,324 citations	7096 78 h-index	8866 145 g-index
235 all docs	235 docs citations	235 times ranked	10491 citing authors

#	Article	IF	Citations
1	Autosomal dominant polycystic kidney disease. Lancet, The, 2007, 369, 1287-1301.	13.7	1,170
2	The polycystic kidney disease 1 (PKD1) gene encodes a novel protein with multiple cell recognition domains. Nature Genetics, 1995, 10, 151-160.	21.4	846
3	Polycystic Kidney Disease. Annual Review of Medicine, 2009, 60, 321-337.	12.2	697
4	The gene mutated in autosomal recessive polycystic kidney disease encodes a large, receptor-like protein. Nature Genetics, 2002, 30, 259-269.	21.4	683
5	Volume Progression in Polycystic Kidney Disease. New England Journal of Medicine, 2006, 354, 2122-2130.	27.0	670
6	Inhibition of renal cystic disease development and progression by a vasopressin V2 receptor antagonist. Nature Medicine, 2003, 9, 1323-1326.	30.7	597
7	Autosomal dominant polycystic kidney disease: the last 3 years. Kidney International, 2009, 76, 149-168.	5.2	491
8	Deletion of the TSC2 and PKD1 genes associated with severe infantile polycystic kidney disease â€" a contiguous gene syndrome. Nature Genetics, 1994, 8, 328-332.	21.4	466
9	Imaging Classification of Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2015, 26, 160-172.	6.1	439
10	Effective treatment of an orthologous model of autosomal dominant polycystic kidney disease. Nature Medicine, 2004, 10, 363-364.	30.7	438
11	Polycystic kidney disease. Nature Reviews Disease Primers, 2018, 4, 50.	30.5	435
12	Blood Pressure in Early Autosomal Dominant Polycystic Kidney Disease. New England Journal of Medicine, 2014, 371, 2255-2266.	27.0	392
13	Comprehensive Molecular Diagnostics in Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2007, 18, 2143-2160.	6.1	372
14	Mutations in GANAB , Encoding the Glucosidase IIÎ± Subunit, Cause Autosomal-Dominant Polycystic Kidney and Liver Disease. American Journal of Human Genetics, 2016, 98, 1193-1207.	6.2	345
15	Renal Cystic Disease in Tuberous Sclerosis: Role of the Polycystic Kidney Disease 1 Gene. American Journal of Human Genetics, 1997, 61, 843-851.	6.2	331
16	Functional polycystin-1 dosage governs autosomal dominant polycystic kidney disease severity. Journal of Clinical Investigation, 2012, 122, 4257-4273.	8.2	321
17	Kidney Volume and Functional Outcomes in Autosomal Dominant Polycystic Kidney Disease. Clinical Journal of the American Society of Nephrology: CJASN, 2012, 7, 479-486.	4.5	305
18	A truncated human chromosome 16 associated with Î± thalassaemia is stabilized by addition of telomeric repeat (TTAGGG)n. Nature, 1990, 346, 868-871.	27.8	300

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19	Characterization of PKD Protein-Positive Exosome-Like Vesicles. Journal of the American Society of Nephrology: JASN, 2009, 20, 278-288.	6.1	300
20	Effectiveness of Vasopressin V2 Receptor Antagonists OPC-31260 and OPC-41061 on Polycystic Kidney Disease Development in the PCK Rat. Journal of the American Society of Nephrology: JASN, 2005, 16, 846-851.	6.1	292
21	Cellular and subcellular localization of the ARPKD protein; fibrocystin is expressed on primary cilia. Human Molecular Genetics, 2003, 12, 2703-2710.	2.9	287
22	The transmembrane protein meckelin (MKS3) is mutated in Meckel-Gruber syndrome and the wpk rat. Nature Genetics, 2006, 38, 191-196.	21.4	266
23	Octreotide Inhibits Hepatic Cystogenesis in a Rodent Model of Polycystic Liver Disease by Reducing Cholangiocyte Adenosine 3â€²,5â€²-Cyclic Monophosphate. Gastroenterology, 2007, 132, 1104-1116.	1.3	261
24	Genetic mechanisms and signaling pathways in autosomal dominant polycystic kidney disease. Journal of Clinical Investigation, 2014, 124, 2315-2324.	8.2	261
25	Mechanisms of Disease: autosomal dominant and recessive polycystic kidney diseases. Nature Clinical Practice Nephrology, 2006, 2, 40-55.	2.0	255
26	Incompletely penetrant PKD1 alleles suggest a role for gene dosage in cyst initiation in polycystic kidney disease. Kidney International, 2009, 75, 848-855.	5.2	248
27	Vasopressin Directly Regulates Cyst Growth in Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2008, 19, 102-108.	6.1	240
28	Cyst Number but Not the Rate of Cystic Growth Is Associated with the Mutated Gene in Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2006, 17, 3013-3019.	6.1	230
29	Strategies Targeting cAMP Signaling in the Treatment of Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2014, 25, 18-32.	6.1	226
30	Genetic Complexity of Autosomal Dominant Polycystic Kidney and Liver Diseases. Journal of the American Society of Nephrology: JASN, 2018, 29, 13-23.	6.1	223
