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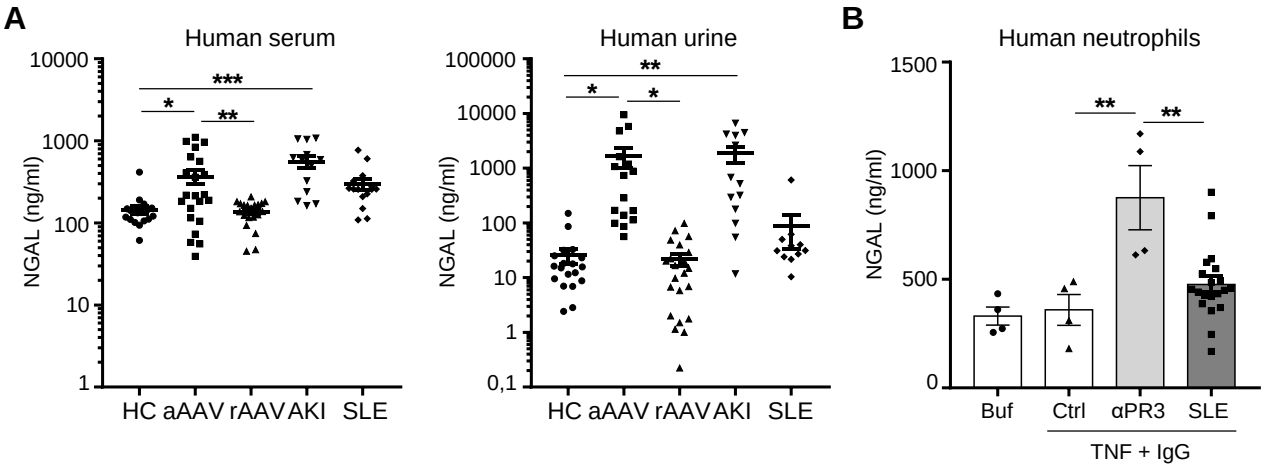

Figure S1. NGAL is increased in serum and urine of patients with ANCA-induced glomerulonephritis and NGAL is released by ANCA-activated neutrophils. (A) NGAL protein levels by ELISA were increased in serum and urine from AAV patients with active renal disease (aAAV) compared to AAV patients in remission (rAAV) and healthy controls (HC). NGAL levels in active AAV patients were similar to those in patients with non-AAV acute kidney injury (AKI) but elevated in comparison to patients with systemic lupus erythematosus (SLE). (B) TNFα-primed human neutrophils were stimulated with IgG (125 µg/ml) isolated from patients with either PR3-ANCA or SLE. Human control IgG preparations served as controls. NGAL protein detected by ELISA was significantly increased in supernatants of ANCA-stimulated human neutrophils but not in SLE-IgG stimulated neutrophils.

Supplemental Table 1. Demographic and clinical features of patients with AAV, AKI, SLE and healthy controls

Characteristics	AAV act.	AAV rem.	AKI	SLE	HC
Females, n (%)	5 (22.7)	12 (52.2)	6 (46,2)	14 (87.5)	10 (50)
Males, n (%)	17 (77.3)	11 (47.8)	7 (53.8)	2 (12.5)	10 (50)
Average age, years, mean ± SD	64.6± 10.3	57.6 ± 15.9	67.7 ± 16.5	39.4 ± 12.6	53.1 ± 16.3
Diagnosis, n (%)					
GPA	16 (72.7)	21 (91.3)	-	-	-
MPA	6 (27.3)	2 (8.7)	-	-	-
ANCA, n (%)					
PR3-ANCA	16 (72.7)	21 (91.3)	-	-	-
MPO-ANCA	6 (27.3)	2 (8.7)	-	-	-
Serum creatinine (mg/dl), mean ± SD	4.0 ± 2.7	1.1 ± 0.4	2.5 ± 1.3	1.4 ± 1.6	-

ANCA: antineutrophil cytoplasmic antibody; AAV: ANCA-associated vasculitis; act.: active; rem.: remission; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis; MPO: myeloperoxidase; PR3: proteinase 3; AKI: acute kidney injury; SLE: systemic lupus erythematosus; HC: healthy controls.

