In vivo Animal Models

Alport Syndrome

Col4a3$^{-/-}$ Mouse Model

Mice homozygous for the Col4a3 targeted mutation are a model for autosomal-recessive Alport syndrome. Animals bred on a 129/SvJ background develop glomerulonephritis and die at about 10 weeks of age. Starting at an age of 4 weeks, Col4a3$^{-/-}$ mice and wild type littermates of mixed sex were treated with Ramipril or vehicle via drinking water. Treatment was continued until humane endpoints were reached.

- Weight loss
- Renal pathology
- Reduced survival
- Phenotype can be rescued by Ramipril
- Severe motor deficits starting at 12 weeks and worsening over age

Survival

Figure 1:

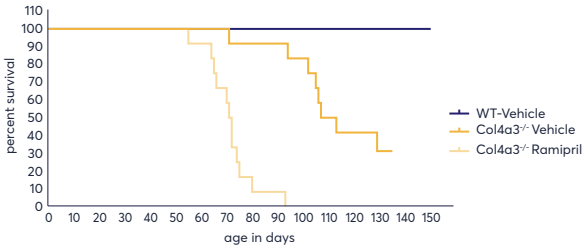


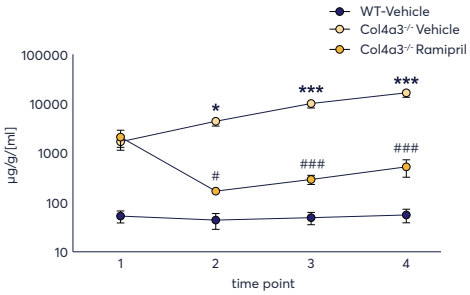
Figure 1:
Survival curve of Col4a3$^{-/-}$ mice after treatment with Ramipril or vehicle. n = 12 at start.

Figure 2:
Quantification of albumin in urine samples. Urine samples collected at treatment day 0, 10, 20, and 28. n = 6; mean ± SEM. Two-way ANOVA with Bonferroni's post hoc test. *Col4a3-/- Vehicle vs. WT-Vehicle; #Col4a3$^{-/-}$ Ramipril vs. Col4a3$^{-/-}$ Vehicle; *p<0.05; ***p<0.001.

Figure 3:
Immunofluorescent labeling of the kidney with a Smooth Muscle Actin (SMA) specific antibody. SMA: red; DAPI: blue; Autofluorescence: green.

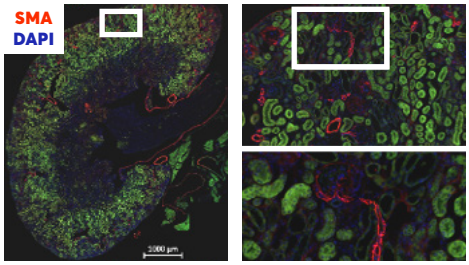
Urine Albumin

Figure 2:



Autofluorescence

Figure 3:



Cosgrove D, Meehan DT, Grunkemeyer JA, Kornak JM, Sayers R, Hunter WJ, Samuelson GC. Collagen COL4A3 knockout: a mouse model for autosomal Alport syndrome. Genes Dev. 1996 Dec 1;10(23):2981-92.

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Discovery

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