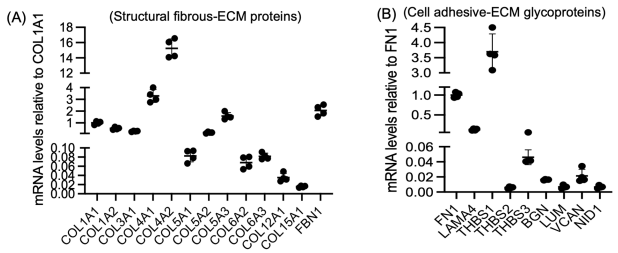
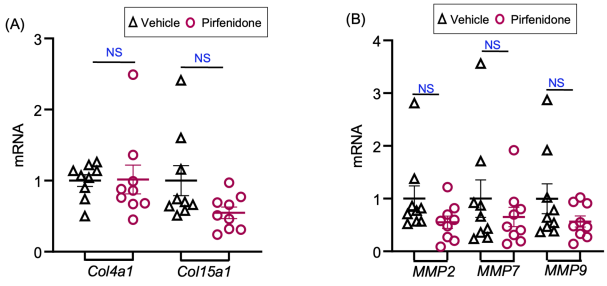
Supplemental Figures

Supplemental Fig. 1



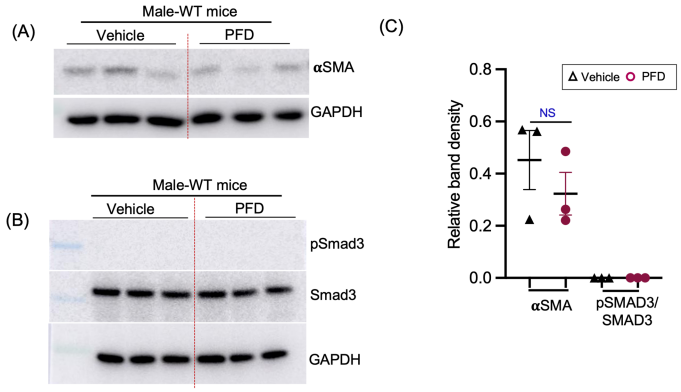
Supplemental Figure 1: (**A**) In primary culture human ADPKD myofibroblasts, relative gene expression of structural fibrous ECM proteins, (**B**) cell adhesive ECM-glycoproteins. Each individual data point represents a biological replicate (cells from individual human ADPKD kidneys).

Supplemental Fig. 2



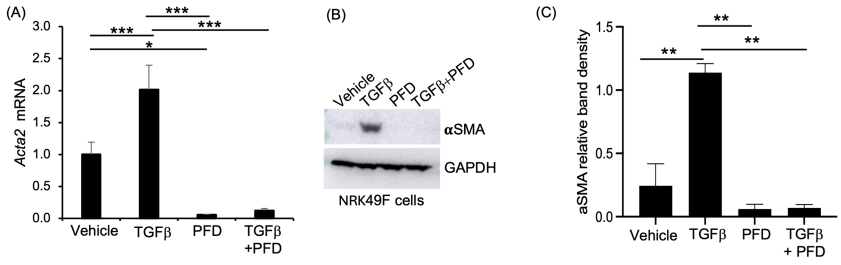
Supplemental Figure 2: Supplemental Figure 2: mRNA levels relative to 18S for (**A**) Col4a1 and Col15a1, and (**B**) MMPs in RC/RC mouse renal tissues.Each each individual datapoint represents a biological replicate derived from an independent mouse kidney.