31	Angiotensin Blockade in Late Autosomal Dominant Polycystic Kidney Disease. New England Journal of Medicine, 2014, 371, 2267-2276.	27.0	221
32	Clinical and Molecular Characterization Defines a Broadened Spectrum of Autosomal Recessive Polycystic Kidney Disease (ARPKD). Medicine (United States), 2006, 85, 1-21.	1.0	215
33	Monoallelic Mutations to DNAJB11 Cause Atypical Autosomal-Dominant Polycystic Kidney Disease. American Journal of Human Genetics, 2018, 102, 832-844.	6.2	208
34	Molecular pathogenesis of ADPKD: The polycystin complex gets complex. Kidney International, 2005, 67, 1234-1247.	5.2	202
35	Association of mutation position in polycystic kidney disease 1 (PKD1) gene and development of a vascular phenotype. Lancet, The, 2003, 361, 2196-2201.	13.7	198
36	Mutation Analysis of the Entire PKD1 Gene: Genetic and Diagnostic Implications. American Journal of Human Genetics, 2001, 68, 46-63.	6.2	196

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37	The Position of the Polycystic Kidney Disease 1 (PKD1) Gene Mutation Correlates with the Severity of Renal Disease. Journal of the American Society of Nephrology: JASN, 2002, 13, 1230-1237.	6.1	195
38	Defects in cholangiocyte fibrocystin expression and ciliary structure in the PCK rat1 1The authors thank Dr. Torra for supplying ARPKD tissue.. Gastroenterology, 2003, 125, 1303-1310.	1.3	194
39	Phenotype-Genotype Correlations and Estimated Carrier Frequencies of Primary Hyperoxaluria. Journal of the American Society of Nephrology: JASN, 2015, 26, 2559-2570.	6.1	185
40	Genotypeâ€“Phenotype Correlations in Autosomal Dominant and Autosomal Recessive Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2007, 18, 1374-1380.	6.1	181
41	Identification, Characterization, and Localization of a Novel Kidney Polycystin-1-Polycystin-2 Complex. Journal of Biological Chemistry, 2002, 277, 20763-20773.	3.4	178
42	Coordinate Expression of the Autosomal Dominant Polycystic Kidney Disease Proteins, Polycystin-2 And Polycystin-1, in Normal and Cystic Tissue. American Journal of Pathology, 1999, 154, 1721-1729.	3.8	174
43	Prevalence Estimates of Polycystic Kidney and Liver Disease by Population Sequencing. Journal of the American Society of Nephrology: JASN, 2018, 29, 2593-2600.	6.1	173
44	Stable length polymorphism of up to 260 kb at the tip of the short arm of human chromosome 16. Cell, 1991, 64, 595-606.	28.9	169
45	Loss of Polycystin-1 in Human Cyst-Lining Epithelia Leads to Ciliary Dysfunction. Journal of the American Society of Nephrology: JASN, 2006, 17, 1015-1025.	6.1	169
46	A Practical Guide for Treatment of Rapidly Progressive ADPKD with Tolvaptan. Journal of the American Society of Nephrology: JASN, 2018, 29, 2458-2470.	6.1	163
47	Pkd2 haploinsufficiency alters intracellular calcium regulation in vascular smooth muscle cells. Human Molecular Genetics, 2003, 12, 1875-1880.	2.9	156
48	Late onset of renal and hepatic cysts in Pkd1-targeted heterozygotes. Nature Genetics, 1999, 21, 160-161.	21.4	149
49	A complete mutation screen of the ADPKD genes by DHPLC. Kidney International, 2002, 61, 1588-1599.	5.2	149
50	Identification of Gene Mutations in Autosomal Dominant Polycystic Kidney Disease through Targeted Resequencing. Journal of the American Society of Nephrology: JASN, 2012, 23, 915-933.	6.1	149
51	microRNA-17 family promotes polycystic kidney disease progression through modulation of mitochondrial metabolism. Nature Communications, 2017, 8, 14395.	12.8	147
52	Comparative analysis of the polycystic kidney disease 1 (PKD1) gene reveals an integral membrane glycoprotein with multiple evolutionary conserved domains. Human Molecular Genetics, 1997, 6, 1483-1489.	2.9	141
53	Genome Duplications and Other Features in 12 Mb of DNA Sequence from Human Chromosome 16p and 16q. Genomics, 1999, 60, 295-308.	2.9	140
54	A polycystin-centric view of cyst formation and disease: the polycystins revisited. Kidney International, 2015, 88, 699-710.	5.2	140

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55	Tuberin-Dependent Membrane Localization of Polycystin-1. Molecular Cell, 2001, 7, 823-832.	9.7	139
56	Food Restriction Ameliorates the Development of Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2016, 27, 1437-1447.	6.1	138
57	Predicted Mutation Strength of Nontruncating PKD1 Mutations Aids Genotype-Phenotype Correlations in Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2016, 27, 2872-2884.	6.1	136
58	Molecular diagnostics for autosomal dominant polycystic kidney disease. Nature Reviews Nephrology, 2010, 6, 197-206.	9.6	134
