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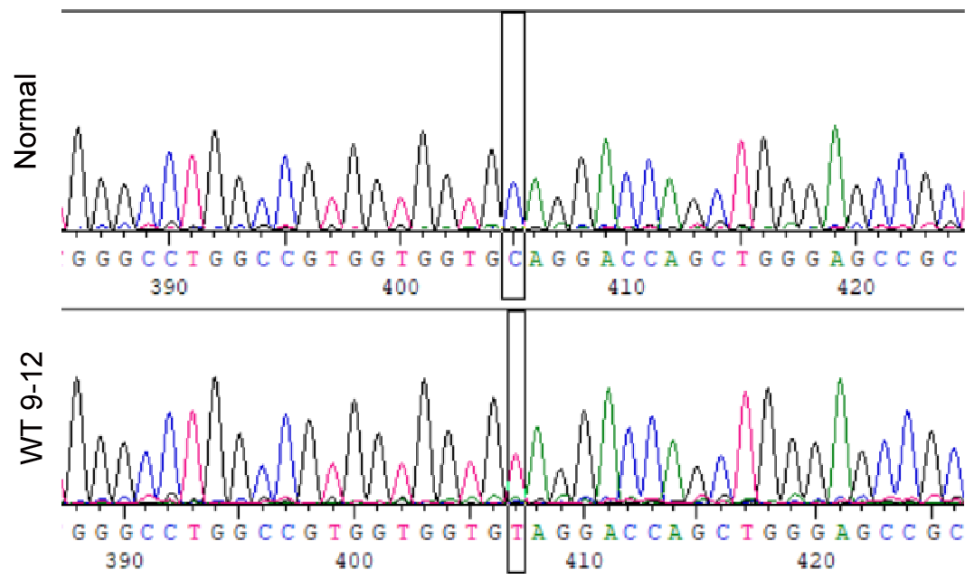
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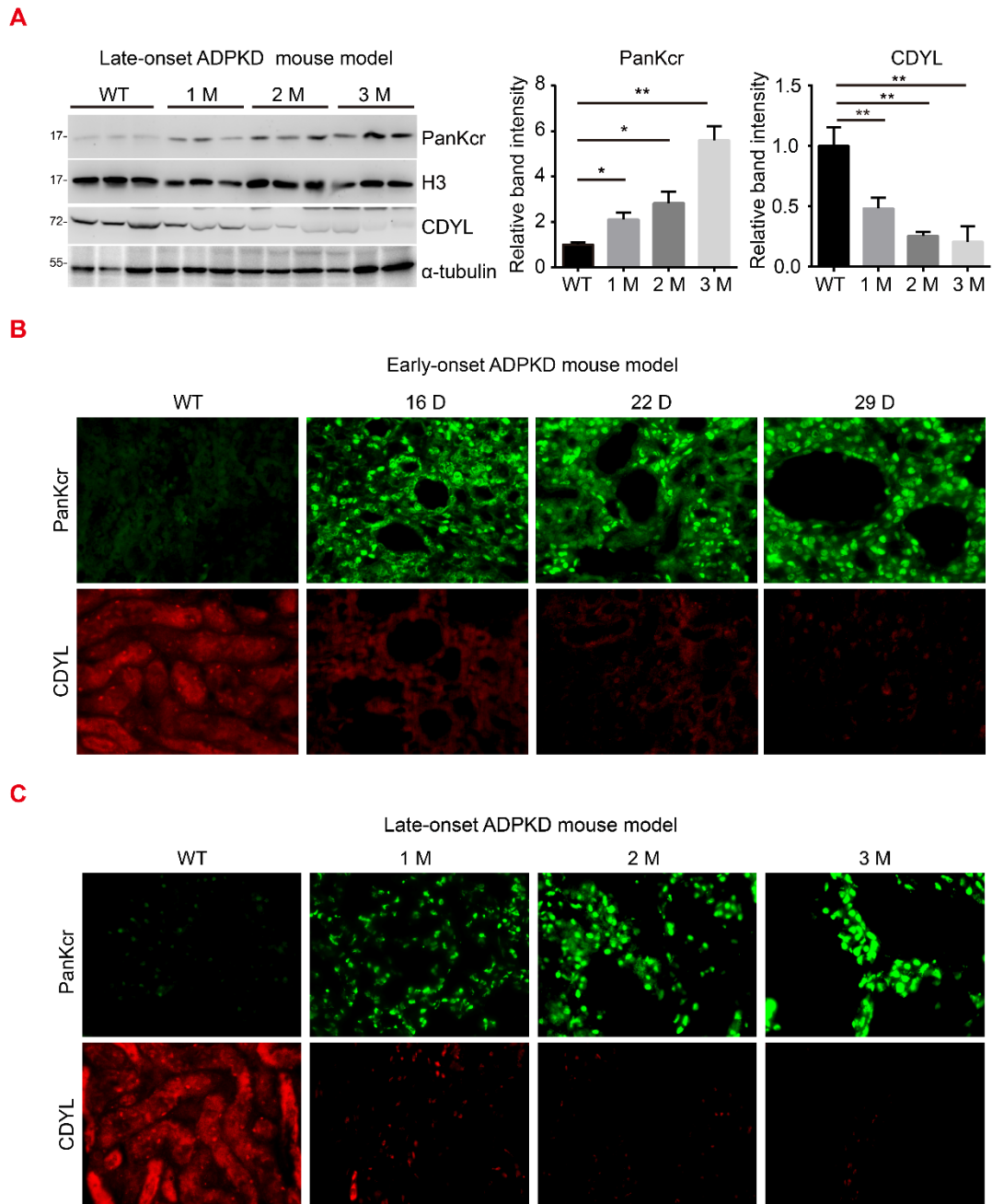
Supplemental Table 1. Primers for RT-PCR.

