globulinemia, Kwok *et al.*[1] present a similar case of falsely elevated creatinine concentration.[2] Interestingly, whereas in our case, Jaffé method was superior to enzymatic creatinine measurement in determining correct renal function, Kwok *et al.* report falsely elevated levels using the Jaffé method. Assay interference depends on the subtype of the elevated monoclonal immunoglobulin M and this may explain the observed differences.[3] Indeed, paraproteinemias, mainly monoclonal immunoglobulin M, have been reported to influence different estimation methods of renal function, putting patients at potential risks.[2–4] Of note, our case and the case by Kwok *et al.* measuring cystatin C revealed a correct measurement of kidney function, suggesting that cystatin C measurement is not affected by paraproteinemia and is thus a suitable method that allows the estimation of renal function in these cases.[2] If this proves true, determining cystatin C in patients with monoclonal gammopathy would prevent potential harm resulting from medication underdosing and renal replacement therapy. Therefore, prospective studies are required to determine the value of cystatin C measurements in patients with paraproteinemias.

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Expression and localization of the ciliary disease protein retinitis pigmentosa GTPase regulator in mammalian kidney

To the Editor: This letter is in reference to the recent article published by Schafer and colleagues,[1] in which they report the association of retinal disease protein retinitis pigmentosa GTPase regulator (RPGR) with the modulation of cystic kidney disease phenotype. We congratulate the authors for this remarkable finding.

RPGR mutations predominantly result in retinal degeneration. Some patients also exhibit sensorineural hearing loss, primary cilia dyskinesia, and sperm defects.[2,3] In light of the recent report,[1] we would like to provide additional corroborating evidence linking RPGR to the manifestation of renal diseases. We showed that RPGR is expressed in mammalian kidney; an RPGR-immunoreactive band can be detected at the expected size of ~170 kDa in mouse and rat kidney extracts (Figure 1a). Immunohistochemical analysis further showed that RPGR localizes to the podocytes in the glomerulus (Figure 1b) as well as to primary cilia in parietal epithelium and tubules (Figure 1c and d).

Given a critical role of ciliary function in regulating renal physiology,[4] these studies should heighten the awareness among clinicians and scientists in this newly recognized cilia associated renal–retinal connection with RPGR. We hope that these studies will also lead to further characterization of the mechanism of RPGR-associated multisystemic pathogenesis.

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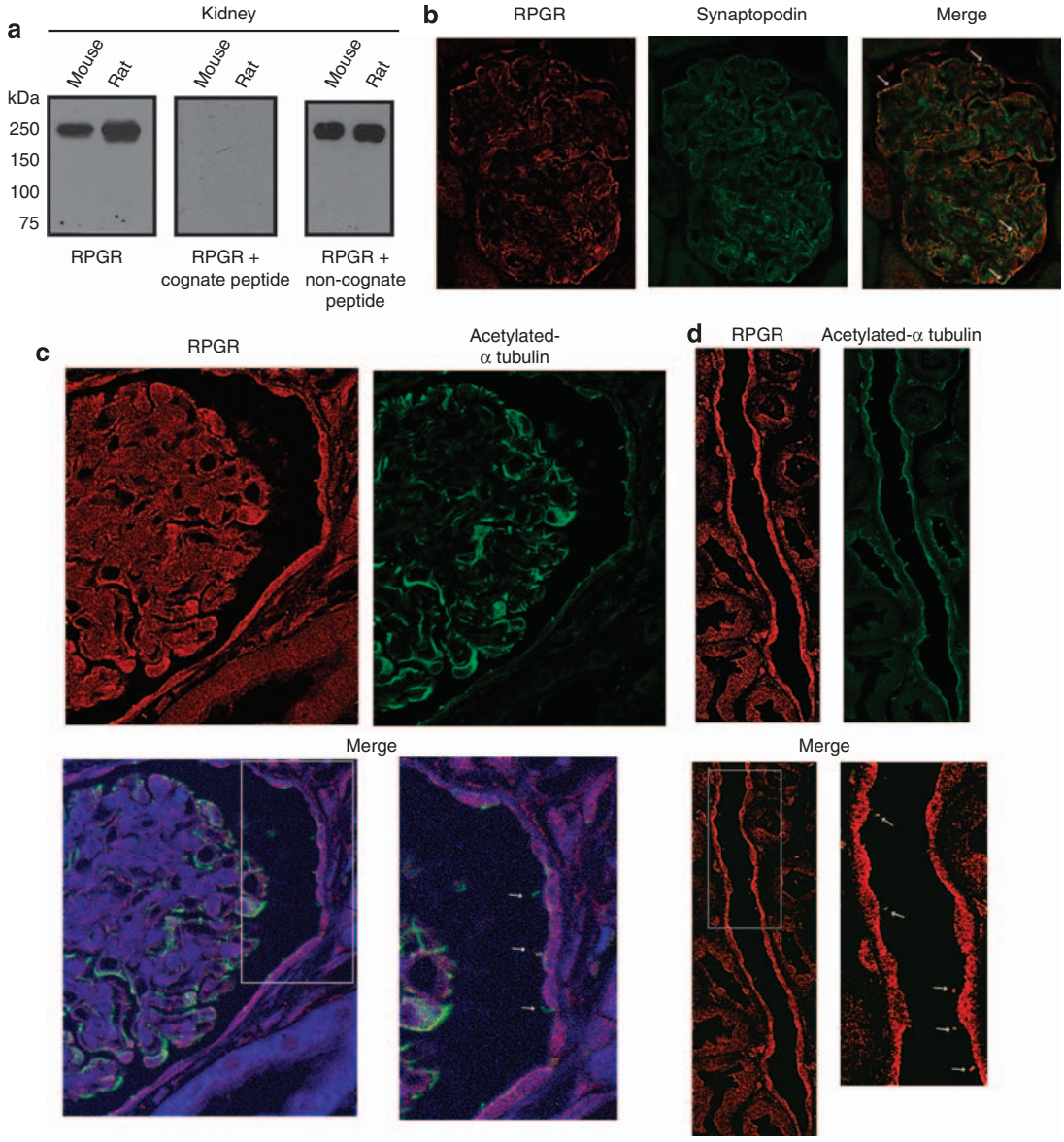

Figure 1 | Expression and localization of retinitis pigmentosa GTPase regulator (RPGR) in mammalian kidney. (**a**) Mouse and rat kidney lysates (20 μg) were analyzed by sodium dodecyl sulfate polyacrylamide gel electrophoresis and immunoblotting using a previously described RPGR antibody. To determine the specificity of the signal, antibody was preincubated with the cognate or noncognate peptide. (**b**) Rat kidney sections were stained with RPGR (red) or synaptopodin (green) (podocye foot process marker) and analyzed by confocal microscopy. Arrows in the Merge image show areas of colocalization. (**c**, **d**) RPGR (red) stains the cilia in parietal epithelium (**c**) and renal tubules (**d**). Cilia are labeled with acetylated α-tubulin (green). Nuclei are stained with 4′,6-diamidino-2-phenylindole (blue). Arrows in the Merge image and inset show colocalization of RPGR with the cilia marker.