59	Incompletely Penetrant PKD1 Alleles Mimic the Renal Manifestations of ARPKD. Journal of the American Society of Nephrology: JASN, 2010, 21, 1097-1102.	6.1	126
60	Imaging-Based Diagnosis of Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2015, 26, 746-753.	6.1	126
61	Refining Genotype-Phenotype Correlation in Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2016, 27, 1861-1868.	6.1	123
62	Characterization of large rearrangements in autosomal dominant polycystic kidney disease and the PKD1/TSC2 contiguous gene syndrome. Kidney International, 2008, 74, 1468-1479.	5.2	120
63	Distinguishing between Hepatic Inflammation and Fibrosis with MR Elastography. Radiology, 2017, 284, 694-705.	7.3	117
64	Ciliary and centrosomal defects associated with mutation and depletion of the Meckel syndrome genes MKS1 and MKS3. Human Molecular Genetics, 2009, 18, 3311-3323.	2.9	115
65	A complete mutation screen of PKHD1 in autosomal-recessive polycystic kidney disease (ARPKD) pedigrees. Kidney International, 2003, 64, 391-403.	5.2	113
66	Performance of an Artificial Multi-observer Deep Neural Network for Fully Automated Segmentation of Polycystic Kidneys. Journal of Digital Imaging, 2017, 30, 442-448.	2.9	112
67	Transition fibre protein FBF1 is required for the ciliary entry of assembled intraflagellar transport complexes. Nature Communications, 2013, 4, 2750.	12.8	110
68	Polycystin-1 maturation requires polycystin-2 in a dose-dependent manner. Journal of Clinical Investigation, 2015, 125, 607-620.	8.2	107
69	Biliary Dysgenesis in the PCK Rat, an Orthologous Model of Autosomal Recessive Polycystic Kidney Disease. American Journal of Pathology, 2004, 165, 1719-1730.	3.8	105
70	Reduced Ciliary Polycystin-2 in Induced Pluripotent Stem Cells from Polycystic Kidney Disease Patients with PKD1 Mutations. Journal of the American Society of Nephrology: JASN, 2013, 24, 1571-1586.	6.1	104
71	Overweight and Obesity Are Predictors of Progression in Early Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2018, 29, 571-578.	6.1	101
72	Tolvaptan plus Pasireotide Shows Enhanced Efficacy in a PKD1 Model. Journal of the American Society of Nephrology: JASN, 2015, 26, 39-47.	6.1	99

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73	DCDC2 Mutations Cause a Renal-Hepatic Ciliopathy by Disrupting Wnt Signaling. American Journal of Human Genetics, 2015, 96, 81-92.	6.2	98
74	Vasopressin and disruption of calcium signalling in polycystic kidney disease. Nature Reviews Nephrology, 2015, 11, 451-464.	9.6	97
75	[Ca ²⁺] _i Reduction Increases Cellular Proliferation and Apoptosis in Vascular Smooth Muscle Cells. Circulation Research, 2005, 96, 873-880.	4.5	89
76	Polycystin-1 expression in PKD1, early-onset PKD1, and TSC2/PKD1 cystic tissue. Kidney International, 1999, 56, 1324-1333.	5.2	87
77	Pregnancy outcomes in autosomal dominant polycystic kidney disease: a case-control study. Journal of Maternal-Fetal and Neonatal Medicine, 2016, 29, 807-812.	1.5	87
78	International consensus statement on the diagnosis and management of autosomal dominant polycystic kidney disease in children and young people. Nature Reviews Nephrology, 2019, 15, 713-726.	9.6	86
79	Proteolytic Cleavage and Nuclear Translocation of Fibrocystin Is Regulated by Intracellular Ca2+ and Activation of Protein Kinase C. Journal of Biological Chemistry, 2006, 281, 34357-34364.	3.4	85
80	B9D1 is revealed as a novel Meckel syndrome (MKS) gene by targeted exon-enriched next-generation sequencing and deletion analysis. Human Molecular Genetics, 2011, 20, 2524-2534.	2.9	79
81	Baseline total kidney volume and the rate of kidney growth are associated with chronic kidney disease progression in Autosomal Dominant Polycystic Kidney Disease. Kidney International, 2018, 93, 691-699.	5.2	76
82	Molecular genetics of autosomal recessive polycystic kidney disease. Molecular Genetics and Metabolism, 2004, 81, 75-85.	1.1	75
83	Polycystic Kidney Disease without an Apparent Family History. Journal of the American Society of Nephrology: JASN, 2017, 28, 2768-2776.	6.1	75
84	Molecular diagnostics of Meckelâ€“Gruber syndrome highlights phenotypic differences between MKS1 and MKS3. Human Genetics, 2007, 121, 591-599.	3.8	74
85	Cyclic nucleotide signaling in polycystic kidney disease. Kidney International, 2010, 77, 129-140.	5.2	67
86	Epitope-Tagged Pkhd1 Tracks the Processing, Secretion, and Localization of Fibrocystin. Journal of the American Society of Nephrology: JASN, 2011, 22, 2266-2277.	6.1	67
87	Autophagy activators suppress cystogenesis in an autosomal dominant polycystic kidney disease model. Human Molecular Genetics, 2016, 26, ddw376.	2.9	67