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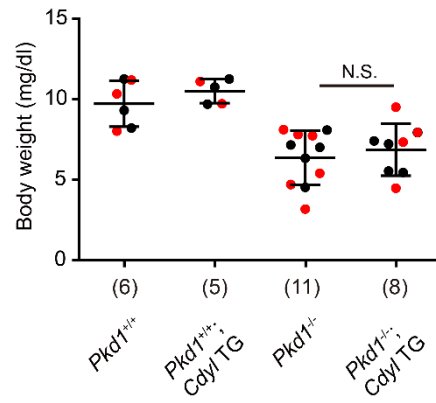
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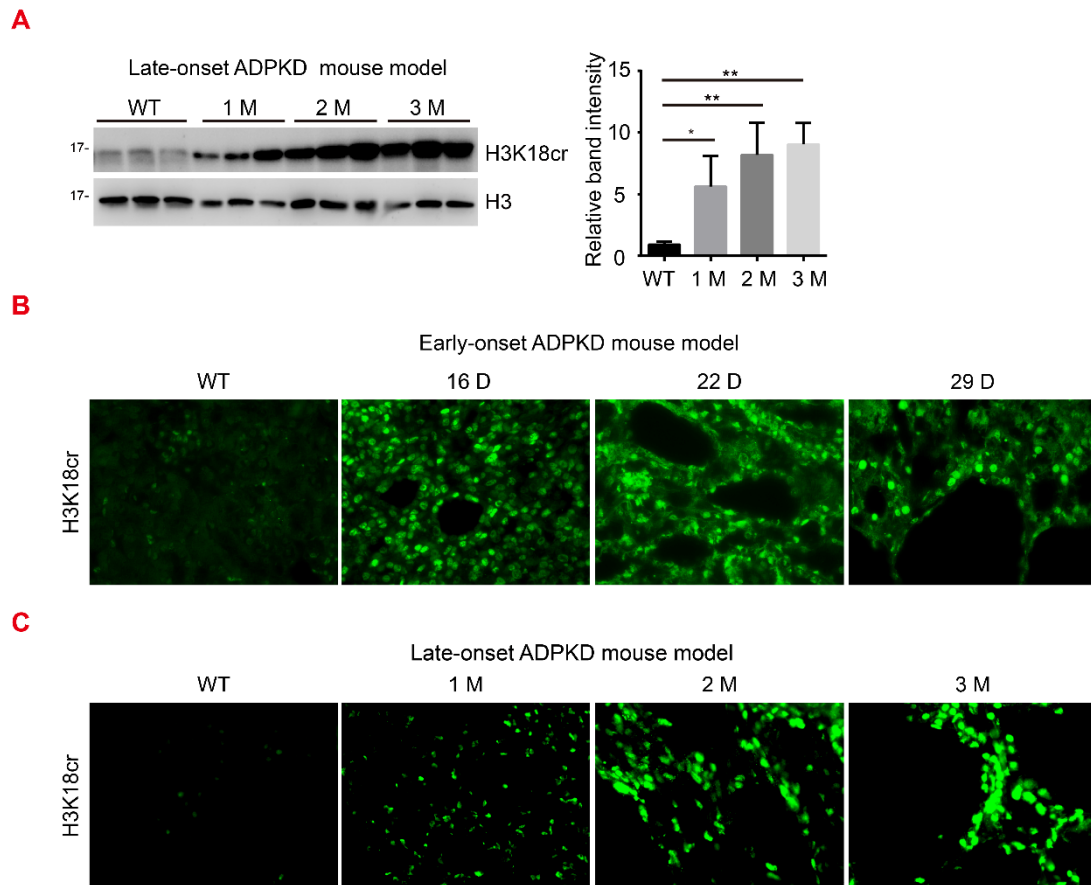
Supplemental Figure 1. Genetic analysis reveals homozygous mutation of *PKD1* in WT 9-12 cells.



