Supplementary figures

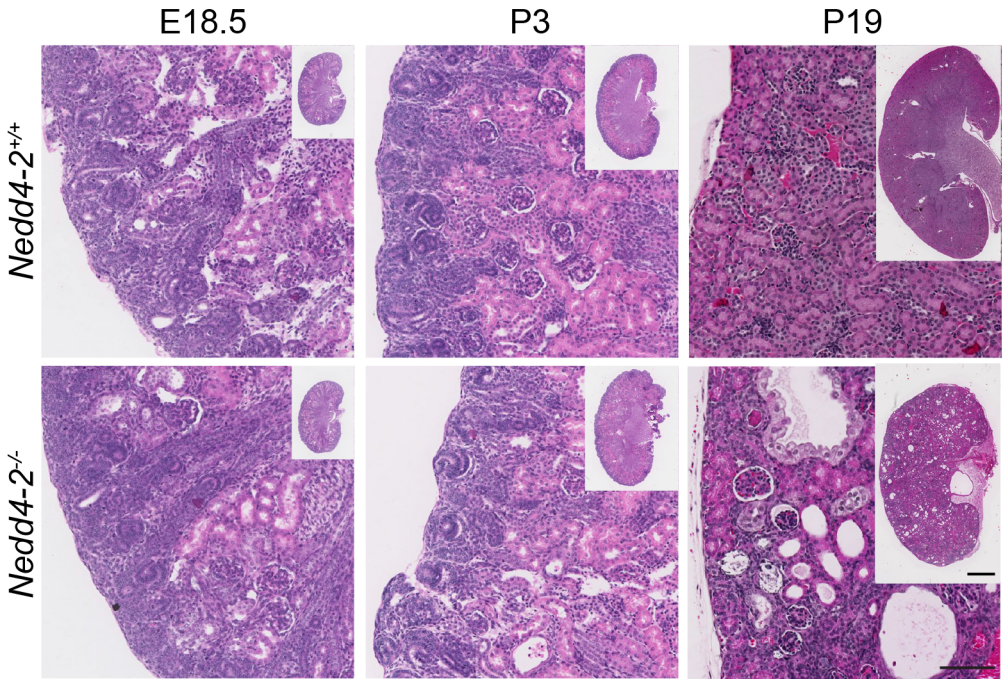

Figure S1. Progressive kidney disease in *Nedd4-2*$^{-/-}$ mice. H & E staining shows normal histology at E18.5 with signs of kidney damage first apparent at P3 in *Nedd4-2*$^{-/-}$ mice. The kidney pathology becomes more pronounced at P19 with dilated tubules and a disorganized structure. Scale bars: 100 μm (inset = 1 mm).