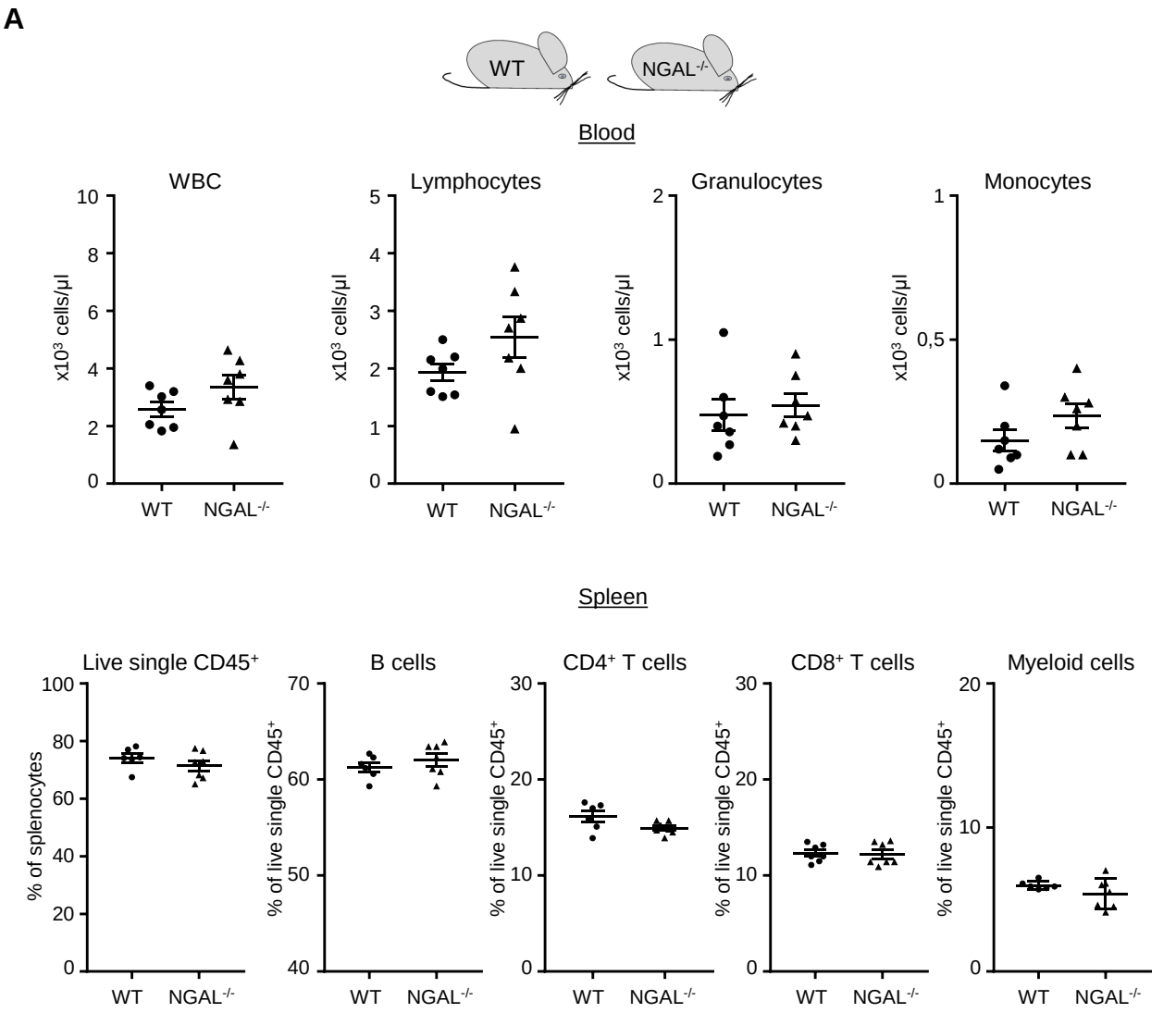

Figure S2. Wildtype and NGAL$^{-/-}$ mice have a similar hematological profile. White blood cell counts (WBC), lymphocyte, granulocyte, and monocyte count in blood was determined by the Pathophysiology Core Facility at the Max-Delbrück Center, Berlin, Germany. Splenic CD45^{+} immune cells, B220^{+} B cells, CD4^{+} and CD8^{+} T cells and CD11b^{+} myeloid cells were characterized by flow cytometry. CD45^{+} immune cells are expressed as percentage of splenocytes whereas B, T and myeloid cells are expressed as percentage of CD45^{+} cells.