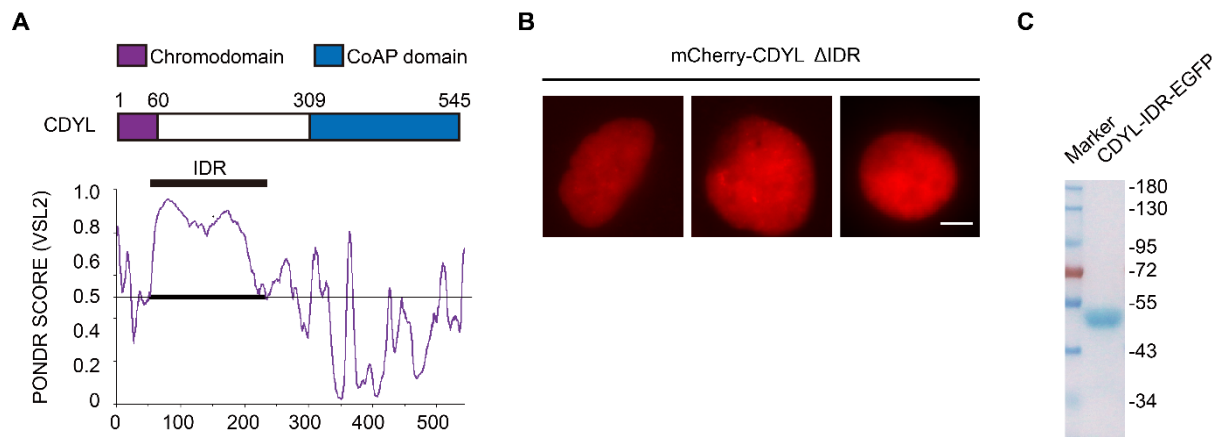
Supplemental Figure 2. Downregulation of CDYL is accompanied by upregulation of histone Kcr in early-onset and late-onset ADPKD mouse model. (A) Western blotting analysis of CDYL and PanKcr in kidneys from WT and late-onset ADPKD mouse model at the indicated times. Data were analyzed by two-tailed unpaired Student's *t*-test and are represented as means \pm SEM. * $P < 0.001$, ** $P < 0.0001$. (B and C) Immunofluorescence imaging of PanKcr and CDYL in mouse kidneys from early-onset (B) or late-onset (C) ADPKD mouse model at the indicated times.



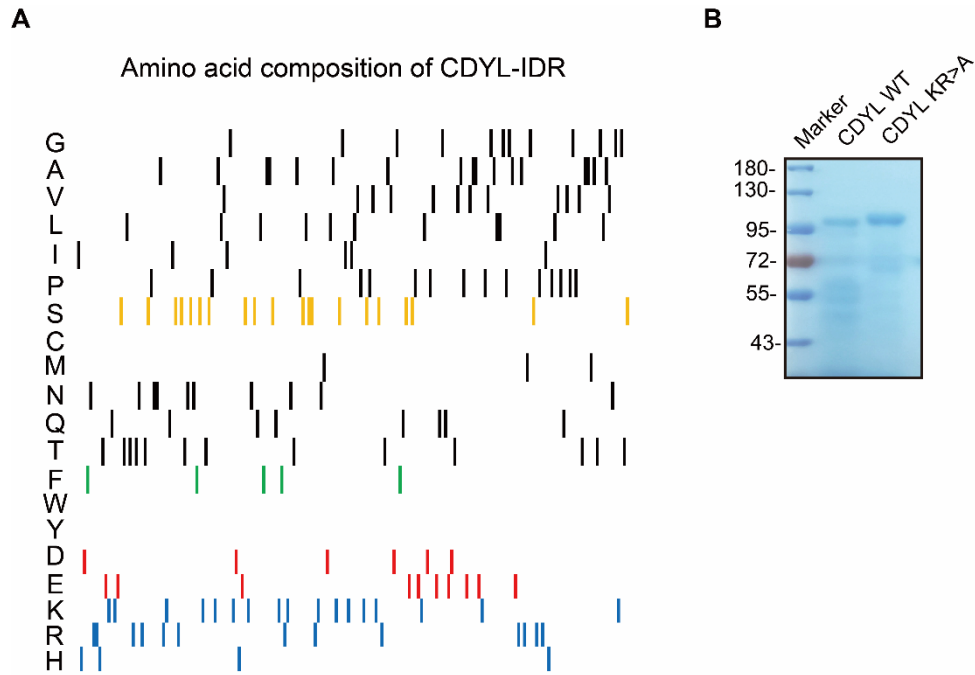
Supplemental Figure 3. Body weight of WT and ADPKD mice in the indicated groups. Red dots represented males and black dots represented females. N.S.: not significant.



