

Supplemental Figure 3 Effect of Intrathecally administrated rhTPP1 on SCMAS storage in liver. Nineteen week-old *Tpp1*^{-/-} mice were administered three daily 80 μ L infusions of artificial CSF (V1-2) or 10 μ g/ μ L rhTPP1 (E1-3) and killed 24 hours after the final dose. Relative intensities (mean \pm standard deviation) of SCMAS staining in liver for vehicle-treated (n=2), enzyme-treated (n=3), and wild-type age controls (n=2), respectively, were as follows: 1.00 \pm 0.10, 0.48 \pm 0.06, 0.30 \pm 0.01. Note that this signal includes both normal mitochondrial and stored SCMAS.

