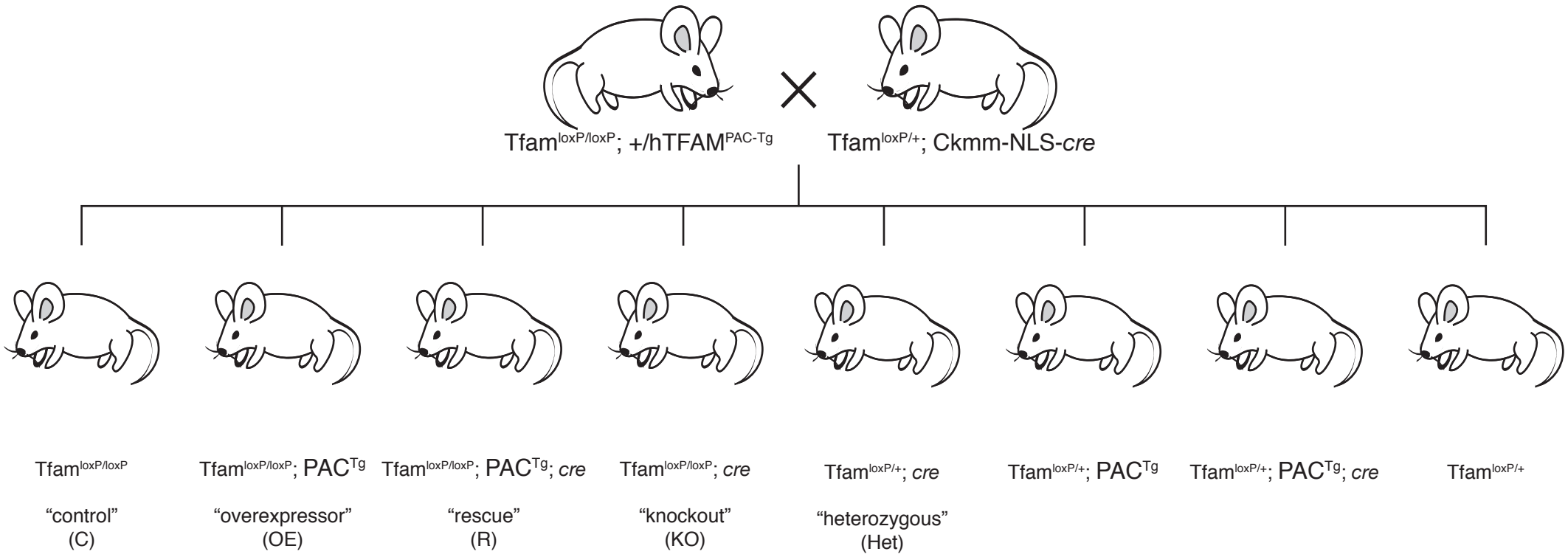
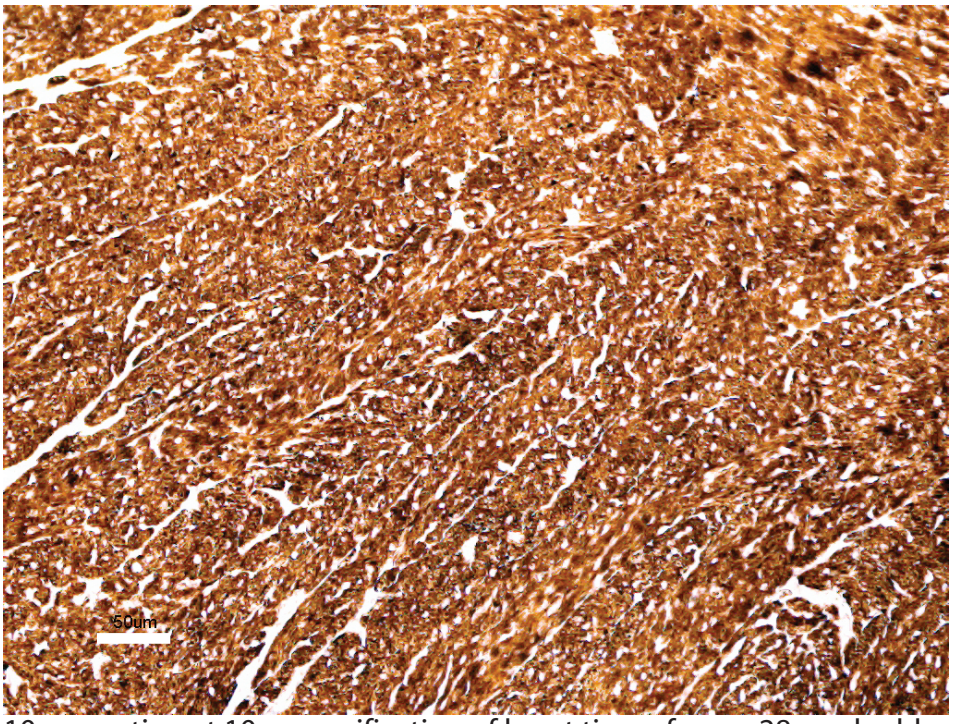