Supplemental Fig. 3

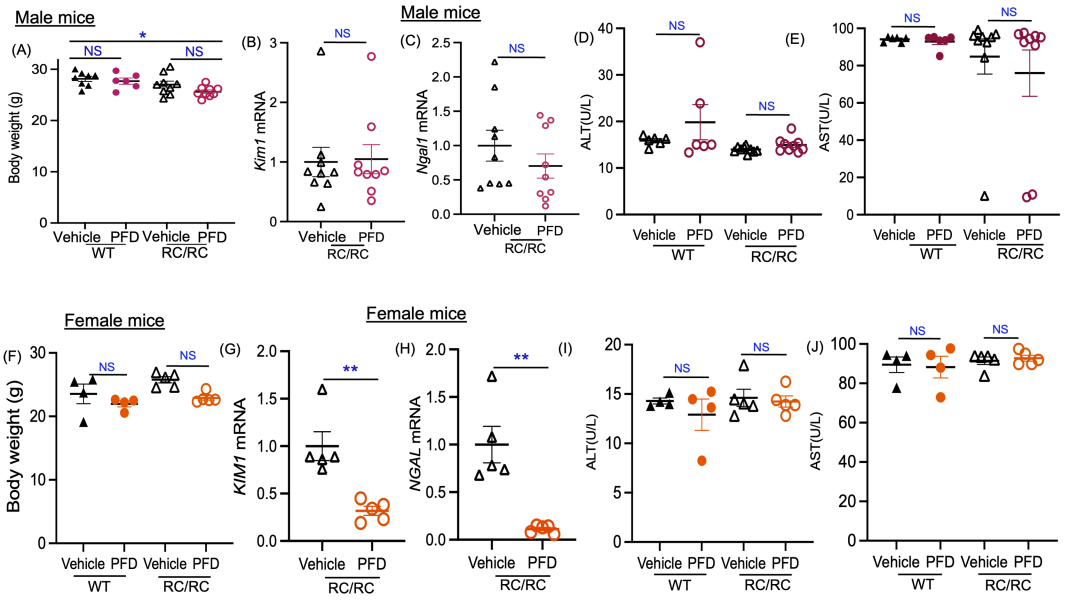


Supplemental Figure 3: Western blot analysis of whole kidney tissue lysate from WT male mice for, (**A**) αSMA (**B**) pSMAD3/SMAD3 and (**C**) Quantification of band density.Each individual datapoint represents a biological replicate derived from an independent mouse kidney.

Supplemental Fig. 4

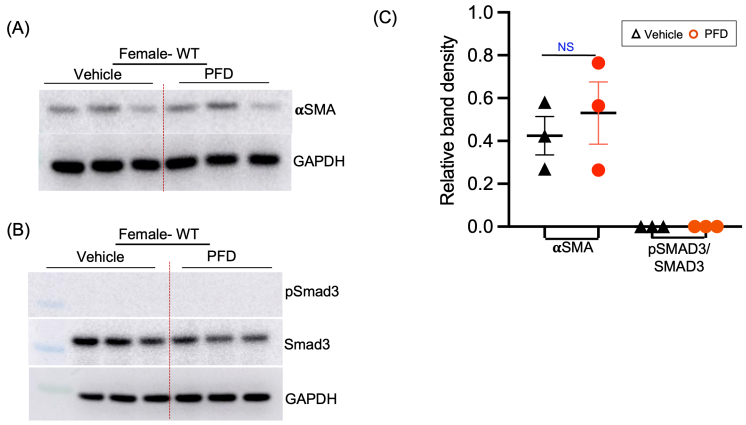


Supplemental Figure 4: (**A**) mRNA levels relative to 18S for αSMA in NRK-49F cells treated with TGFβ and pirfenidone for 24h. (**B**) Western blot analysis of NRK-49F cells treated with TGFβ (2ng/ml) and pirfenidone (0.5mg/ml) for 24h.(**C**) quantification of band density. *P<0.05, **P<0.01, ***P<0.001 by two-tailed unpaired t-test with Welch's correction.



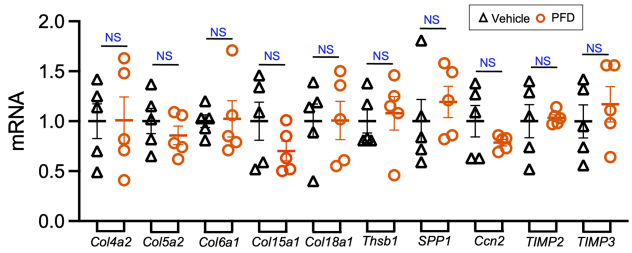
Supplemental Figure 5: (**A**) Body weight of male mice. (**B**) mRNA levels relative to 18S for *Kim1* and (**C**) *Ngal* in male mouse kidey tissue. (**D**) Serum ALT (U/L) in male mice (E) Serum AST (U/L) in male mice.(**F**) Body weight of female mice. (**G**) mRNA levels relative to 18S for *Kim1* and (**H**) *Ngal* in female mouse kidey tissue. (**I**) Serum ALT (U/L) in female (J) Serum AST (U/L) in female mice.Each individual datapoint represents a biological replicate derived from an independent mouse kidney.