88	Molecular basis of polycystic kidney disease: PKD1, PKD2 and PKHD1. Current Opinion in Nephrology and Hypertension, 2002, 11, 309-314.	2.0	64
89	Deficiency of polycystinâ€2 reduces Ca 2+ channel activity and cell proliferation in ADPKD lymphoblastoid cells. FASEB Journal, 2004, 18, 884-886.	0.5	63
90	Monoallelic IFT140 pathogenic variants are an important cause of the autosomal dominant polycystic kidney-spectrum phenotype. American Journal of Human Genetics, 2022, 109, 136-156.	6.2	62

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91	Long-Term Administration of Tolvaptan in Autosomal Dominant Polycystic Kidney Disease. Clinical Journal of the American Society of Nephrology: CJASN, 2018, 13, 1153-1161.	4.5	60
92	Genetic Variation of DKK3 May Modify Renal Disease Severity in ADPKD. Journal of the American Society of Nephrology: JASN, 2010, 21, 1510-1520.	6.1	59
93	Long-term trajectory of kidney function in autosomal-dominant polycystic kidney disease. Kidney International, 2019, 95, 1253-1261.	5.2	59
94	Pkd1 transgenic mice: adult model of polycystic kidney disease with extrarenal and renal phenotypes. Human Molecular Genetics, 2010, 19, 1174-1189.	2.9	58
95	The genetics of kidney stone disease and nephrocalcinosis. Nature Reviews Nephrology, 2022, 18, 224-240.	9.6	57
96	Determinants of Renal Disease Variability inÂADPKD. Advances in Chronic Kidney Disease, 2010, 17, 131-139.	1.4	55
97	PKHDL1, a homolog of the autosomal recessive polycystic kidney disease gene, encodes a receptor with inducible T lymphocyte expression. Human Molecular Genetics, 2003, 12, 685-698.	2.9	54
98	The Meckel syndrome protein meckelin (TMEM67) is a key regulator of cilia function but is not required for tissue planar polarity. Human Molecular Genetics, 2013, 22, 2024-2040.	2.9	54
99	Effect of genotype on the severity and volume progression of polycystic liver disease in autosomal dominant polycystic kidney disease. Nephrology Dialysis Transplantation, 2016, 31, 952-960.	0.7	54
100	Image texture features predict renal function decline in patients with autosomal dominantÂpolycystic kidney disease. Kidney International, 2017, 92, 1206-1216.	5.2	54
101	CD8+ T cells modulate autosomal dominant polycystic kidney disease progression. Kidney International, 2018, 94, 1127-1140.	5.2	54
102	2008 Homer W. Smith Award. Journal of the American Society of Nephrology: JASN, 2009, 20, 1188-1198.	6.1	51
103	PKD2 -Related Autosomal Dominant Polycystic Kidney Disease: Prevalence, Clinical Presentation, Mutation Spectrum, andÂPrognosis. American Journal of Kidney Diseases, 2017, 70, 476-485.	1.9	50
104	Primary results of the randomized trial of metformin administration in polycystic kidney disease (TAME PKD). Kidney International, 2021, 100, 684-696.	5.2	48
105	Effects of hydration in rats and mice with polycystic kidney disease. American Journal of Physiology - Renal Physiology, 2015, 308, F261-F266.	2.7	47
106	Functional characterization of PKDREJ, a male germ cell-restricted polycystin. Journal of Cellular Physiology, 2006, 209, 493-500.	4.1	46
107	What Is the Role of Somatic Mutation in Autosomal Dominant Polycystic Kidney Disease?. Journal of the American Society of Nephrology: JASN, 2010, 21, 1073-1076.	6.1	46
108	Spatiotemporal dynamics and heterogeneity of renal lymphatics in mammalian development and cystic kidney disease. ELife, 2019, 8, .	6.0	46

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109	Autosomal dominant polycystic kidney disease coexisting with cystic fibrosis. Journal of Nephrology, 2006, 19, 529-34.	2.0	46
110	PKD1 Duplicated regions limit clinical Utility of Whole Exome Sequencing for Genetic Diagnosis of Autosomal Dominant Polycystic Kidney Disease. Scientific Reports, 2019, 9, 4141.	3.3	44
111	Detection and characterization of mosaicism in autosomal dominant polycystic kidney disease. Kidney International, 2020, 97, 370-382.	5.2	44
112	Autosomal Dominant Polycystic Kidney Patients May Be Predisposed to Various Cardiomyopathies. Kidney International Reports, 2017, 2, 913-923.	0.8	42
113	Extracellular vesicles and exosomes generated from cystic renal epithelial cells promote cyst growth in autosomal dominant polycystic kidney disease. Nature Communications, 2021, 12, 4548.	12.8	42
114	Novel stop and frameshifting mutations in the autosomal dominant polycystic kidney disease 2 (PKD2) gene. Human Genetics, 1997, 101, 229-234.	3.8	41
115	Development of multiorgan pathology in the wpk rat model of polycystic kidney disease. The Anatomical Record, 2004, 277A, 384-395.	1.8	41
