

Expanded View Figures

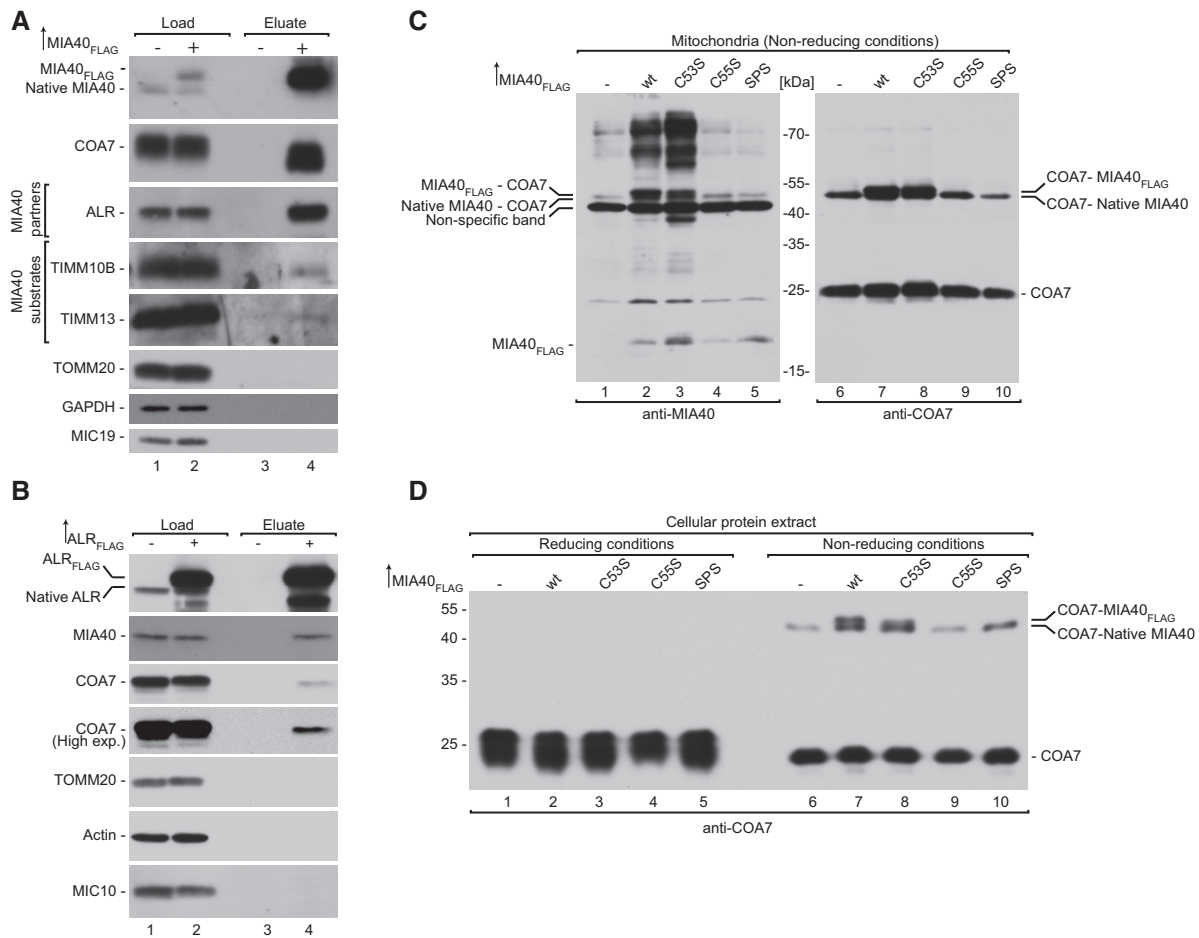


Figure EV1. COA7 interacts with MIA40 by disulfide bonding.

- A Flp-In T-REx 293 cells induced to express MIA40_{FLAG} were solubilized, and the affinity purification of MIA40_{FLAG} was performed. The fractions were analyzed by SDS-PAGE and Western blot. Load: 2.5%. Eluate: 100%.
- B Flp-In T-REx 293 cells induced to express ALR_{FLAG} were solubilized, and the affinity purification of ALR_{FLAG} was performed. The fractions were analyzed by SDS-PAGE and Western blot. Load: 2.5%. Eluate: 100%.
- C Mitochondria were isolated from Flp-In T-REx 293 cells induced to express wild-type and mutant forms of MIA40_{FLAG} under non-reducing conditions. The extract was analyzed by non-reducing SDS-PAGE and Western blot.
- D Cellular protein extracts were isolated from Flp-In T-REx 293 cells induced to express wild-type and mutant forms of MIA40_{FLAG} under non-reducing conditions. The extract was analyzed by non-reducing SDS-PAGE and Western blot.