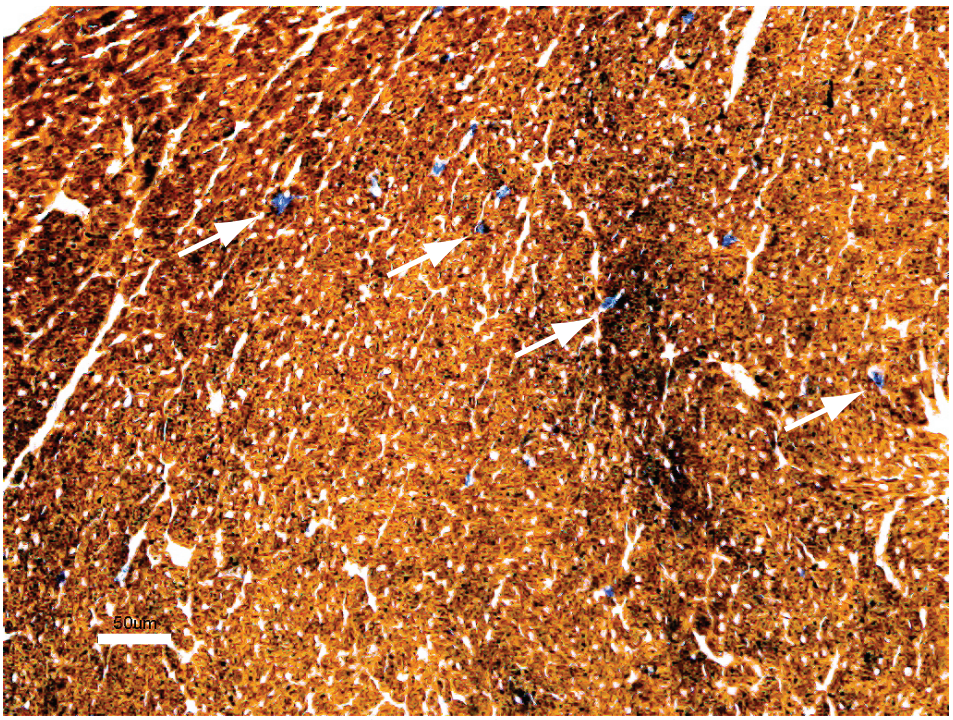