116	Haplotype analysis improves molecular diagnostics of autosomal recessive polycystic kidney disease. American Journal of Kidney Diseases, 2005, 45, 77-87.	1.9	41
117	The value of genotypic and imaging information to predict functional and structural outcomes in ADPKD. JCI Insight, 2020, 5, .	5.0	41
118	Synergistic Genetic Interactions between Pkhd1 and Pkd1 Result in an ARPKD-Like Phenotype in Murine Models. Journal of the American Society of Nephrology: JASN, 2019, 30, 2113-2127.	6.1	39
119	Epidemiology of Autosomal Dominant Polycystic Kidney Disease in Olmsted County. Clinical Journal of the American Society of Nephrology: CJASN, 2020, 15, 69-79.	4.5	39
120	Clinical spectrum, prognosis and estimated prevalence of DNAJB11-kidney disease. Kidney International, 2020, 98, 476-487.	5.2	38
121	The ENOS polymorphism is not associated with severity of renal disease in polycystic kidney disease 1. American Journal of Kidney Diseases, 2003, 41, 90-94.	1.9	37
122	Understanding pathogenic mechanisms in polycystic kidney disease provides clues for therapy. Current Opinion in Nephrology and Hypertension, 2006, 15, 456-463.	2.0	37
123	Evidence of a third ADPKD locus is not supported by re-analysis of designated PKD3 families. Kidney International, 2014, 85, 383-392.	5.2	37
124	Prognostic Enrichment Design in Clinical Trials for Autosomal Dominant Polycystic Kidney Disease: The TEMPO 3:4 Clinical Trial. Kidney International Reports, 2016, 1, 213-220.	0.8	37
125	Quantitative MRI of kidneys in renal disease. Abdominal Radiology, 2018, 43, 629-638.	2.1	37
126	Prognostic enrichment design in clinical trials for autosomal dominant polycystic kidney disease: the HALT-PKD clinical trial. Nephrology Dialysis Transplantation, 2017, 32, gfw294.	0.7	36

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127	Modulation of Polycystic Kidney Disease Severity by Phosphodiesterase 1 and 3 Subfamilies. Journal of the American Society of Nephrology: JASN, 2016, 27, 1312-1320.	6.1	36
128	Recent advances in the identification and management of inherited hyperoxalurias. Urolithiasis, 2019, 47, 79-89.	2.0	36
129	Identification of a subtle t(16;19)(p13.3;p13.3) in an infant with multiple congenital abnormalities using a 12-colour multiplex FISH telomere assay, M-TEL. European Journal of Human Genetics, 2000, 8, 903-910.	2.8	35
130	Utilizing magnetization transfer imaging to investigate tissue remodeling in a murine model of autosomal dominant polycystic kidney disease. Magnetic Resonance in Medicine, 2016, 75, 1466-1473.	3.0	35
131	Polycystin-1 regulates amphiregulin expression through CREB and AP1 signalling: implications in ADPKD cell proliferation. Journal of Molecular Medicine, 2012, 90, 1267-1282.	3.9	34
132	The Mutation, a Key Determinant of Phenotype in ADPKD. Journal of the American Society of Nephrology: JASN, 2013, 24, 868-870.	6.1	34
133	GTP-binding of ARL-3 is activated by ARL-13 as a GEF and stabilized by UNC-119. Scientific Reports, 2016, 6, 24534.	3.3	34
134	Presymptomatic Screening for Intracranial Aneurysms in Patients with Autosomal Dominant Polycystic Kidney Disease. Clinical Journal of the American Society of Nephrology: CJASN, 2019, 14, 1151-1160.	4.5	34
135	The genetic background significantly impacts the severity of kidney cystic disease in the Pkd1RC/RC mouse model of autosomal dominant polycystic kidney disease. Kidney International, 2021, 99, 1392-1407.	5.2	32
136	Can we further enrich autosomal dominant polycystic kidney disease clinical trials for rapidly progressive patients? Application of the PROPKD score in the TEMPO trial. Nephrology Dialysis Transplantation, 2018, 33, 645-652.	0.7	31
137	Progress in the understanding of polycystic kidney disease. Nature Reviews Nephrology, 2019, 15, 70-72.	9.6	31
138	Cross-talk between CDK4/6 and SMYD2 regulates gene transcription, tubulin methylation, and ciliogenesis. Science Advances, 2020, 6, .	10.3	31
139	Biallelic inheritance of hypomorphic PKD1 variants is highly prevalent in very early onset polycystic kidney disease. Genetics in Medicine, 2021, 23, 689-697.	2.4	31
140	Identification of a leader exon and a core promoter for the rat tuberous sclerosis 2 (Tsc2) gene and structural comparison with the human homolog. Mammalian Genome, 1997, 8, 554-558.	2.2	30
141	The Genetics of Vascular Complications in Autosomal Dominant Polycystic Kidney Disease (ADPKD). Current Hypertension Reviews, 2013, 9, 37-43.	0.9	30
142	Patterns of Kidney Function Decline in Autosomal Dominant Polycystic Kidney Disease: A Post Hoc Analysis From the HALT-PKD Trials. American Journal of Kidney Diseases, 2018, 71, 666-676.	1.9	30