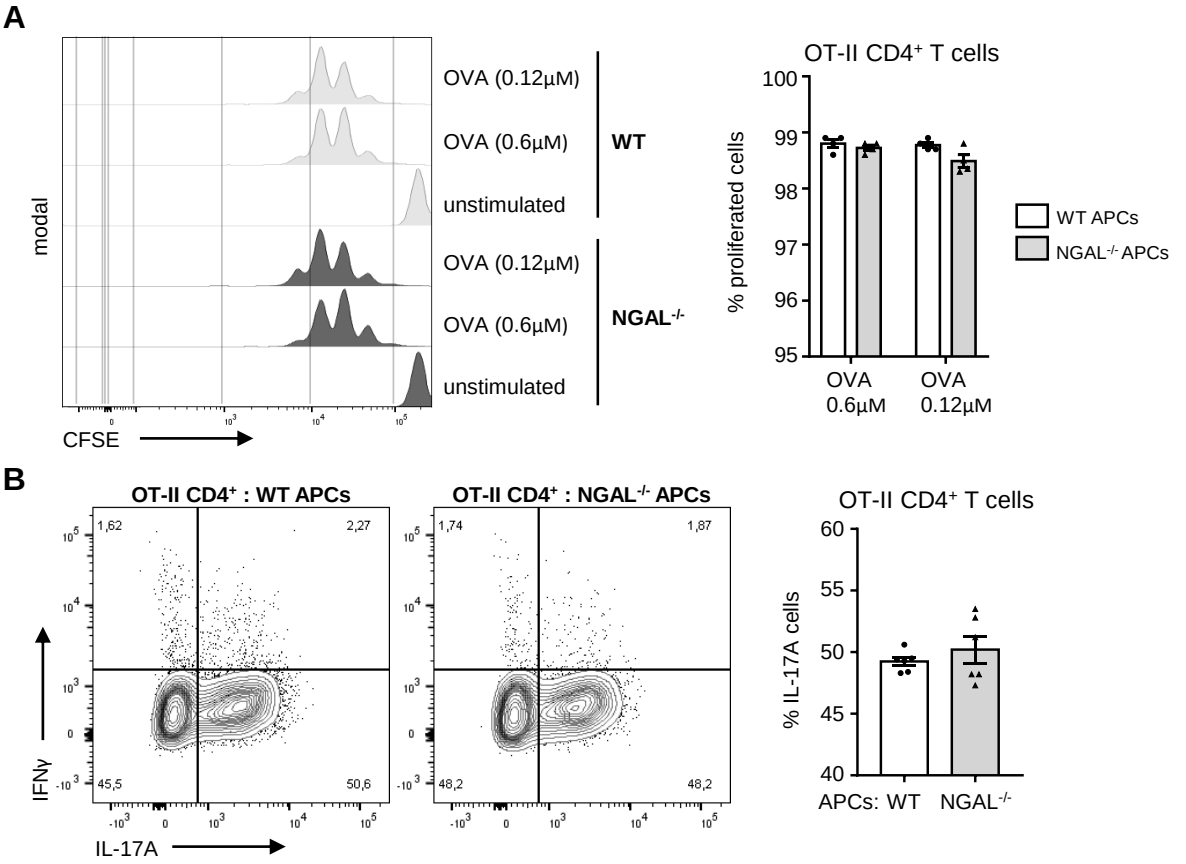

Figure S3. Antigen-presenting cells from NGAL$^{-/-}$ mice have normal functions. (A) Antigen-specific proliferation in response to two ova peptide$_{323-339}$ (OVA) concentrations was tested in CFSE-labeled CD4^{+} T cells isolated from spleens of OT-II transgenic mice and co-incubated with splenic antigen-presenting cells (APCs) from either WT or NGAL$^{-/-}$ mice for 3 days. Representative histograms and corresponding percentages of proliferated CD4^{+} T cells are shown. (B) Splenic CD4^{+} T cells from OT-II transgenic mice and splenic antigen-presenting cells (APCs) from either WT or NGAL$^{-/-}$ mice were cultured in presence of 0.6 µM ova peptide$_{323-339}$ in T_H17 polarization medium. Representative histograms and corresponding percentages of T_H17 T cells (IL-17A^{+}IFNγ^{-}) are shown.

