Supplemental Fig. 1. Analysis of the UMOD mouse antibody in cell culture

UMOD immunofluorescence in IMCD3 cells. Staining of IMCD3 with antibodies directed against UMOD and acetylated tubulin results in staining of primary cilia. Blocking the staining by preincubating the antibodies with purified UMOD decreases the staining for UMOD significantly indicating the specifity of the antibody. Preincubation with UMOD does not affect the cilia staining with antibodies against acetylated tubulin. The scale bar represents 10 µm.

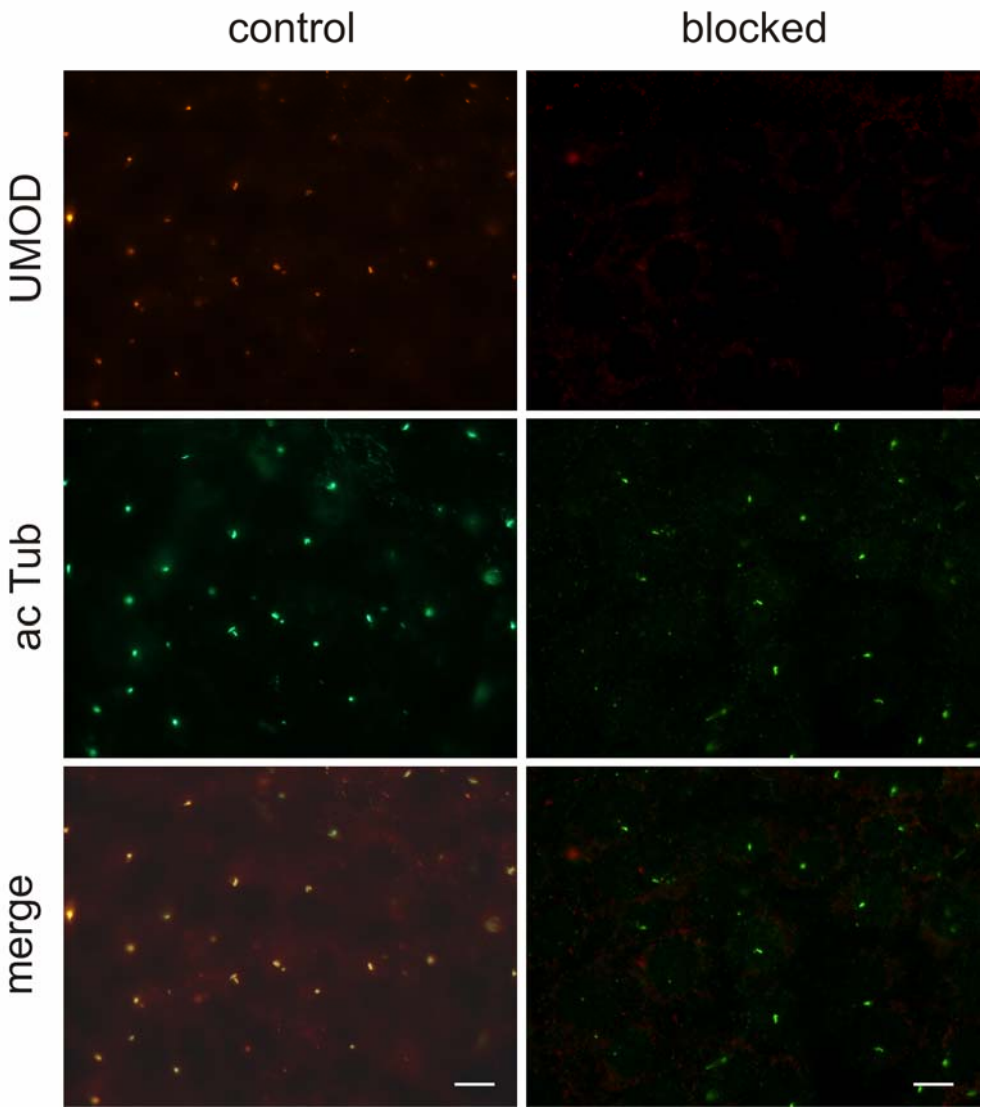
Supplemental Fig. 2. Ciliary UMOD expression in human renal biopsies of patients with other tubulo-interstitial kidney diseases

Renal samples from human control individuals (C1-C3) with tubulo-intersitial kidney diseases were stained for UMOD (left column) and acetylated tubulin (right column). In all kidney biopsy sections of the three patients with tubulo-intersitial kidney disease hair-like structures were identified protruding into the tubular lumen of a TAL (arrows), indicating a UMOD stained cilium (left column). Co-staining for acetylated tubulin detected exactly the same structures confirming ciliary expression of UMOD (right column). The patients with tubulo-intersitial kidney dieases did not show a significant different ciliary UMOD staining compared to healthy control individuals (Table 2). Control patients were diagnosed with primary hyperoxaluria (C1) and interstitial nephritis (C2, C3).