143	Generation and phenotypic characterization of Pde1a mutant mice. PLoS ONE, 2017, 12, e0181087.	2.5	29
144	The Value of Genetic Testing in Polycystic Kidney Diseases Illustrated by a Family With PKD2 and COL4A1 Mutations. American Journal of Kidney Diseases, 2018, 72, 302-308.	1.9	29

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145	Transcriptome analysis reveals manifold mechanisms of cyst development in ADPKD. Human Genomics, 2016, 10, 37.	2.9	28
146	The PKD1 gene product. Nature Medicine, 1995, 1, 493-493.	30.7	27
147	MicroRNA501â€5p induces p53 proteasome degradation through the activation of the mTOR/MDM2 pathway in ADPKD cells. Journal of Cellular Physiology, 2018, 233, 6911-6924.	4.1	27
148	Parallel microarray profiling identifies ErbB4 as a determinant of cyst growth in ADPKD and a prognostic biomarker for disease progression. American Journal of Physiology - Renal Physiology, 2017, 312, F577-F588.	2.7	26
149	Oxidative Stress and Mitochondrial Abnormalities Contribute to Decreased Endothelial Nitric Oxide Synthase Expression and Renal Disease Progression in Early Experimental Polycystic Kidney Disease. International Journal of Molecular Sciences, 2020, 21, 1994.	4.1	26
150	The regulatory 1Î± subunit of protein kinase A modulates renal cystogenesis. American Journal of Physiology - Renal Physiology, 2017, 313, F677-F686.	2.7	25
151	Growth Pattern of Kidney Cyst Number and Volume in Autosomal Dominant Polycystic Kidney Disease. Clinical Journal of the American Society of Nephrology: CJASN, 2019, 14, 823-833.	4.5	25
152	Regulation of polycystin expression, maturation and trafficking. Cellular Signalling, 2020, 72, 109630.	3.6	25
153	A novel PKD1 variant demonstrates a disease-modifying role in trans with a truncating PKD1 mutation in patients with Autosomal Dominant Polycystic Kidney Disease. BMC Nephrology, 2015, 16, 26.	1.8	24
154	Pansomatostatin Agonist Pasireotide Long-Acting Release for Patients with Autosomal Dominant Polycystic Kidney or Liver Disease with Severe Liver Involvement. Clinical Journal of the American Society of Nephrology: CJASN, 2020, 15, 1267-1278.	4.5	24
155	Clinical characterization of primary hyperoxaluria type 3 in comparison with types 1 and 2. Nephrology Dialysis Transplantation, 2022, 37, 869-875.	0.7	23
156	Volume regression of native polycystic kidneys after renal transplantation. Nephrology Dialysis Transplantation, 2016, 31, 73-79.	0.7	22
157	Expanded Imaging Classification of Autosomal Dominant Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2020, 31, 1640-1651.	6.1	22
158	Phosphodiesterase 1A Modulates Cystogenesis in Zebrafish. Journal of the American Society of Nephrology: JASN, 2014, 25, 2222-2230.	6.1	21
159	Pyridoxine Responsiveness in a Type 1 Primary Hyperoxaluria Patient With a Rare (Atypical) AGXT Gene Mutation. Kidney International Reports, 2020, 5, 955-958.	0.8	20
160	Genomic diagnostics in polycystic kidney disease: an assessment of real-world use of whole-genome sequencing. European Journal of Human Genetics, 2021, 29, 760-770.	2.8	20
161	PKHDL1, a homolog of the autosomal recessive polycystic kidney disease gene, encodes a receptor with inducible T lymphocyte expression. Human Molecular Genetics, 2003, 12, 685-98.	2.9	20
162	Endothelial Dysfunction Occurs prior to Clinical Evidence of Polycystic Kidney Disease. American Journal of Nephrology, 2013, 38, 233-240.	3.1	19

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163	Characterization of three ciliopathy pedigrees expands the phenotype associated with biallelic C2CD3 variants. European Journal of Human Genetics, 2018, 26, 1797-1809.	2.8	19
164	Metalloproteinase PAPP-A regulation of IGF-1 contributes to polycystic kidney disease pathogenesis. JCI Insight, 2020, 5, .	5.0	19
165	Recurrence of the PKD1 nonsense mutation Q4041X in Spanish, Italian, and British families. Human Mutation, 1998, 11, S117-S120.	2.5	16
166	Insight into response to mTOR inhibition when PKD1 and TSC2 are mutated. BMC Medical Genetics, 2015, 16, 39.	2.1	15
167	Semantic Instance Segmentation of Kidney Cysts in MR Images: A Fully Automated 3D Approach Developed Through Active Learning. Journal of Digital Imaging, 2021, 34, 773-787.	2.9	15
168	Comprehensive Genetic Analysis Reveals Complexity of Monogenic Urinary Stone Disease. Kidney International Reports, 2021, 6, 2862-2884.	0.8	15
169	NF-ÎºB activation is required for apoptosis in fibrocystin/polyductin-depleted kidney epithelial cells. Apoptosis: an International Journal on Programmed Cell Death, 2010, 15, 94-104.	4.9	14