Figure S1: Schematic breading scheme used. Floxed Tfam mice, containing the hTFAM PAC transgene were crossed to heterozygous floxed Tfam mice, expressing *cre* recombinase from the Ckmm promoter. The offspring could be genotyped into: Tfam$^{loxP/loxP}$; Tfam$^{loxP/+}$; Tfam$^{loxP/loxP}$, PAC^{Tg}; Tfam$^{loxP/+}$, PAC^{Tg}; Tfam$^{loxP/loxP}$, *cre*; Tfam$^{loxP/+}$, *cre*; Tfam$^{loxP/loxP}$, PAC^{Tg}, *cre;* and Tfam$^{loxP/+}$, PAC^{Tg}, *cre*.



10um section at 10x magnification of heart tissue from a 28 week-old control animal. Size bar in 50μm.



10um section at 10x magnification of heart tissue from a 28 week-old rescue animal. COX-deficient cells are shown in blue (arrows). Size bar in 50μm.

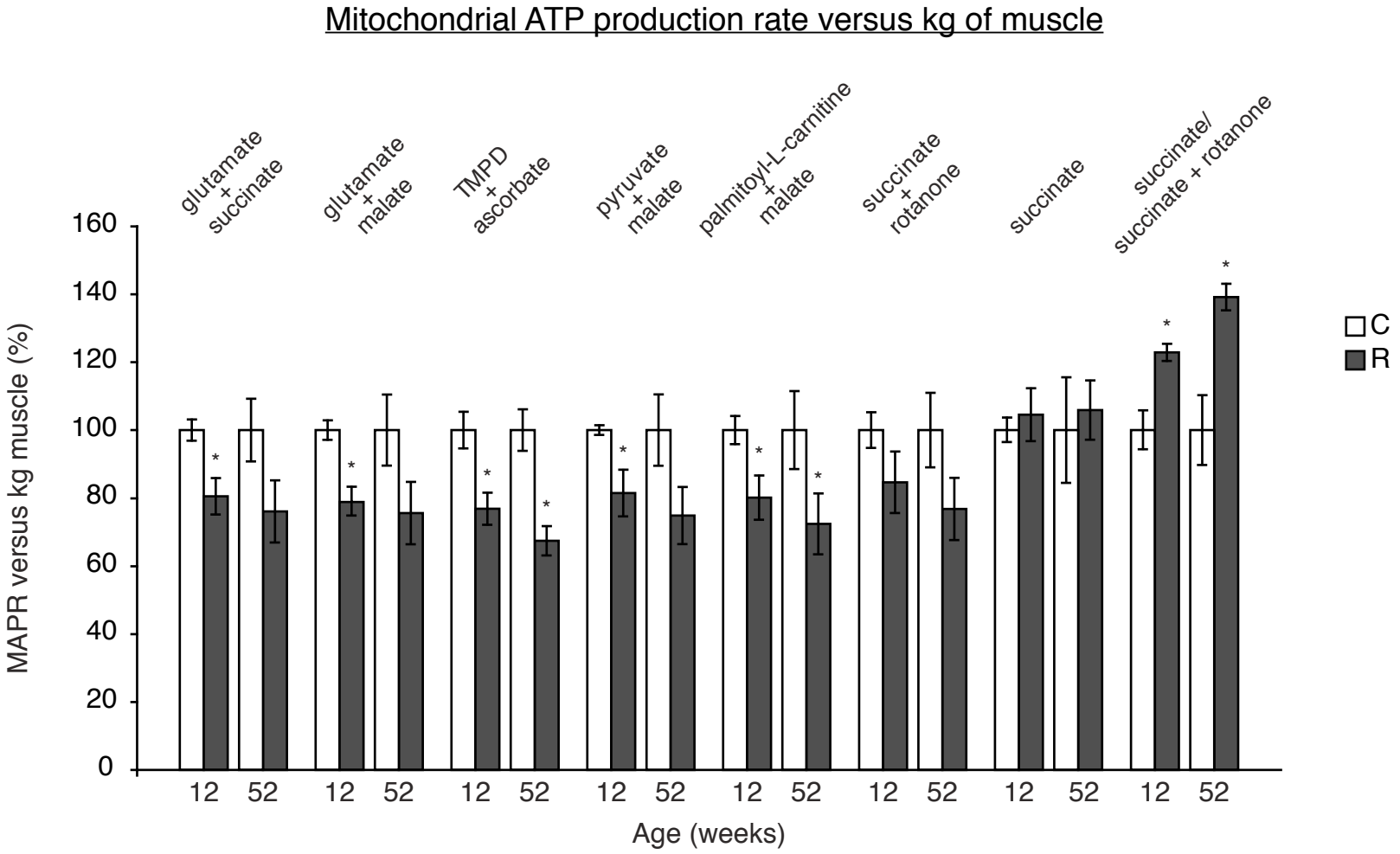


Figure S2. Results from measurements of MAPR per kg of heart tissue of control (white bars) and rescue (dark bars) mice at 12 and 52weeks of age. The relative MAPR per kg of tissue are presented as 100% to control. N=5 for all samples, except control animals at 52 weeks, where 4 samples were included. Data are presented as means ± SEM percentage of controls; * is $P < 0.05$.

