

**Supplementary Figure S2.** Q-PCR analysis of tfam expression. Q-PCR analysis of expression of tfam exon 2,5 and 6-7 at day 4 in zebrafish injected with 2 ng tfam splice-MO compared to the Ctrl-MO (n= 6 per injected condition from gene expression analysis samples). Analysis of tfam exon 5 and exon 6-7 showed respectively 41% and 40% expression in tfam splice-MO injected embryos at day 4. This indicates that ~60% of tfam RNA is subjected to nonsense-mediated decay and that only half of the residual 40% tfam RNA is exon 2 containing wild-type RNA. Bars indicate mean values with SEM. P-values are calculated using ANOVA, \* p-value < 0.05.