170	Strategy and rationale for urine collection protocols employed in the NEPTUNE study. BMC Nephrology, 2015, 16, 190.	1.8	14
171	Epidemiology of autosomal-dominant polycystic liver disease in Olmsted county. JHEP Reports, 2020, 2, 100166.	4.9	14
172	Establishing a nephrology genetic clinic. Kidney International, 2021, 100, 254-259.	5.2	14
173	The utility of a genetic kidney disease clinic employing a broad range of genomic testing platforms: experience of the Irish Kidney Gene Project. Journal of Nephrology, 2022, 35, 1655-1665.	2.0	14
174	Functional and transport analyses of CLCN5 genetic changes identified in Dent disease patients. Physiological Reports, 2016, 4, e12776.	1.7	13
175	Determinants of Progression in Early Autosomal Dominant Polycystic Kidney Disease: Is it Blood Pressure or Renin-Angiotensin-Aldosterone- System Blockade?. Current Hypertension Reviews, 2018, 14, 39-47.	0.9	13
176	Genomics Integration Into Nephrology Practice. Kidney Medicine, 2021, 3, 785-798.	2.0	13
177	RNA helicase p68 inhibits the transcription and post-transcription of Pkd1 in ADPKD. Theranostics, 2020, 10, 8281-8297.	10.0	12
178	Relationship between caffeine intake and autosomal dominant polycystic kidney disease progression: a retrospective analysis using the CRISP cohort. BMC Nephrology, 2018, 19, 378.	1.8	11
179	Population data improves variant interpretation in autosomal dominant polycystic kidney disease. Genetics in Medicine, 2019, 21, 1425-1434.	2.4	11
180	The genetic landscape of polycystic kidney disease in Ireland. European Journal of Human Genetics, 2021, 29, 827-838.	2.8	11

#	Article	IF	Citations
181	Pain and Obesity in Autosomal Dominant Polycystic Kidney Disease: A Post Hoc Analysis of the Halt Progression of Polycystic Kidney Disease (HALT-PKD) Studies. Kidney Medicine, 2021, 3, 536-545.e1.	2.0	11
182	Somatotroph pituitary adenoma with acromegaly and autosomal dominant polycystic kidney disease: SSTR5 polymorphism and PKD1 mutation. Pituitary, 2012, 15, 342-349.	2.9	10
183	Inherited renal cystic diseases. Abdominal Radiology, 2016, 41, 1035-1051.	2.1	10
184	mtor Haploinsufficiency Ameliorates Renal Cysts and Cilia Abnormality in Adult Zebrafish tmem67 Mutants. Journal of the American Society of Nephrology: JASN, 2021, 32, 822-836.	6.1	10
185	Characteristics of Patients with End-Stage Kidney Disease in ADPKD. Kidney International Reports, 2021, 6, 755-767.	0.8	10
186	Up-Regulation of DNA Damage Response Signaling in Autosomal Dominant Polycystic Kidney Disease. American Journal of Pathology, 2021, 191, 902-920.	3.8	10
187	Primary Hyperoxaluria Type 3 Can Also Result in Kidney Failure: A Case Report. American Journal of Kidney Diseases, 2022, 79, 125-128.	1.9	10
188	The role of DNA damage as a therapeutic target in autosomal dominant polycystic kidney disease. Expert Reviews in Molecular Medicine, 2019, 21, e6.	3.9	9
189	Prognostic Value of Fibroblast Growth Factor 23 in Autosomal Dominant Polycystic Kidney Disease. Kidney International Reports, 2021, 6, 953-961.	0.8	9
190	Digenic mutations of human OCRL paralogs in Dentâ€™s disease type 2 associated with Chiari I malformation. Human Genome Variation, 2016, 3, 16042.	0.7	8
191	The time for next-generation molecular genetic diagnostics in nephrology is now!. Kidney International, 2018, 94, 237-239.	5.2	8
192	Clâ¯ and H+ coupling properties and subcellular localizations of wildtype and disease-associated variants of the voltage-gated Clâ¯/H+ exchanger ClC-5. Journal of Biological Chemistry, 2020, 295, 1464-1473.	3.4	8
193	Ciliopathy protein HYLS1 coordinates the biogenesis and signaling of primary cilia by activating the ciliary lipid kinase PIPKIÎ³. Science Advances, 2021, 7, .	10.3	8
194	High Prevalence of Kidney Cysts in Patients With CYP24A1 Deficiency. Kidney International Reports, 2021, 6, 1895-1903.	0.8	8
195	Genotype Phenotype Correlation in Dent Disease 2 and Review of the Literature: OCRL Gene Pleiotropism or Extreme Phenotypic Variability of Lowe Syndrome?. Genes, 2021, 12, 1597.	2.4	8
196	Kidney Cysts in Hypophosphatemic Rickets With Hypercalciuria: A Case Series. Kidney Medicine, 2022, 4, 100419.	2.0	8
197	Tuberous sclerosis complex and polycystic kidney disease together: An exception to the contiguous gene syndrome. Genetics in Medicine, 2006, 8, 197-198.	2.4	7
198	B-type natriuretic peptide overexpression ameliorates hepatorenal fibrocystic disease inÂaÂratÂmodel of polycystic kidney disease. Kidney International, 2017, 92, 657-668.	5.2	7

