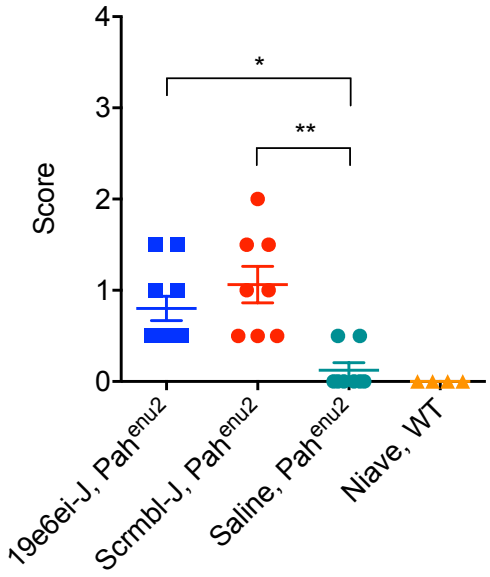
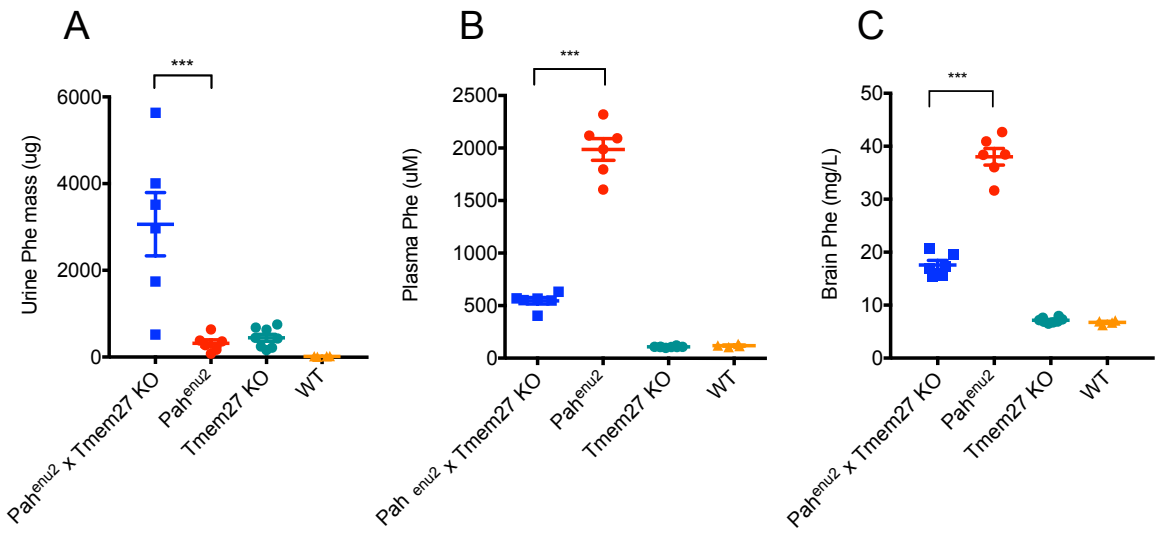
Supplementary Figures



Score	Criteria
0	No lesions observed
1	Mild: scattered small foci of tubular epithelial necrosis/regeneration affecting <25% of the tubules
2	Moderate: tubular epithelial necrosis/regeneration affecting 25-50% of the tubules
3	Marked: tubular epithelial necrosis/regeneration affecting 51-75% of the tubules
4	Severe: tubular epithelial necrosis/regeneration affecting 76-100% of the tubules

Supplementary Figure 1. Histopathology score after intravenous administration of peptide-conjugated morpholino oligonucleotides. Animals were injected intravenously (7.5 mg/kg) with *Slc6a19* PPMO (19e6ei-J), control PPMO (Scrmbl-J), or saline on days 0, 1, 8, 15, and 22. Kidneys were harvested on day 25, fixed in formalin, embedded in paraffin, sectioned at 5 microns and stained with hematoxylin and eosin. Slides were evaluated semi-quantitatively for the presence of renal tubular necrosis and regeneration using the scoring system shown above. *P<0.05, **P=0.0011 versus wild-type.



Supplementary Figure 2. Decreased Phe levels in *Pah*^{enu2} mice lacking *Tmem27*. (**A**) Excretion of Phe in the urine of *Pah*^{enu2} x *Tmem27* KO mice. Mice were placed in metabolic cages and urine was collected for 24h. (**B**) Phe levels in plasma. (**C**) Phe levels in brain. Animals were perfused with phosphate-buffered saline and whole brains were harvested. Phe was quantitated by HPLC/tandem MS/MS. Phe is expressed as mg Phe per liter of brain homogenate. N=4-8 animals/group. One-way ANOVA was performed, follow by Tukey's multiple comparisons test. ***P < 0.0001. Shown are means +/- SEM.