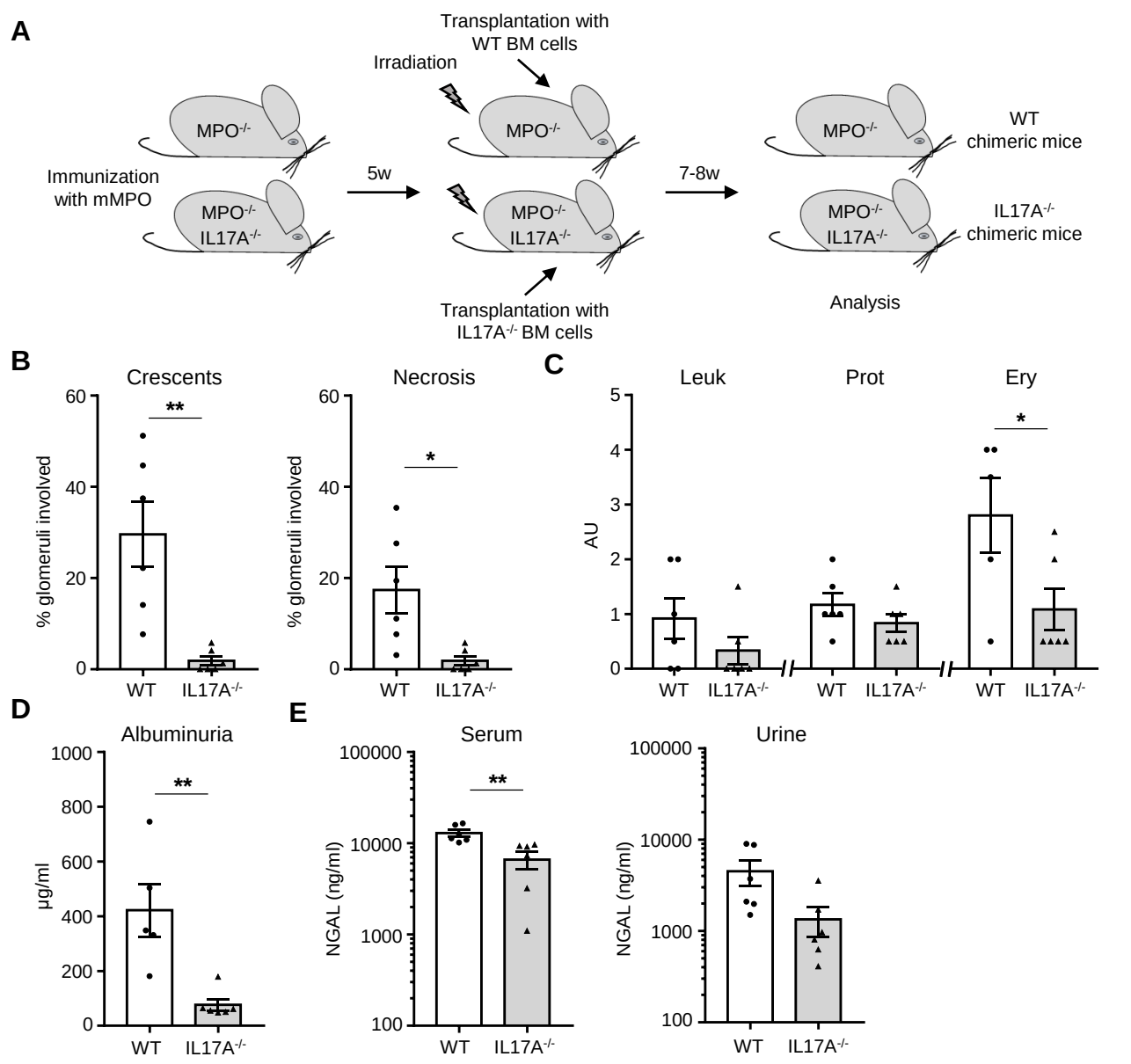

Figure S4. IL-17A genetic deletion protect mice from anti-MPO induced NCGN. (A) Experimental scheme for the anti-MPO NCGN induction in WT and IL-17A$^{-/-}$/MPO$^{-/-}$ chimeric mice. MPO$^{-/-}$ mice were immunized with murine MPO (mMPO), irradiated, and transplanted with wild-type (WT) bone marrow (BM). Immunized IL-17A$^{-/-}$/MPO$^{-/-}$ mice were irradiated and transplanted with IL-17A$^{-/-}$ BM. All chimeric mice were analyzed 7-8 weeks after transplantation. (B) Glomerular damage was assessed by histology and expressed as the mean percentage of glomeruli with crescents and necrosis. The percentage of glomeruli with crescents and necrosis was strongly reduced in the IL-17A$^{-/-}$ compared to the WT chimeric mice. (C) Leukocyturia, proteinuria, and erythrocyturia were measured in urine by dipstick. (D) Albuminuria was quantified by ELISA. (E) Serum and urinary NGAL levels were measured by ELISA.