#	Article	IF	Citations
199	Disrupting Polycystin-2 EF hand Ca2+ affinity does not alter channel function or contribute to polycystic kidney disease. Journal of Cell Science, 2020, 133, .	2.0	7
200	Congenital Heart Disease in Adults with Autosomal Dominant Polycystic Kidney Disease. American Journal of Nephrology, 2022, 53, 316-324.	3.1	7
201	A potentially crucial role of the PKD1 C-terminal tail in renal prognosis. Clinical and Experimental Nephrology, 2018, 22, 395-404.	1.6	6
202	Pancreatic Cysts and Intraductal Papillary Mucinous Neoplasm in Autosomal Dominant Polycystic Kidney Disease. Pancreas, 2019, 48, 698-705.	1.1	6
203	Characterization of Primary Cilia in Osteoblasts Isolated From Patients <scp>With ADPKD</scp> and <scp>CKD</scp>. JBMR Plus, 2021, 5, e10464.	2.7	6
204	Volume Progression and Imaging Classification of Polycystic Liver in Early Autosomal Dominant Polycystic Kidney Disease. Clinical Journal of the American Society of Nephrology: CJASN, 2022, 17, 374-384.	4.5	6
205	ADPKD Progression in Patients With No Apparent Family History and No Mutation Detected by Sanger Sequencing. American Journal of Kidney Diseases, 2018, 71, 294-296.	1.9	5
206	The Underestimated Burden of Monogenic Diseases in Adult-Onset ESRD. Journal of the American Society of Nephrology: JASN, 2018, 29, 1583-1584.	6.1	5
207	Protein Kinase A Downregulation Delays the Development and Progression of Polycystic Kidney Disease. Journal of the American Society of Nephrology: JASN, 2022, 33, 1087-1104.	6.1	5
208	Differential Expression of Renal Proteins in a Rodent Model of Meckel Syndrome. Nephron Experimental Nephrology, 2011, 117, e31-e38.	2.2	4
209	Bacterial Cholangitis in Autosomal Dominant Polycystic Kidney and Liver Disease. Mayo Clinic Proceedings Innovations, Quality & Outcomes, 2019, 3, 149-159.	2.4	4
210	Multiple unilateral subcapsular cortical hemorrhagic cystic disease of the kidney: CT and MRI findings and clinical characteristic. European Radiology, 2019, 29, 4843-4850.	4.5	4
211	Interactions between FGF23 and Genotype in Autosomal Dominant Polycystic Kidney Disease. Kidney360, 2020, 1, 648-656.	2.1	4
212	Functional megalin is expressed in renal cysts in a mouse model of adult polycystic kidney disease. CKJ: Clinical Kidney Journal, 2021, 14, 2420-2427.	2.9	4
213	Autosomal Dominant Polycystic Kidney Disease. , 2010, , 529-542.		3
214	Impaired Hedgehog-Gli1 Pathway Activity Underlies the Vascular Phenotype of Polycystic Kidney Disease. Hypertension, 2020, 76, 1889-1897.	2.7	3
215	Autosomal dominant polycystic kidney disease: A genetic perspective. Nephrology, 1997, 3, 387-395.	1.6	2
216	Genetic Etiologies, Diagnosis, and Management of Neonatal Cystic Kidney Disease. NeoReviews, 2022, 23, e175-e188.	0.8	2

#	Article	IF	Citations
217	Asymptomatic Pyuria as a Prognostic Biomarker in Autosomal Dominant Polycystic Kidney Disease. Kidney360, 2022, 3, 465-476.	2.1	2
218	Cardiovascular Outcomes in Kidney Transplant Recipients With ADPKD. Kidney International Reports, 2022, 7, 1991-2005.	0.8	2
219	Molecular analysis of a consanguineous Iranian polycystic kidney disease family identifies a PKD2mutation that aids diagnostics. BMC Nephrology, 2013, 14, 190.	1.8	1
220	Closeout of the HALT-PKD trials. Contemporary Clinical Trials, 2015, 44, 48-55.	1.8	1
221	SP003GENETIC TESTING IN SUSPECTED HEREDITARY PROTEINURIC KIDNEY DISEASES. Nephrology Dialysis Transplantation, 2018, 33, i346-i347.	0.7	1
222	Classical Polycystic Kidney Disease: Gene Structures and Mutations and Protein Structures and Functions. , 2018, , 3-26.		1
223	CYP24A1 deficiency causing persistent hypercalciuria in a stone former. Journal of Nephrology, 2021, 34, 949-951.	2.0	1
224	PKD1 Compared With PKD2 Genotype and Cardiac Hospitalizations in the Halt Progression of Polycystic Kidney Disease Studies. Kidney International Reports, 2022, 7, 117-120.	0.8	1
225	Genetic studies: a key to understanding pathogenesis in PKD. Nephrology News & Issues, 2006, 20, 20-2.	0.1	1
226	FC044: Heterozygous Variants in Kinase Domain of NEK8 cause an Autosomal-Dominant Ciliopathy. Nephrology Dialysis Transplantation, 2022, 37, .	0.7	1
227	Disease Stage Characterization of Hepatorenal Fibrocystic Pathology in the PCK Rat Model of ARPKD. Anatomical Record, 2010, 293, spc1-spc1.	1.4	0
228	Bariatric surgery in a patient with cystinuria. Clinical Nephrology Case Studies, 2021, 9, 54-58.	0.7	0
229	Cystâ€lining epithelial cells from ADPKD kidneys have a mechanoâ€ciliary dysfunction. FASEB Journal, 2006, 20, A339.	0.5	0
230	Netrin-1 overexpression induces polycystic kidney disease - a novel mechanism contributing cystogenesis in ADPKD.. American Journal of Pathology, 2022, , .	3.8	0