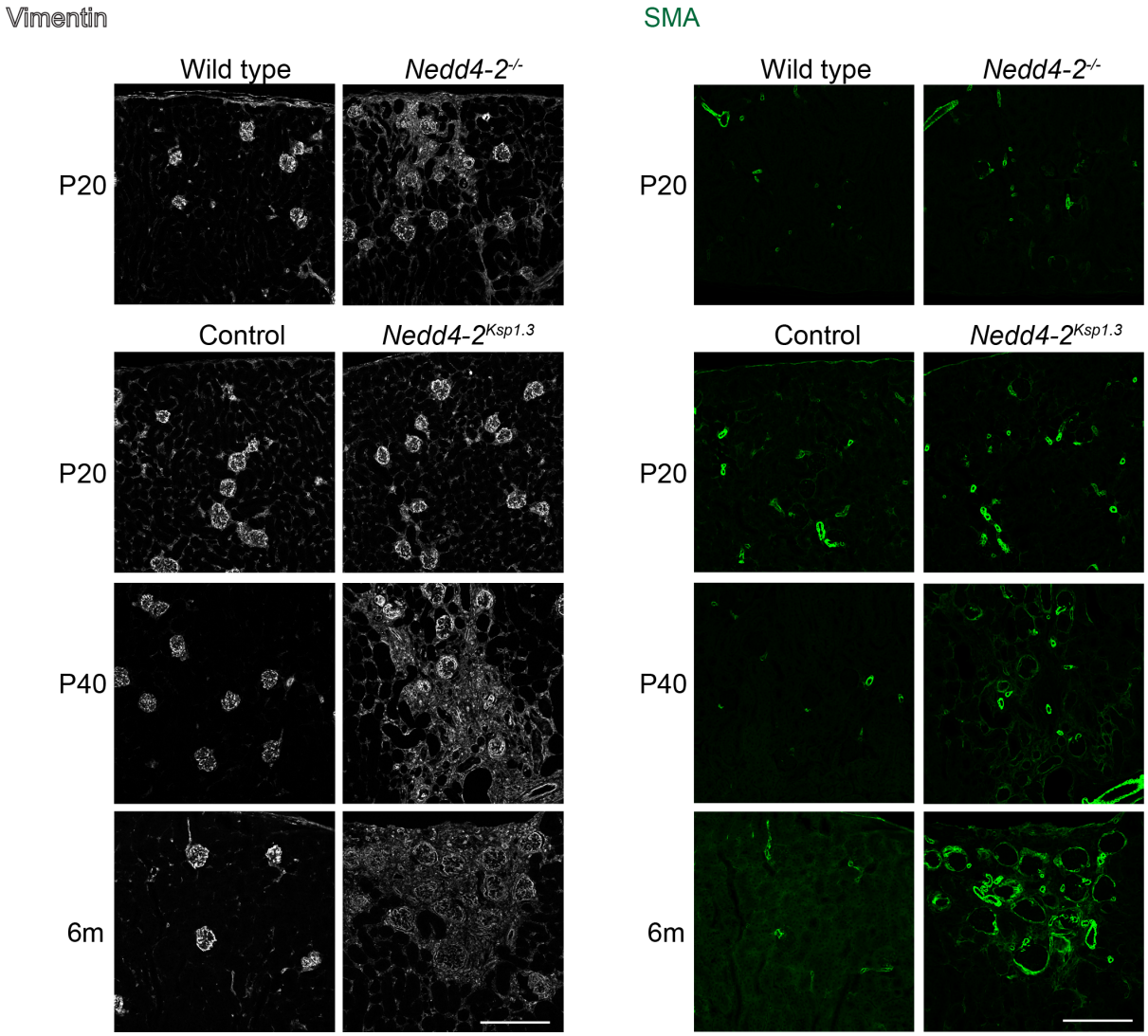

Figure S2. Disease progression in *Nedd4-2*$^{Ksp1.3}$ kidneys demonstrated by increasing expression of markers of kidney injury, vimentin and SMA. Immunohistochemistry of vimentin (white) and SMA (green) at P20 in *Nedd4-2*$^{-/-}$ kidneys are roughly equivalent to expression levels of these markers at P40 in *Nedd4-2*$^{Ksp1.3}$ kidneys. There is further increase in kidney damage at 6m of age. Scale bars: 100 μm.

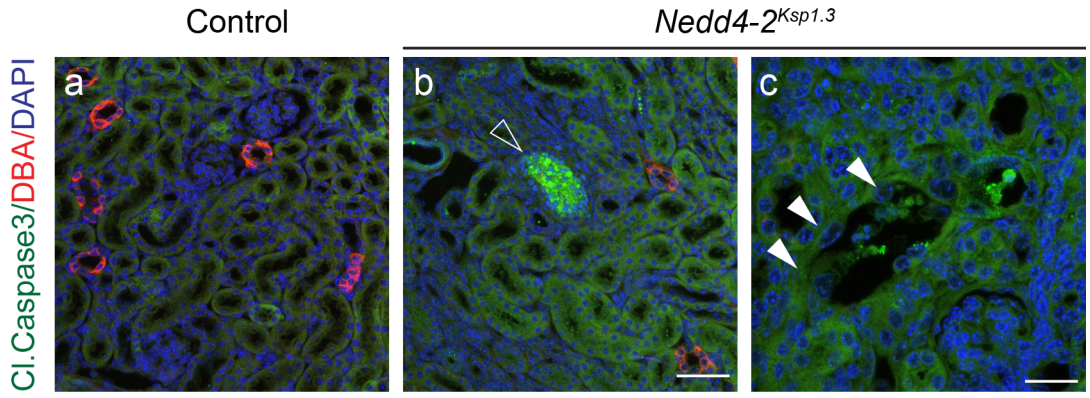

Figure S3. Increased cleaved caspase-3 staining in *Nedd4-2* KO mice at P20. (**a**) Very few positive cleaved caspase-3 cells (green) are seen in control kidneys at P20. In *Nedd4-2*$^{Ksp1.3}$ kidneys, cleaved caspase-3-positive cells can be seen within tubule lumens (**b**, empty arrow head) and in the cells lining the tubules (**c**). Scale bars: **a** and **b** = 50 μm, **c** = 25 μm. Arrowhead shows enlarged nuclei in tubule epithelium.

Supplementary Table 1. Primers used for qPCR. All primers were designed to amplify the mouse sequences.

Gene	**Primer sequence (5'-3')**
Collagen-1 (*Col1a1*)	F: CGGAGAAGAAGGAAAACGAGGAG
	R: CACCATCAGCACCAGGGAAAC
Vimentin (*Vim*)	F: CGGCTGCGAGAGAAATTGC
	R: CCACTTTCCGTTCAAGGTCAAG
α-smooth muscle actin (*αSMA*)	F: CCCAGACATCAGGGAGTAATGG
	R: TCTATCGGATACTTCAGCGTCA
Kidney injury molecule 1 (*KIM-1*)	F: TGGTTGCCTTCCGTGTCTCT
	R: TCAGCTCGGGAATGCACAA
TATA-box binding protein (*TBP*)	F: CAAACCCAGAATTGTTCTCCTT
	R: ATGTGGTCTTCCTGAATCCCT