Supplemental Figure 4. Upregulation of H3K18cr in early-onset and late-onset ADPKD mouse model. (A) Western blotting analysis of H3K18cr in kidneys from WT and late-onset ADPKD mouse model at the indicated times. Data were analyzed by two-tailed unpaired Student's *t*-test and are represented as means \pm SEM. * $P < 0.001$, ** $P < 0.0001$. (B and C) Immunofluorescence imaging of H3K18cr in mouse kidneys from early-onset (B) or late-onset (C) ADPKD mouse model at the indicated times.

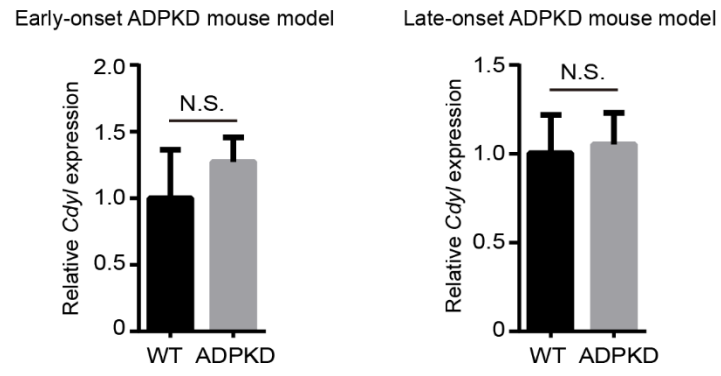


Supplemental Figure 5. Intrinsic disorder of CDYL. (A) For PONDOR prediction, a VSL2 score above 0.5 reflects a high degree of disorder. The black bar designates the IDR under investigation. (B) Immunofluorescence imaging of mCherry-CDYL Δ IDR in WT 9-12 cells. Scale bar, 5 μ m. (C) Purified CDYL-IDR-EGFP fusion proteins stained with Coomassie Brilliant Blue.



Supplemental Figure 6. CDYL WT and KR>A mutant fusion proteins. **(A)** Amino acid composition of the CDYL-IDR protein. Each row represents a single amino acid. Black bars represent the occurrence of the indicated amino acid at that position in CDYL-IDR. **(B)** Purified CDYL WT and KR>A mutant fusion proteins stained with Coomassie Brilliant Blue.

A



Supplemental Figure 7. *Cdy1* mRNA in ADPKD mice. N.S.: not significant.

Supplemental Table 1. Primers for RT-PCR

Gene name	Forward	Reverse
Mus. <i>Stat5a</i>	CACTCCTGTACTTGGTTCGTCA	CCAGGTCAAACCTCGCCATCT
Mus. <i>Col5a1</i>	GACTTCCCAGATGGCGAATA	CCAAGAAGTGATTCTGGCTCC
Mus. <i>Tlr4</i>	TCCCTGCATAGAGGTAGTTCC	TCAAGGGGTTGAAGCTCAGA
Mus. <i>Igf2</i>	CGGCTTCTACTTCAGCAGGC	TATCTGGGGAAGTCGTCCG
Mus. <i>Actin</i>	TTGCTGACAGGATGCAGAAG	ATCCACATCTGCTGGAAGGT
Homo. <i>STAT5A</i>	GAAGATCAAGCTGGGGCACT	CGGACCAGCCTCTGTTCATT
Homo. <i>COL5A1</i>	CTTCCCTGACAAGAAGTCCGAA	CCACATAGGAGAGCAGTTTCC
Homo. <i>TLR4</i>	CTGCGTGGAGGTGGTTCC	AGCTCAGGTCCAGGTTCTTG
Homo. <i>SEMA5A</i>	GAACCTGCCACCCACC	TCTCACACACCAACACAGGT
Homo. <i>GAPDH</i>	TTGCCCTCAACGACCACTTT	TGGTCCAGGGGTCTTACTCC