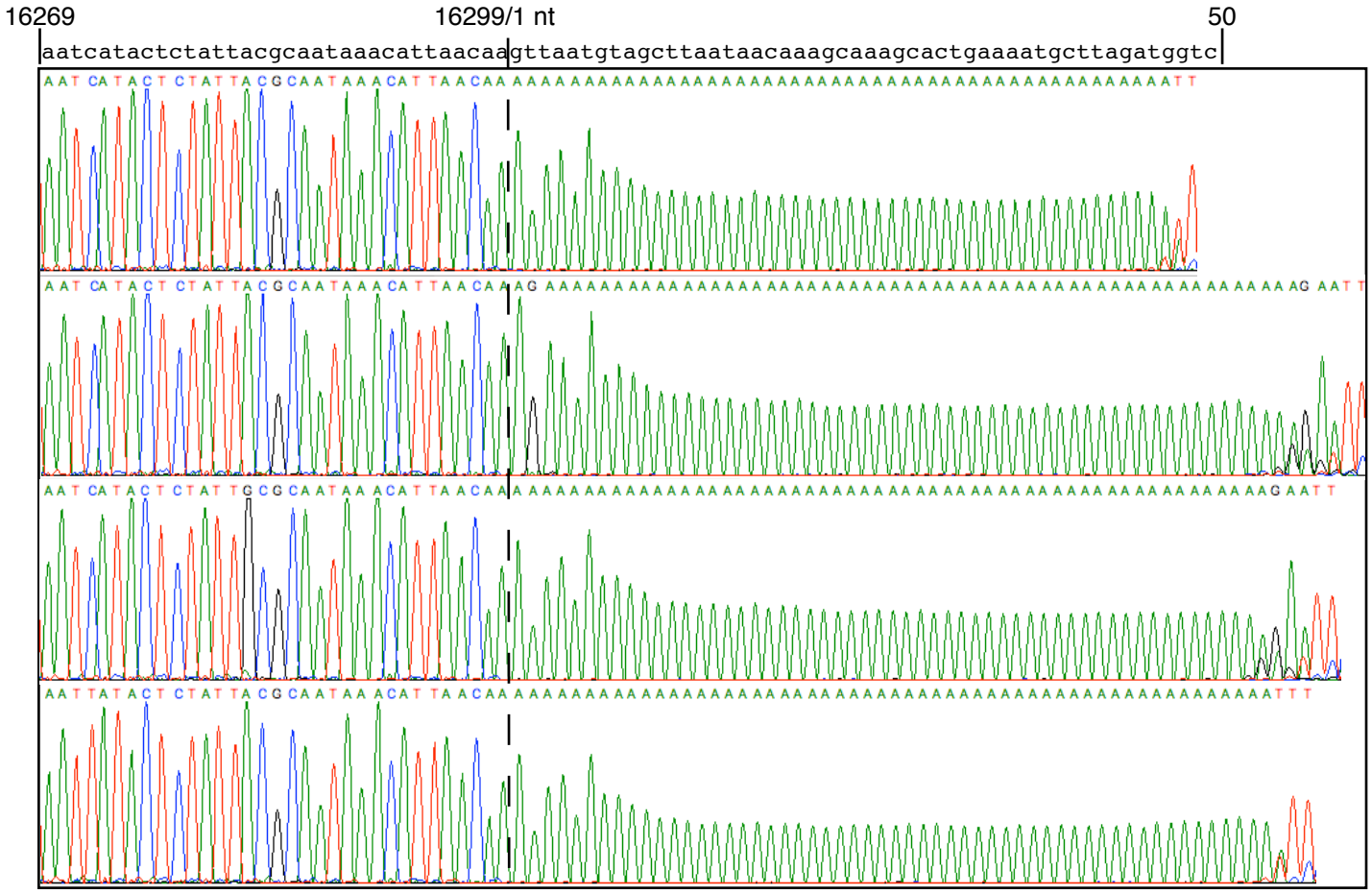


Figure S4. Electropherogram of 4 representative sequences obtained from 3' RACE experiments of the anti-control region transcript. Sequence and nucleotide position of the corresponding mouse mtDNA sequence are given in black.