Supplemental Fig. 6

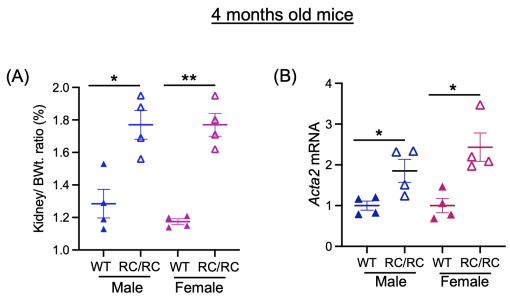


Supplemental Figure 6: Western blot analysis of whole kidney tissue lysate from WT female mice for, (**A**) αSMA (**B**) pSMAD3/SMAD3 and (**C**) Quantification of band density.Each individual datapoint represents a biological replicate derived from an independent mouse kidney.

Supplemental Fig. 7



Supplemental Figure 7: mRNA levels relative to 18S for Col4a2, Col5a2, Col6a1, Col15a, Col18a1, Thsb1, SPP1,SPP1,Ccn2,TIMP2 and TIMP3 in female RC/RC mouse renal tissues.Each each individual datapoint represents a biological replicate derived from an independent mouse kidney.



Supplemental Figure 8: Male and female RC/RC mice at 4 months of age. (**A**) Kidney to body weight ratio, (**B**) mRNA levels relative to 18S for *αSMA*. P<0.05 by two-tailed unpaired t-test with Welch's correction. Each individual datapoint represents a biological replicate derived from an independent mouse kidney.

Supplemental Table-1

Human primer sequence used for QRT-PCR

	Gene Name	Primer Forward (F) and Reverse (R)
1.	*MMP-2*	F-AGCGAGTGGATGCCGCCTTTAA R-CATTCCAGGCATCTGCGATGAG
2.	*MMP-7*	F-TCGGAGGAGATGCTCACTTCGA R- GGATCAGAGGAATGTCCCATACC
3.	*LAMA-4*	F-GAGATGACTCTCTGCTGGACCT R-AGTTCCAGGCAGCCAACAAAGC
4.	*TIMP-1*	F-GGAGAGTGTCTGCGGATACTTC R-GCAGGTAGTGATGTGCAAGAGTC
5.	*TIMP-2*	F-ACCCTCTGTGACTTCATCGTGC R-GGAGATGTAGCACGGGATCATG
6.	*TIMP-3*	F-TACCGAGGCTTCACCAAGATGC R-CATCTTGCCATCATAGACGCGAC
8.	*BGN*	F-TTGAACCTGGAGCCTTCGATGG R-TTGGAGTAGCGAAGCAGGTCCT
9.	*LUM*	F-AACATACCAACTGTCAATGAAAACC R-TGCCATCCAAACGCAAATGCTTG
10.	*FBN1*	F-GGATACACAGGTGATGGCTTCAC R-GTCGCATTCACAGCGGTATCCT
11.	*VCAN*	F-TTGGACCTCAGGCGCTTTCTAC R-GGATGACCAATTACACTCAAATCAC
12.	*NID1*	F-ACATTGAGCCCTACACGGAGCT R-GCCACTGGTAAGTGTAGATGCG
13.	*ADAM12*	F-ATGGCATCTGCCAGACTCACGA R-GGAACTCTTCGAGACTTTGCCAC
14.	*ADAM17*	F-AACAGCGACTGCACGTTGAAGG R-CTGTGCAGTAGGACACGCCTTT
15.	*ADAM19*	F-CGAGAAGGTGAATGTGGCAGGA R-AGCTCTGACACTGGATCTTCCC
16.	*THBS1*	F-GCTGGAAATGTGGTGCTTGTCC R-CTCCATTGTGGTTGAAGCAGGC
17.	*THBS2*	F-CAGTCTGAGCAAGTGTGACACC R-TTGCAGAGACGGATGCGTGTGA
18.	*THBS3*	F-GGAAGGAGATGCCTGTGACAAC R-GGTAGGATTGCTCATTTCAGGGC
19.	*SPARC*	F-TGCCTGATGAGACAGAGGTGGT R-CTTCGGTTTCCTCTGCACCATC
20.	*FBLN1*	F-GTGGCTACCATCTCAACGAGGA

