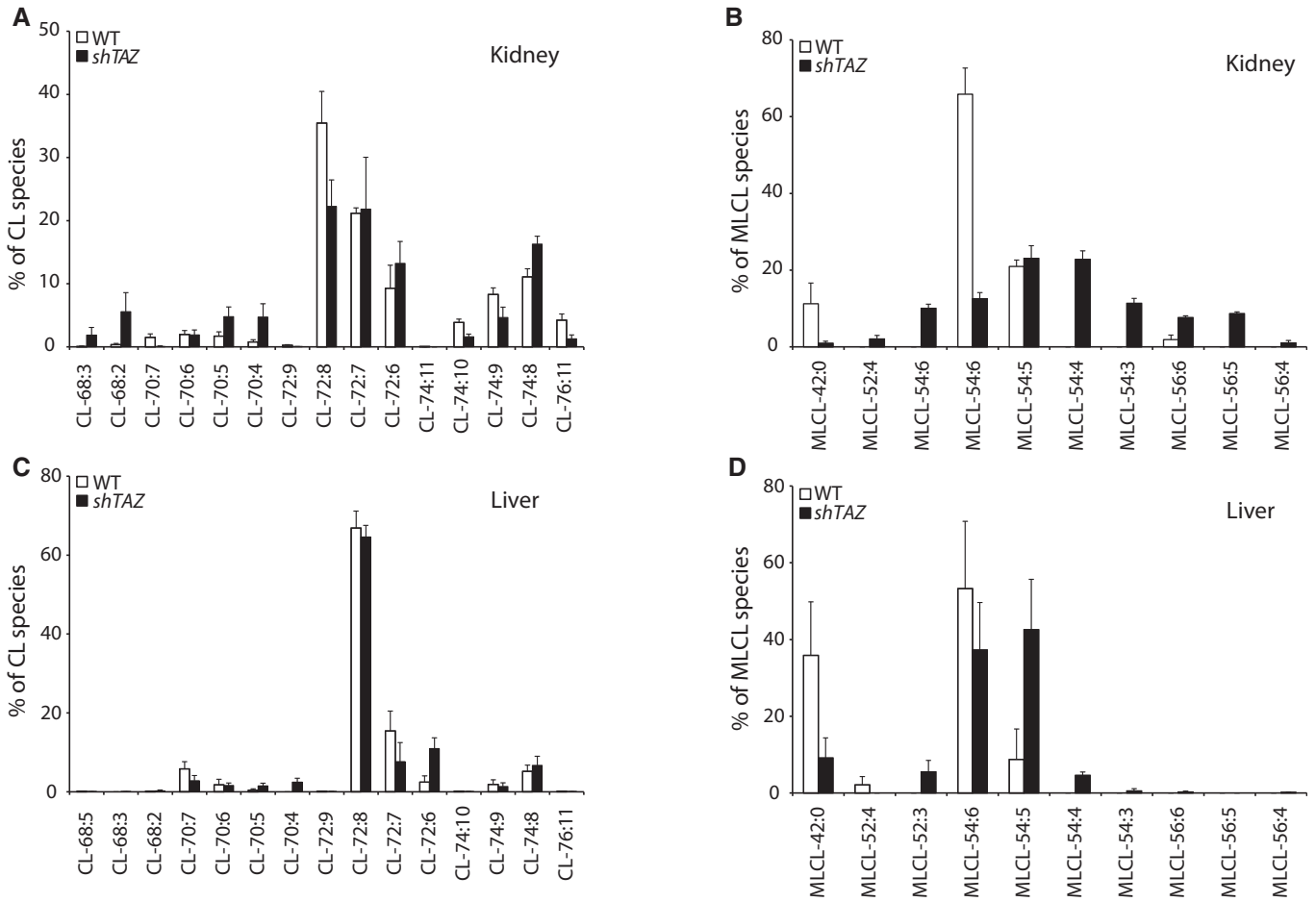


Expanded View Figures

**Figure EV1. Cardiolipin and monolysocardiolipin profiles in kidney and liver.**

A–D Lipid species profiles of cardiolipin (A, C) and monolysocardiolipin (B, D) in the mitochondria isolated from indicated tissues analyzed by mass spectrometry. Mean of $n = 4$ per genotype, \pm SEM.

