

Figure S1

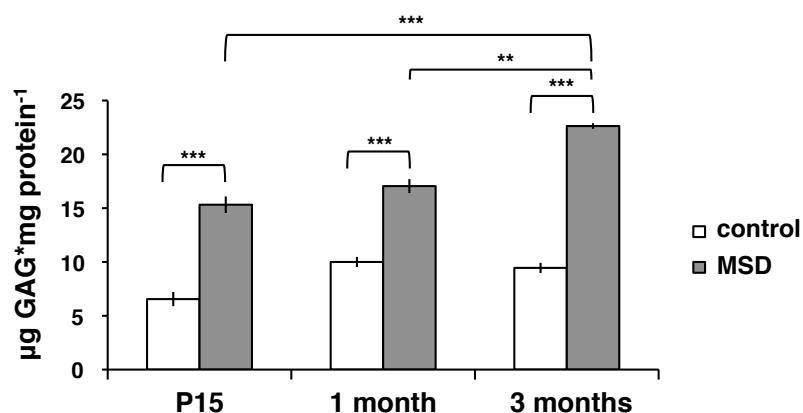
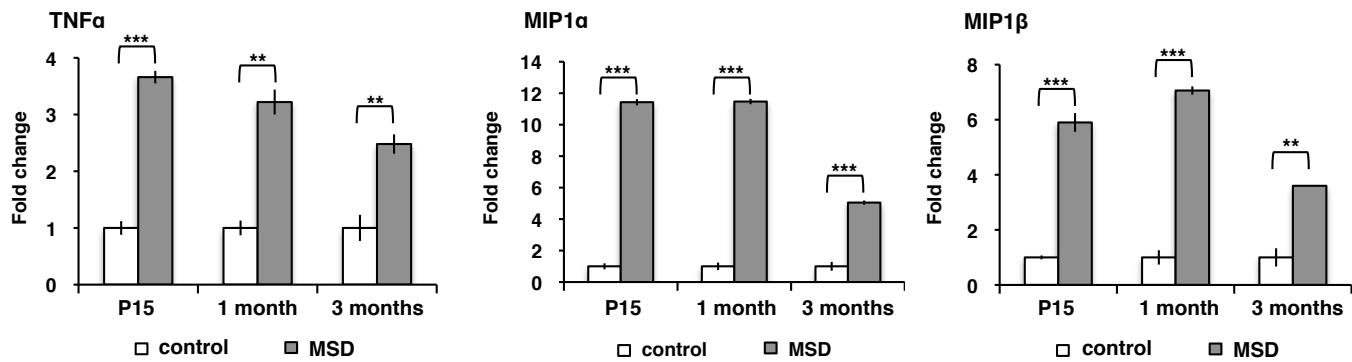


Figure S1. Progressive GAG accumulation in MSD liver. Quantification of glycosaminoglycans (GAGs) was performed using the DMB-Tris/Formate assay in liver samples from MSD (n=4) and control mice (n=4) at P15, 1 month and 3 months of age. Results are represented as the amount (μg) of GAG per mg of total protein. ANOVA p-value= 5.5E-09; ** p-value< 0.01; *** p-value< 0.005.

Figure S2

Liver



Brain

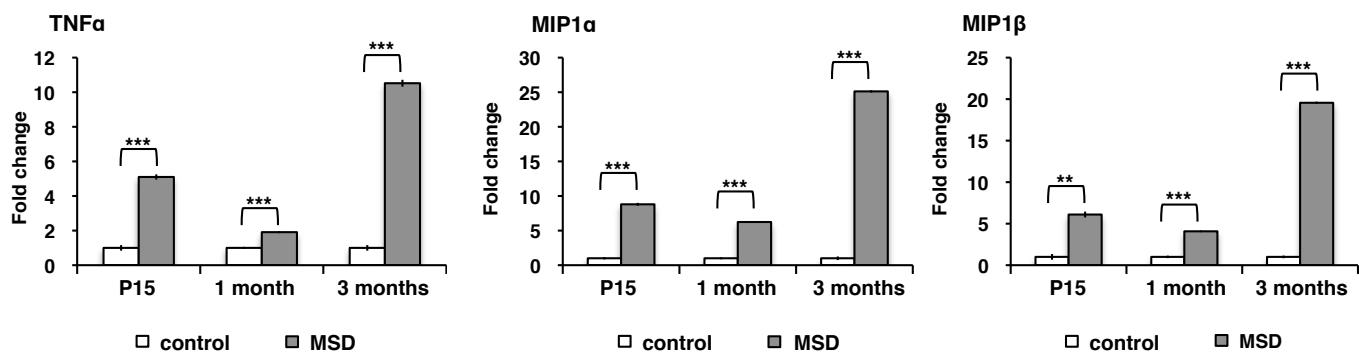


Figure S2. Expression of pro-inflammatory cytokines in MSD. Real-time PCR of pro-inflammatory cytokines (TNF α , MIP1 α and MIP1 β) in liver (top) and brain (bottom) of MSD (n=4) and control (n=4) mice at P15, 1 month and 3 months of age. Results are expressed as fold-change compared to control. ANOVA Liver TNF α p-value=3.09E-04, MIP1 α p-value=1.85E-04, MIP1 β p-value=2.36E-03; ANOVA Brain TNF α p-value=2.03E-03, MIP1 α p-value=1.37E-05, MIP1 β p-value=5.35E-05. ** p-value< 0.05; *** p-value< 0.001.