		R-CTTGCATTCGCAGCGGAAACTG
21.	*FBLN5*	F-CTGCTGGATGACAACCGAAGCT R-GATAAGGCTCCTCACAGCGGAT
22.	*CCN1*	F-GGAAAAGGCAGCTCACTGAAGC R-GGAGATACCAGTTCCACAGGTC
23.	*CCN2*	F-CTTGCGAAGCTGACCTGGAAGA R-CCGTCGGTACATACTCCACAGA
24.	*COL1a1*	F-TGACGTGATCTGTGACGAGAC R-GGTTTCTTGGTCGGTGGGT
25.	*COL1a2*	F-CCTGGTGCTAAAGGAGAAAGAGG R-ATCACCACGACTTCCAGCAGGA
26.	*COL3a1*	F- TGG TCT GCA AGG AAT GCC TGGA R- TCT TTC CCT GGG ACA CCA TCAG
27.	*COL4a1*	F-TGTTGACGGCTTACCTGGAGAC R-GGTAGACCAACTCCAGGCTCTC
28.	*COL4a2*	F-GGATAACAGGCGTGACTGGAGT R-CTTTGCCACCAGGCAGTCCAAT
29.	*COL5a1*	F- GGAGATGATGGTCCCAAAGGCA R- CCATCATCTCCTTTGTCACCAGG
30.	*COL5a2*	F- CAGGCTCCATAGGAATCAGAGG R- CCAGCATTTCCTGCTTCTCCAG
31.	*COL5a3*	F-GAGAGGAGAACTGGGCTTCCAA R-TAGAGGTCCCACTTCTCCTGTC
32.	*COL6a2*	F-CGTGGAGACTCAGGACAGCCA R-CCTTTCAAGCCAAAGTCGCCTC
33.	*COL6a3*	F-CCTGGTGTAACTGATGCTGCCA R-AAGATGGCGTCCACCTTGGACT
34.	*COL12a1*	F-CAGTGCCTGTAGTCAGCCTGAA R-GGTCTTGTTGGCTCTGTGTCCT
35.	*COL15a1*	F- GGTGACACTGGTTTACCTGGCT R-GCCTTTCCAGAGGAATGTCCTC
36.	*POSTN*	F- TGCCCAGCAGTTTTGCCCAT R- CGTTGCTCTCCAAACCTCTA
37.	*SERPINE1*	F- CTCATCAGCCACTGGAAAGGCA R- GACTCGTGAAGTCAGCCTGAAAC
38.	*FBLN1*	F- GTGGCTACCATCTCAACGAGGA R- CTTGCATTCGCAGCGGAAACTG
39.	*FBLN5*	F- CTGCTGGATGACAACCGAAGCT R- GATAAGGCTCCTCACAGCGGAT
40.	*18s*	F-ACCGCGGTTCTATTTTGTTG R- CCCTCTTAATCATGGCCTCA

Mouse primer sequence used for QRT-PCR

	Gene Name	Primer Forward (F) and Reverse (R)
1.	*Adam12*	F-TGCTACAACGGCATCTGCCAGA R-GCTCTTGGAGTCTTTGCCACAG
2.	*Adam17*	F-TGTGAGCGGTGACCACGAGAAT R-TTCATCCACCCTGGAGTTGCCA
3.	*Adam19*	F-GTGCCTCACTTACCAGGAACAG R-GGACTGCACTTCCTGTATTGGC
4.	*Thbs1*	F-GGTAGCTGGAAATGTGGTGCGT R-GCACCGATGTTCTCCGTTGTGA
5.	*Thbs2*	F-GTATGGAGGGAAGGACTGTGTC R-ACTTGGCTCCAGGAAAACACGG
6.	*SPARC*	F-CACCTGGACTACATCGGACCAT R-CTGCTTCTCAGTGAGGAGGTTG
7.	*Lama4*	F-CAGTTTGTCCTCTACCTCGGAAG R-CTCACAGGCTTGGAATCCAGGA
8.	*Bgn*	F-TGAACCAGGAGCCTTTGATGGC R-GTCCTCCAACTCAATAGCCTGG
9.	*Vcan*	F-GGACCAAGTTCCACCCTGACAT R-CTTCACTGCAAGGTTCCTCTTCT
10.	*Col1a1*	F - AGACATGTTCAGCTTTGTGGAC R - GCA GCT GAC TTC AGG GATG
11.	*Col1a2*	F-TTCTGTGGGTCCTGCTGGGAAA R-TTGTCACCTCGGATGCCTTGAG
12.	*Col3a1*	F - TCC CCT GGA ATC TGT GAA TC R - TGA GTC GAA TTG GGG AGA AT
13.	*Col4a1*	F-CGGGTGTGAAAAGACCTATCGG R-CTGGCATTCCTCTGACGCCTTT
14.	*Col4a2*	F-CGGGTGTGAAAAGACCTATCGG R-CTGGCATTCCTCTGACGCCTTT
15.	*Col5a1*	F-AGATGGCATCCGAGGTCTGAAG R-GACCTTCAGGACCATCTTCTCC
16.	*Col5a3*	F-GGCAAAGATGGTATTCCAGGACC R-TGCTTCCTTTGTGACCAGGCATC
17.	*Col5a2*	F-GTGGCATAGGAGAGAAAGGTGC R-GCCAACTAAGCCTCTAGGACCA
18.	*Col6a1*	F-GACACCTCTCAGTGTGCTCTGT R-GCGATAAGCCTTGGCAGGAAATG
19.	*Col6a3*	F-CCTGGTGTAACTGATGCTGCCA R-AAGATGGCGTCCACCTTGGACT
20.	*Col8a1*	F-GGAAATCCCACCTGTGCCAAGA R-TCCTCTTGGTCCAGGTTCTCCA
21.	*Col12a1*	F-CAGCACCATGAATGTCGTCTGG R-GGTCTTTGAGGATAGTGCTGGC
22.	*Col15a1*	F-ACACCCACAGTGACTCCCAAGA R-TCCTCATTGCCCACGATGTCTC
23.	*Col18a1*	F-GTGACACTGGACCTCAAGGCTT R-TTGTCTGAAGGAGGGTCCTGGT
24.	*CCN1*	F-GTGAAGTGCGTCCTTGTGGACA R-CTTGACACTGGAGCATCCTGCA
25.	*CCN2*	F - GTGCCAGAACGCACACTG R - CCCCGGTTACACTCCAAA
26.	*aSMA*	F - TCAGGGAGTAATGGTTGGAATG

