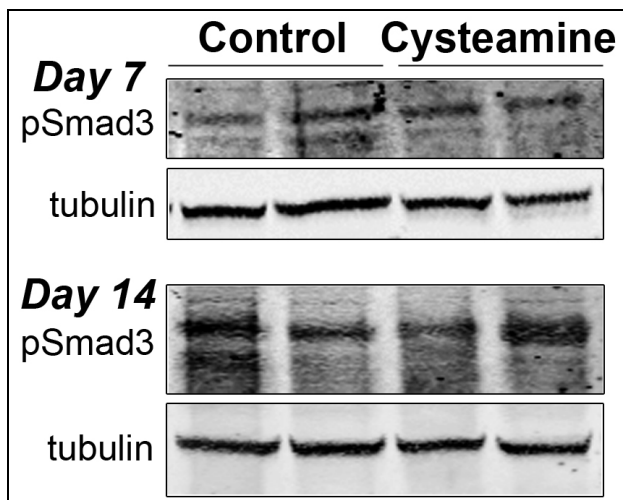
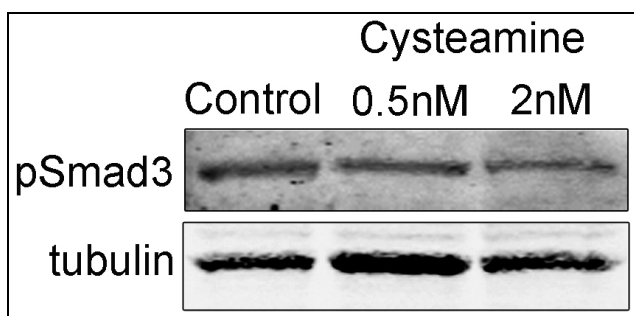


Supplemental Figure A1. Anti-fibrotic dose dependent effects of cysteamine with UUO. A pilot study was performed to determine the efficacy of three different doses of oral cysteamine 200, 400, and 600mg/kg. Mice were placed on cysteamine starting on the day of surgery and sacrificed 14 days after UUO (n=2-4/group).



Supplemental Figure A2. Cysteamine mediated attenuation of fibrosis is independent of TGF- β . Protein levels of phosphorylated Smad3 (pSmad3) measured by Western blotting and normalized to α -tubulin was performed on whole kidney homogenate in control and cysteamine treated mice (n=4/group).



Supplemental Figure A3. TGF- β activity is unaltered by cysteamine with myofibroblast activation. Normal rat kidney fibroblasts were transformed into α SMA+ myofibroblasts by exposure to transforming growth factor beta (TGF- β). Protein levels of phosphorylated Smad3 (pSmad3) measured by Western blotting and normalized to α -tubulin was performed on lysates from cells treated with cysteamine or vehicle alone (n=4/group).

SUPPLEMENTAL TABLES

Gene	Relative expression level (cysteamine:control)
<i>TNF-α</i>	1.31 \pm 0.34
<i>TNF-α receptor</i>	0.99 \pm 0.22
<i>IL-1β</i>	0.77 \pm 0.36
<i>IL-1β receptor</i>	0.91 \pm 0.30
<i>Nox4</i>	1.34 \pm 0.35
<i>Nox2</i>	0.80 \pm 0.21

Supplemental Table A1 No difference in pro-inflammatory and oxidative gene expression profiles after UUO with cysteamine treatment. Semi-quantitative real time RT qPCR was performed on total RNA from whole kidney homogenate from Day 14 UUO kidneys. Genes of interest were normalized to two housekeeping genes, 18S and GAPDH.

Gene	Relative expression level (cysteamine:control)		
	Day 3	Day 7	Day 14
<i>PDGF-Rα</i>	0.97 \pm 0.48	1.00 \pm 0.27	0.89 \pm 0.25
<i>PDGF-Rβ</i>	0.97 \pm 0.51	1.47 \pm 0.43	0.93 \pm 0.31

Supplemental Table A4 No difference in PDGF receptor gene expression profiles after UUO with cysteamine treatment. Semi-quantitative real time RT qPCR was performed on total RNA from whole kidney homogenate from control and cysteamine treated mice UUO kidneys. Genes of interest were normalized to two housekeeping genes, 18S and GAPDH.

Gene	Forward	Reverse
<i>TNF-α</i>	CATCTTCTCAAAATTCGAGTGACAA	TGGGAGTAGACAAGGTACAACCC
<i>TNF-α R</i>	GCTGACCTCTGCTCTACGAA	GCCATCCACCACAGCATACA
<i>IL-1β</i>	CAACCAACAAGTGATATTCTCCATG	GATCCACACTCTCCAGCTGCA
<i>IL-1β R</i>	CCTCGGGATGGAAATCTGCT	CCAGATTCTGAAGGCTTGCAT
<i>Nox2</i>	GGGAGACTGGACGGAGGGGC	ATGCGTGTCCCTGCACAGCC
<i>Nox4</i>	CACCAAACACAGAAGCACAAAG	AGAAAGCAAAGCAGGGTATCA
<i>PDGFR-α</i>	ATGGACGCACGCCAGACTGTG	TCTCGACGAAGCCTTTCTCGTGG
<i>PDGFR-β</i>	CACCTTCTCCAGTGTGCTGA	GGAGTCCATAGGGAGGAAGC
<i>Fibronectin</i>	AGACTGCAGTGACCACCATTC	AATGTGTCCTTGAGAGCATAGAC
<i>Procollagen 1</i>	AGAAGTCTCAAGATGGTGGCCG	GGTCACGAACCACGTTAGCATC
<i>Procollagen 3</i>	CAGCTATGGCCCTCCTGATCTT	GTAATGTTCTGGGAGGCCCG
<i>GAPDH</i>	ACTTTGTCAAGCTCATTTCC	TGCAGCGAACTTTATTGATG
<i>18S</i>	GGTGAAATTCTTGGACCGGC	GACTTTGGTTTCCCGGAAGC

Supplemental Table A3: qPCR primer sequences