**Supplemental Table 1. Biodistribution of TPP1 preparations 24 hours following IV administration.** Data are expressed as pmol TPP1 per mg protein and also compared to levels present in control *Tpp1*(+/+) mice.

|  |  |  |
| --- | --- | --- |
| Genotype | *Tpp1*(+/+) | *Tpp1*(-/-) |
| Preparation | - | Unmodified TPP1 | OX-TPP1 | DG-TPP1 |
| Dose (mg) | - | 0.12 | 2.0 | 0.12 | 2.0 | 0.12 |
| Sample size | 5 | 5 | 8 | 2 | 3 | 2 |
|  | TPP1 specific activity (pmol/mg protein)(% of wild-type) |
| Brain | 2.18±0.45(100%) | 0.06±0.02(3%) | 0.23±0.13(11%) | 0.01±0.02(1%) | 0.11±0.04(5%) | 0.03±0.001(1%) |
| Liver | 5.80±0.94(100%) | 5.84±5.62(101%) | 78.3±18.0(1349%) | 2.78±0.54(48%) | 32.63±8.86(562%) | 4.64±0.06(80%) |
| Spleen | 5.76±1.48(100%) | 6.16±3.27(107%) | 57.6±11.7(1001%) | 0.88±0.26(15%) | 14.52±4.30(252%) | 2.98±0.19(52%) |
| Kidney | 7.46±1.54(100%) | 0.51±0.27(7%) | 3.21±0.78(43%) | 0.23±0.06(3%) | 3.20±0.66(43%) | 0.33±0.04(4%) |
| Heart | 1.12±0.19(100%) | 0.45±0.46(40%) | 8.33±3.90(746%) | 0.42±0.05(38%) | 7.30±1.57(654%) | 0.24±0.03(21%) |
| Lung | 4.78±0.52(100%) | 0.43±0.37(9%) | 6.44±2.75(135%) | 0.31±0.12(6%) | 5.46±1.11(114%) | 0.23±0.06(5%) |