		R - GGTGATGATGCCGTGTTCTA
27.	*18s*	F - GTAACCCGTTGAACCCGA R - CCATCCAATCGGTAGTAGCG
28.	*Timp1*	F - CAAGGATGGACTCCTGGCACAT R - TACTCGCCATCAGCGTTCCCAT
29.	*Timp2*	F - AGCCAAAGCAGTGAGCGAGAAG R - GCCGTGTAGATAAACTCGATGTC
30.	*Timp3*	F- AGGATGCCTTCTGCAACTCCGA R- GTGTAGACCAGAGTGCCAAAGG
31.	*Pai-1*	F - GAGGTGGAAAGAGCCAGATTTA R- CCACTGAAGTAGAGGGCATTC
32.	*Mmp2*	F - CAAGGATGGACTCCTGGCACAT R - TACTCGCCATCAColGCGTTCCCAT
33.	*Mmp7*	F - AGGTGTGGAGTGCCAGATGTTG R - CCACTACGATCCGAGGTAAGTC
34.	*Mmp9*	F - GCTGACTACGATAAGGACGGCA R - TAGTGGTGCAGGCAGAGTAGGA
35.	*Hai1*	F- CTTCGTGAGGAAGAGTGCATGC R-TCACACTCCAGGAAGCCATCGA
36.	*Areg*	F CAGAAGAATGGAAGAGTCAG R CAGATATGCAGGGAGTCACC
37.	*Opn*	F TGAGAGCAATGAGCATTCCGATG R CAGGGAGTTTCCATGAAGCCAC
38.	*He4*	F - AGGTCAAGTCTCCACGAAGCCA R - AGAACACTGGCTGTCCACCTGA
39.	*Ngal*	F - ATGTCACCTCCATCCTGGTCAG R - GCCACTTGCACATTGTAGCTCTG
40.	*Kim1*	F-AAACCAGAGATTCCCACACG R-GTCGTGGGTCTTCCTGTAGC
41.	*Fn1*	F -ATGTGGACCCCTCCTGATAGT R -GCCCAGTGATTTCAGCAAAGG
42.	*Icam1*	F -CTTCCAGCTACCATCCCAAA R -CTTCAGAGGCAGGAAACAGG
43.	*IL-6*	F- CTTCCATCCAGTTGCCTTCT R- CTCCGACTTGTGAAGTGGTATAG
44.	*IL-10*	F-TTTGAATTCCCTGGGTGAGAA R-ACAGGGGAGAAATCGATGACA
45.	*Tnf*	F-ACCCTCACACTCAGATCATCTTC R-TGGTGGTTTGCTACGACGT
46.	*Ifn*	F -GGCCATCAGCAACAACATAAGCGT R-TGGGTTGTTGACCTCAAACTTGGC
47.	*Ccl2*	F -CTCGGACTGTGATGCCTTAAT R- TGGATCCACACCTTGCATTTA
48.	*Ccl3*	F-GAAGATTCCACGCCAATTCATC R-GATCTGCCGGTTTCTCTTAGTC
49.	*IL1b*	F -TTGACGGACCCCAAAAGAT R - GAAGCTGGATGCTCTCATCTG
50.	*Postn*	F-CAAAGCACACAGTTACCTTTCCAGGG R-GCAGGAAACCCACATTGCATGAGA