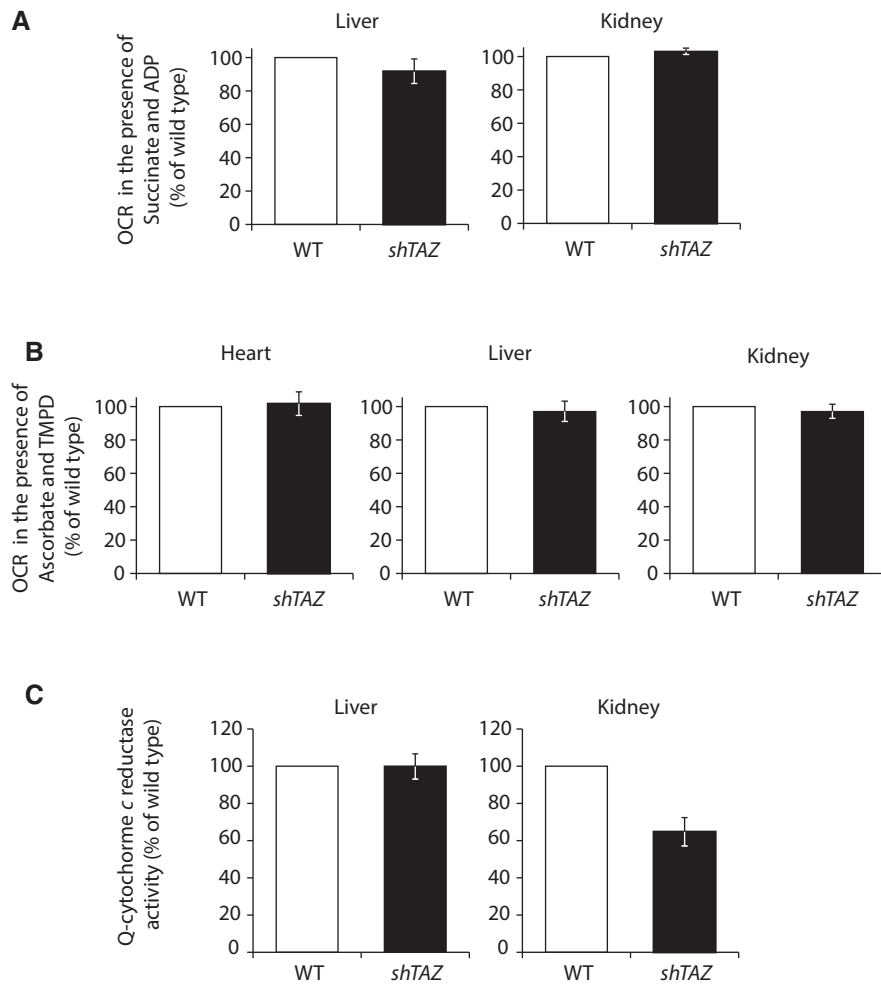


Figure EV2. Respiration is not affected in kidney and liver.

A Measurement of the oxygen consumption rate (OCR) of isolated mitochondria after the administration of succinate and ADP as substrate. Mitochondria from the liver (left) and kidney (right) were measured from 3 different animals per genotype, mean \pm SEM.

B OCRs of mitochondria after the administration of TMPD and ascorbate, isolated from the heart (left; $n = 8$ per genotype), liver (central; $n = 3$ per genotype), and kidney (right; $n = 3$ per genotype); data represented in mean, \pm SEM.

C Enzymatic activity of complex III in the liver (left; $n = 3$ per genotype) and kidney (right; $n = 2$ per genotype) normalized to MDH activity, in percent with control set to 100% (mean \pm SEM). Complex III activity was measured spectrophotometrically as an increase in reduced cytochrome c over time.