Supplemental Table 2. Characteristics of ADPKD patients

Age (yr)	sex	Family history	Hepatic cyst	AST (U/L)	ALT (U/L)	eGFR (mL/min/1.73m ²)	BUN (mg/dL)	SCr (mg/dL)	CKD stages
49	Male	Y	Y	-	-	52	9.9	137.7	III
55	Female	Y	Y	22	18	107	7.4	47.4	I
21	Male	Y	N	14	9	96	5.5	96.94	I
63	Female	Y	N	21	12	35	14	138.8	III
35	Male	Y	N	21	26	94	6.4	92.66	I
47	Female	N	N	13	10	48	6.1	116.28	III
60	Male	N	N	16	20	80	4.6	89.3	II
71	Male	N	Y	27	29	71	7.2	92.81	II
59	Female	N	N	18	15	57	11.7	94.09	III
45	Male	N	N	19	12	64	9.5	118	II
55	Female	N	Y	30	31	46	7.3	126.31	III
51	Female	N	Y	19	12	77	6.6	77.3	II
52	Male	N	N	33	66	65	9.8	111.2	II
32	Female	Y	Y	16	8	116	5.1	60.5	I
72	Male	N	N	22	15	68	7.5	96.6	II
23	Male	N	N	29	69	131	5.9	70.1	I
52	Male	N	N	24	25	60	7.2	118.45	II
45	Female	N	Y	19	9	86	6.5	73	II
61	Male	N	Y	24	17	49	10.7	132.2	III
42	Male	Y	Y	17	17	76	5.1	104	II
49	Male	N	Y	21	32	132	8	63.3	I
65	Male	N	N	21	11	84	5.2	83.6	II
49	Female	Y	Y	13	5	13	23.1	344.7	V
48	Male	Y	Y	23	24	63	7.6	117.3	II
51	Female	Y	Y	17	12	26	11	206.8	IV

Y: yes, N: no.

Supplemental Table 3. Characteristics of ADPKD mice

Phenotype	sex	Body weight (g)	Kidney weight (g)	KW/BW (%)	cystic index (%)	BUN (mg/dL)	SCr (mg/dL)
<i>Pkd1^{+/+}</i>	Female	9.31	0.125	1.34		17.25	0.16
	Female	8.21	0.124	1.51		19.72	0.16
	Male	8.02	0.117	1.46		22.90	0.33
	Male	10.33	0.1398	1.35		17.13	0.66
	Female	11.26	0.166	1.47		17.92	0.33
	Male	11.19	0.1538	1.37		19.34	0.33
<i>Pkd1^{+/+}; Cdy1 TG</i>	Male	9.72	0.124	1.28		19.18	0.33
	Male	11.1	0.1422	1.28		18.88	0.16
	Female	10.75	0.166	1.54		23.53	0.33
	Female	11.26	0.114	1.01		21.48	0.16
	Female	9.7	0.1555	1.60		21.82	0.49
<i>Pkd1^{-/-}</i>	Female	4.52	0.412	9.12	36.91	53.55	1.81
	Male	4.7	0.303	6.45	43.69	56.28	1.97
	Male	8.1	0.7584	9.36	64.43	67.20	0.99
	Female	8.08	0.63	7.80	48.87	61.38	1.97
	Female	6.33	0.63	9.95	48.81	53.93	2.30
	Female	7.16	0.678	9.47	55.82	73.95	1.48
	Male	7.8	0.6558	8.41	32.17	62.35	0.82
	Male	5.39	0.499	9.26	44.57	76.45	1.15
	<i>Pkd1^{-/-}; Cdy1 TG</i>	Male	7.93	0.54	6.81	37.76	57.74
Female		5.45	0.311	5.71	32.58	50.87	0.82
Female		5.53	0.36	6.51	16.05	44.97	0.49
Male		9.51	0.3703	3.89	23.96	40.32	0.33
Male		7.73	0.5001	6.47	41.86	60.38	2.14
Female		7.01	0.7023	10.02	47.59	56.95	1.32
Female		7.22	0.5504	7.62	27.92	48.15	0.33
Male		4.465	0.1628	3.65	23.84	52.85	0.16
Male		3.165	0.2291	7.24	35.80	78.80	2.47
Female		7.41	0.2196	2.96	30.76	21.15	0.49
Male		7.33	0.2307	3.15	32.31	36.08	0.66