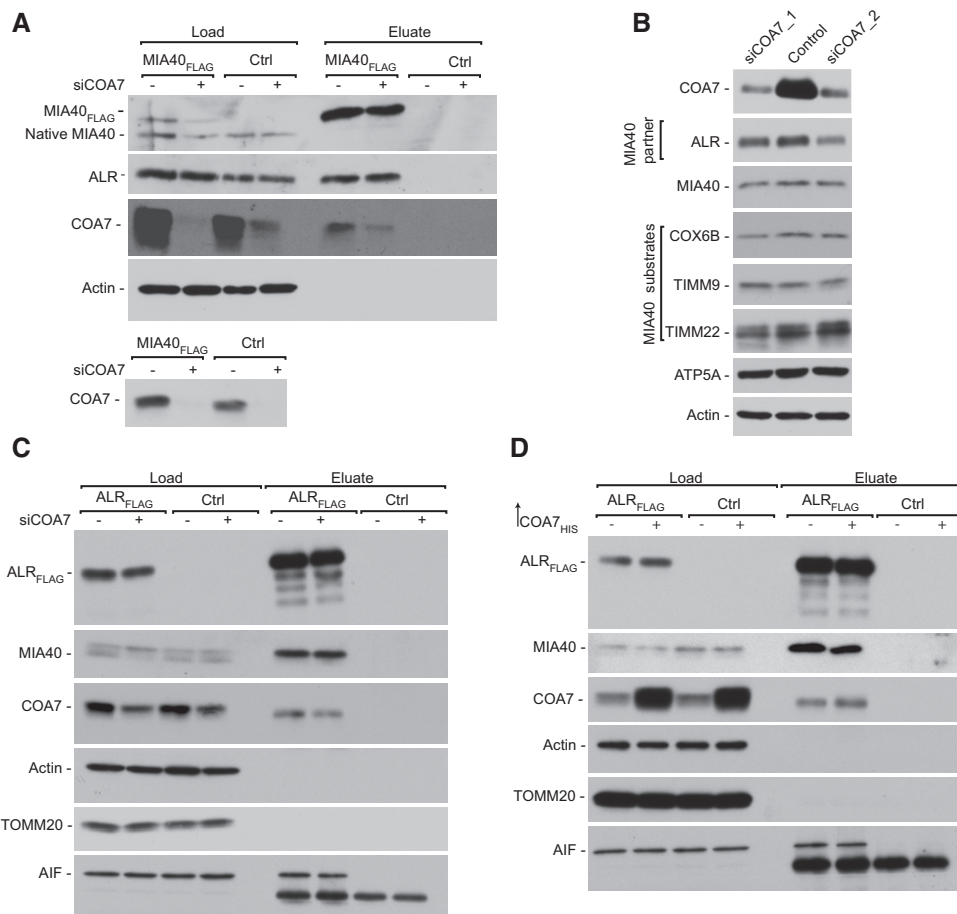


Figure EV2. COA7 does not affect MIA40-ALR interaction.

- A Protein extracts were isolated from Flp-In T-REX 293 cells induced to express MIA40_{FLAG} that was transfected with oligonucleotides that targeted COA7 mRNA or control oligonucleotides and subjected to affinity purification. The samples were analyzed by SDS-PAGE and Western blot. Load: 2.5%. Eluate: 100%.
- B Cellular protein extracts were isolated from HeLa cells that were transfected with oligonucleotides that targeted different regions of COA7 mRNA or control oligonucleotides. The samples were analyzed by reducing SDS-PAGE and Western blot.
- C Protein extracts were isolated from Flp-In T-REX 293 cells induced to express ALR_{FLAG} that was transfected with oligonucleotides that targeted COA7 mRNA or control oligonucleotides and subjected to affinity purification. The samples were analyzed by SDS-PAGE and Western blot. Load: 2.5%. Eluate: 100%.
- D Flp-In T-REX 293 cells induced to express ALR_{FLAG} were transfected with a plasmid that encoded COA7_{HIS} or an empty vector. The affinity purification of ALR_{FLAG} was performed, and samples were analyzed by SDS-PAGE and Western blot.

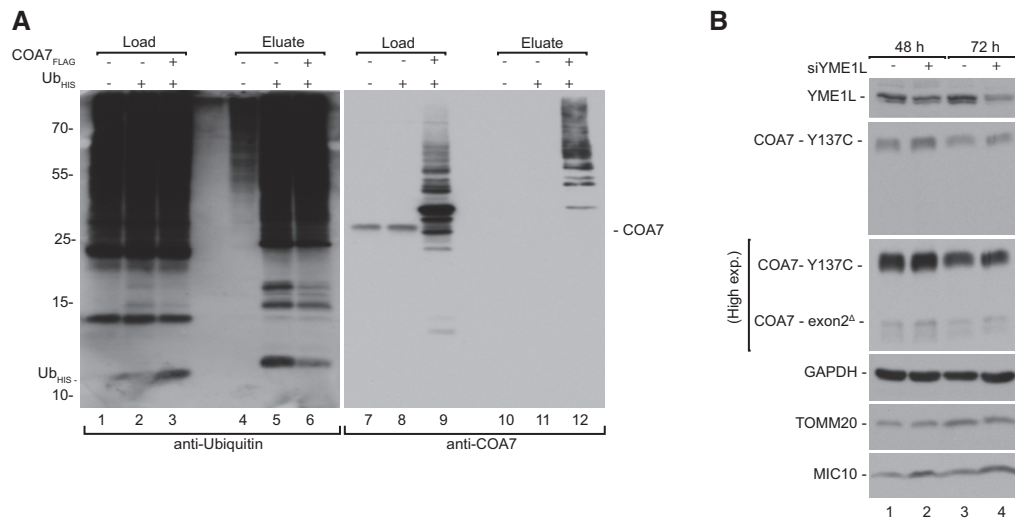


Figure EV3. Involvement of ubiquitin–proteasome system and YME1L in degradation of COA7 and its mutants.

- A Cellular protein extracts were isolated from HEK293 cells that expressed ubiquitin_{HIS} (Ub_{HIS}) and COA7_{FLAG} and subjected to affinity purification. Load: 2.5%. Eluate: 100%.
- B Cellular protein extracts were isolated after 48 and 72 h from patient fibroblast that were transfected with oligonucleotides that targeted different regions of YME1L mRNA or control oligonucleotides. The samples were analyzed by reducing SDS–PAGE and Western blot.