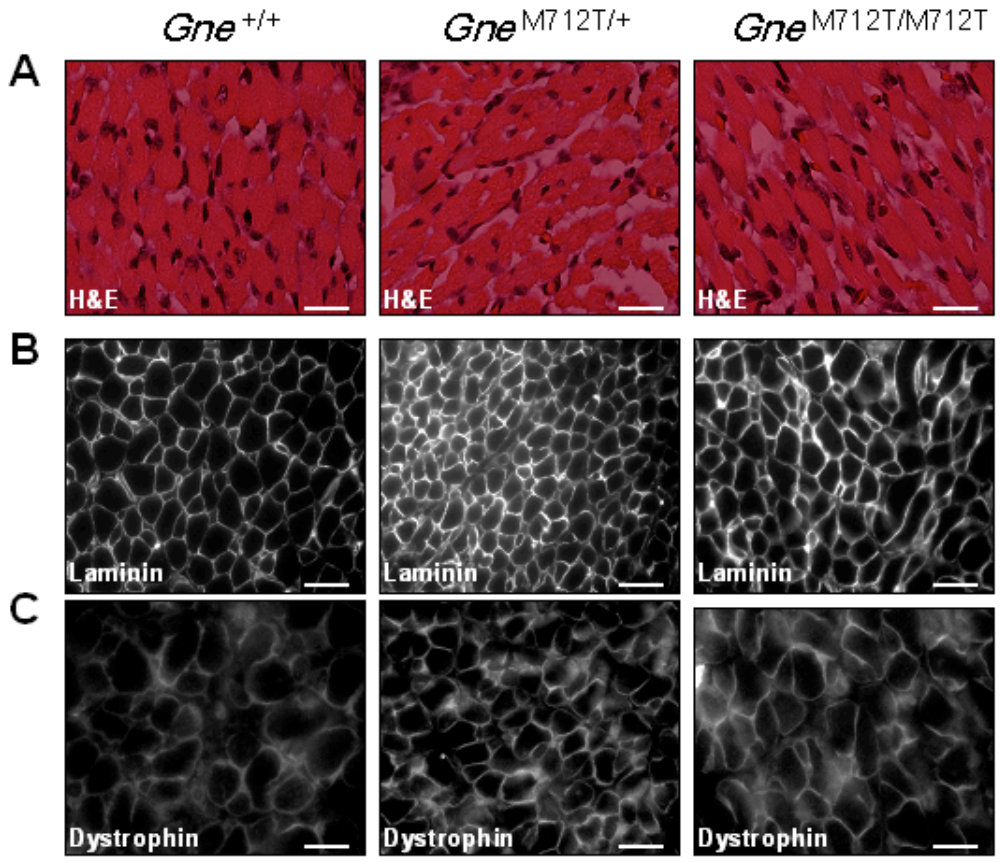
