

## Supplementary figure 1: mTOR pathway activation in proximal tubule cells and cyst lining epithelial cells.

Kidney sections from *Six2 Cre TSC1<sup>f/f</sup>* embryos at E17.5 were co-stained for phosphor S6 and LTL as indicated, showing mTOR pathway activation in proximal tubule cells and cyst lining epithelial cells. Scale bar =50µm.



Supplementary Figure 2: mTOR activation in proximal tubule cells of Six2 Cre<sup>tg/+</sup> TSC1<sup>t/f</sup> mice at P14, treated with Rapamycin during embryogenesis. Kidney sections of Six2 CreTSC1<sup>f/f</sup> mice treated with Rapamycin during embryogenesis. Six2 CreTSC1<sup>f/f</sup> and WT mice kidney sections at P14, were co-stained for phospho S6 and LTL, showing mTOR pathway activation in cyst lining epithelial cells. H&E staining scale bar=500µm, fluorescence staining scale bar =50µm.

WΤ



Supplementary figure 3: mTOR pathway is hyper activated in kidney extracts obtained from Six2 Cre TSC1 <sup>f/f</sup> mice compared to

WT mice. Kidneys from WT and Six2Cre TSC1<sup>#/f</sup> mice (n=5 in each group) were homogenized and immunoblotted for pS6 (as an indication for mTOR activity) and GAPDH. mTOR pathway is significantly activated in Six2Cre TSC1<sup>#/f</sup> kidney extracts compared to WT kidney extracts.