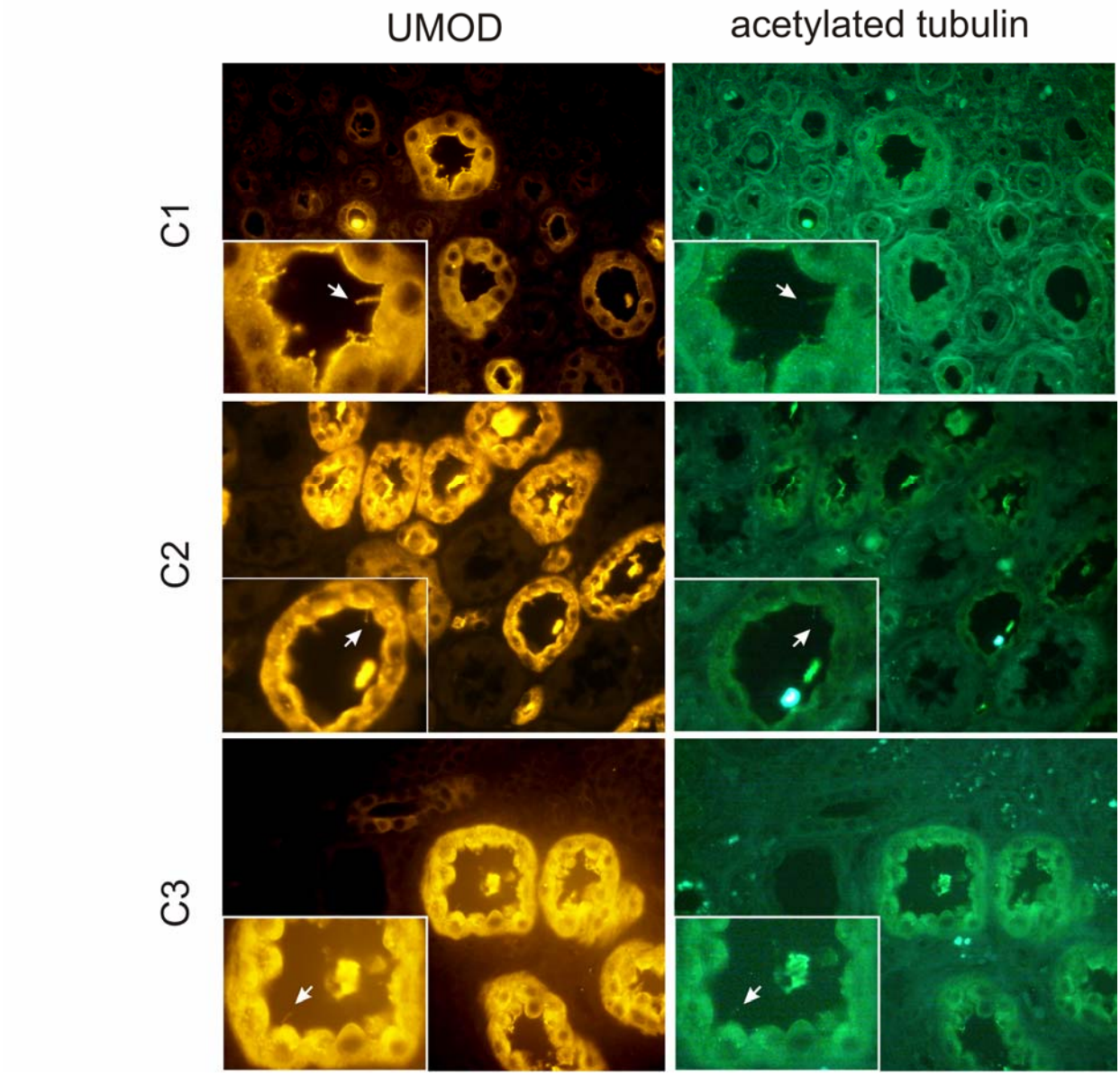
Supplemental Fig. 3. Overexpression of wildtype and mutant T225K UMOD in IMCD3 cells

IMCD3 cells were transfected with wildtype (a,c,e) or T225K mutant UMOD-EGFP fusion constructs (b,d,f) and processed for immunofluorescence staining. Using an antibody directed against GFP, both wildtype and mutant UMOD was mainly localized to the cell surface (a,b) However, cilia-like structures were observed in addition in cells transfected with the wildtype construct (a, marked by arrows). An antibody directed against acetylated tubulin was used to visualize cilia (c,d). The merge of GFP-UMOD and acetylated tubulin signal demonstrates that wildtype GFP-UMOD is localized to the cilia while mutant UMOD is not (e,f). However, mutant *UMOD* expressing cells still display cilia as indicated by arrows (f). The scale bar represents 10 μm.

Supplemental Fig. 1



Supplemental Fig. 2



Supplemental Fig. 3