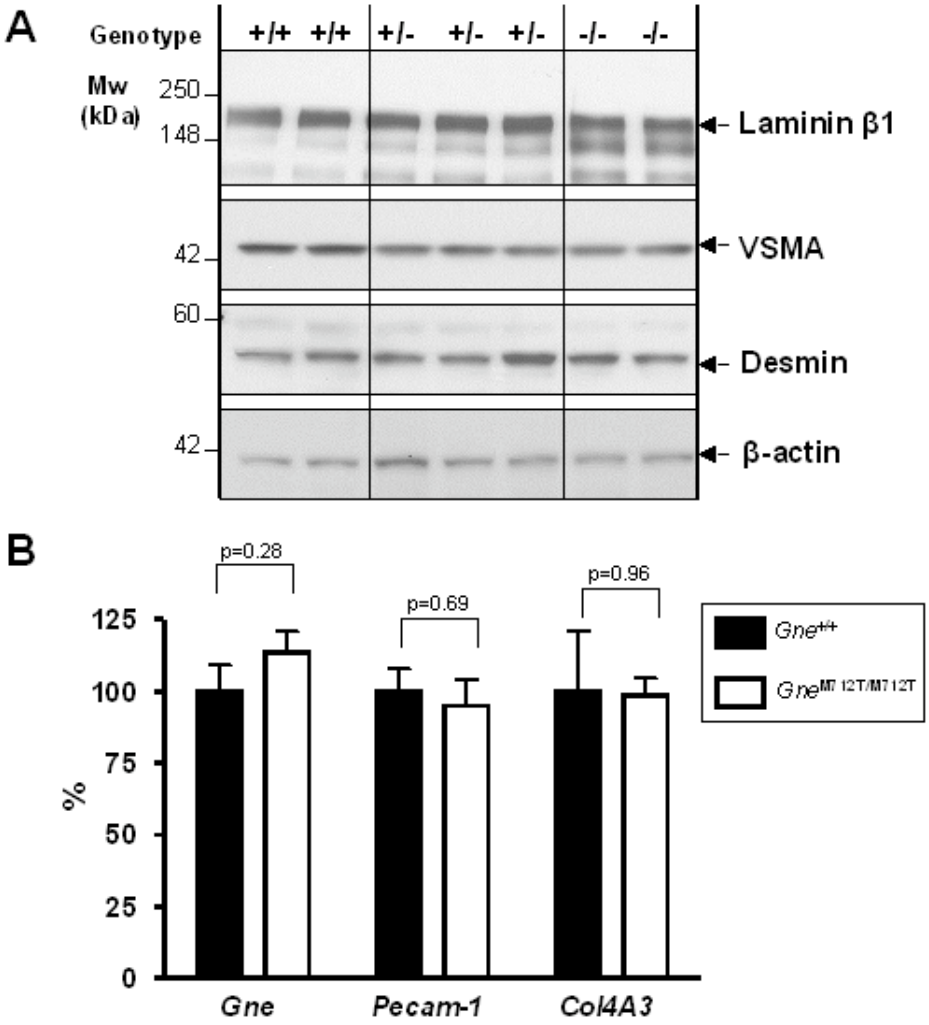
Supplementary Figure 1



Supplementary Figure 1: Muscle histology.

(**A-C**) Representative sections of skeletal muscle from wild type $Gne^{+/+}$, $Gne^{M712T/+}$, and homozygous mutated littermates at age P2. Scale bars, 20µm. (**A**) H&E stained sections show no difference in muscle histology among genotypes. (**B**) Laminin (extracellular component of the dystrophin-glycoprotein complex (DGC)) and (**C**) Dystrophin (intracellular component of the DGC) were normally distributed in all genotypes.

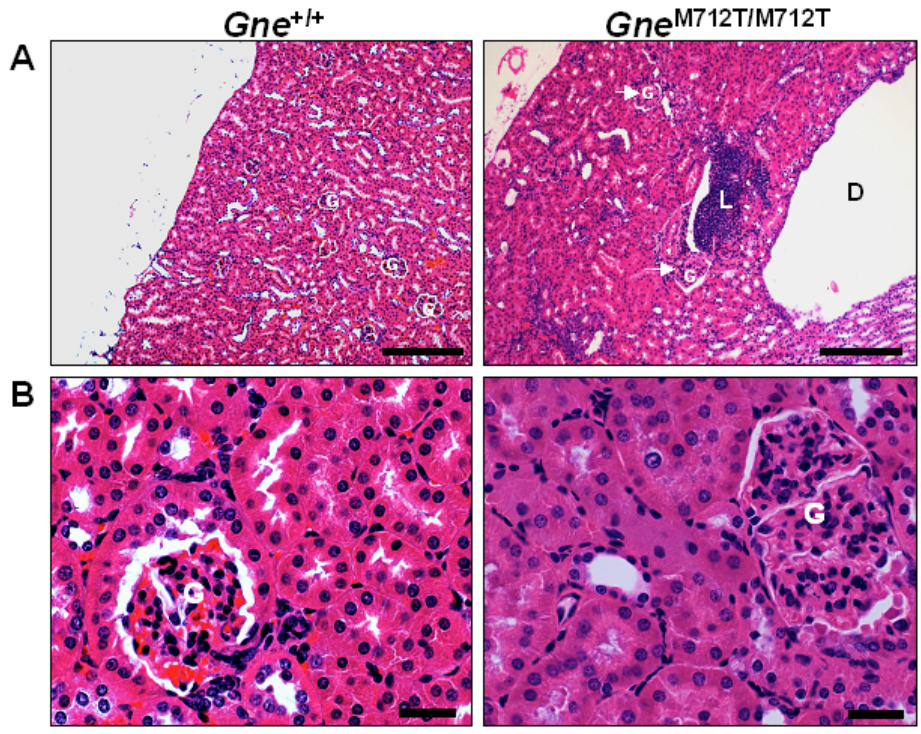
Supplementary Figure 2



Supplementary Figure 2: Expression levels of glomerular compartment markers.

(**A**) Representative immunoblots of kidney extracts (age P1) labeled with antibodies to GBM (laminin β1) and glomerular mesangial cell (VSMA and desmin) markers. After normalization to β-actin (~ 41 kDa), no difference in intensity or size was apparent in any of these markers between $Gne^{+/+}$ (+/+), $Gne^{M712T/+}$ (+/-), or $Gne^{M712T/M712T}$ (-/-): Laminin β1 (~ 210 kDa), VSMA (~42 kDa; n=3, p=0.65 (comparison of $Gne^{+/+}$ and $Gne^{M712T/M712T}$)), and Desmin (~ 53 kDa; n=3, p=0.93 (comparison of $Gne^{+/+}$ and $Gne^{M712T/M712T}$)). (**B**) mRNA expression levels in kidney of *Gne*, *CD31/Pecam-1* (endothelial cell marker), and *Col4A3* (GBM marker) were measured by real-time qPCR (n=3) and normalized to *β-actin* expression levels and $Gne^{+/+}$ expression levels for each gene were set at 100%. None of the tested genes differed significantly in mRNA expression in tissues of $Gne^{M712T/M712T}$ compared to wild type littermates.

Supplementary Figure 3



C

	BUN[a] (mg/dL)	Creatinine (mg/ dL)	Creatine Kinase (U/L)
+/+ [b]	18 (± 2)	0.33 (± .10)	410 (± 308)
+/-	21 (± 2)[b]	0.33 (± .10)[b]	525 (± 482)[c]
-/-	39 (± 10)[d]	0.40 (± .17)[d]	575[e]

[a] BUN= Blood Urea Nitrogen; [b] 3 mice, n= 9; [c] 3 mice, n=5; [d] one mouse, n=3; [e] one mouse, n=2

Supplementary Figure 3: Histological kidney analyses.

(**A** and **B**) Representative H&E stained kidney sections of the only *Gne*$^{M712T/M712T}$ (male) survivor (right panels) and a wild type male littermate (left panels), euthanized at age 8.5 months. (**A**) Low magnification views demonstrated markedly cystic dilations of the renal pelves (D), multifocal lymphoplasmacytic infiltrates (L), and expanded glomeruli (G) (arrows in right panel) in the *Gne*$^{M712T/M712T}$ mouse, which were not present in its wild type littermate. Scale bars, 500µm. (**B**) Representative higher magnification images demonstrated multiple *Gne*$^{M712T/M712T}$ glomeruli with mild to moderate mesangial matrix increase, and/or mesangial cell hyperplasia/hypertrophy (representative expanded glomerulus, right panel). These abnormalities were not observed in the wild type mouse (left panel). Scale bars, 100µm. **(C)** Average serum metabolite levels in weaned mice age 4 months. +/+: *Gne*$^{+/+}$; +/-: *Gne*$^{M712T/+}$; -/-: *Gne*$^{M712T/M712T}$ (the only mutant survivor without ManNAc treatment).