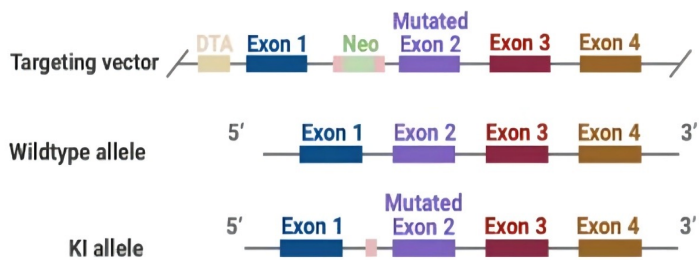
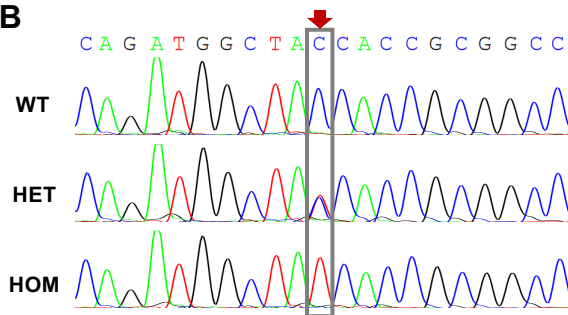


Supplementary Figures

A



B



C

WT 5' ... CTGATGGCC CAGATGGCTACCACCGCGGCCGGTGTGGCT ...3'

#

T61I 5' ... CTGATGGCC CAGATGGCTATCACCAGCGGCCGGTGTGGCT ...3'

D

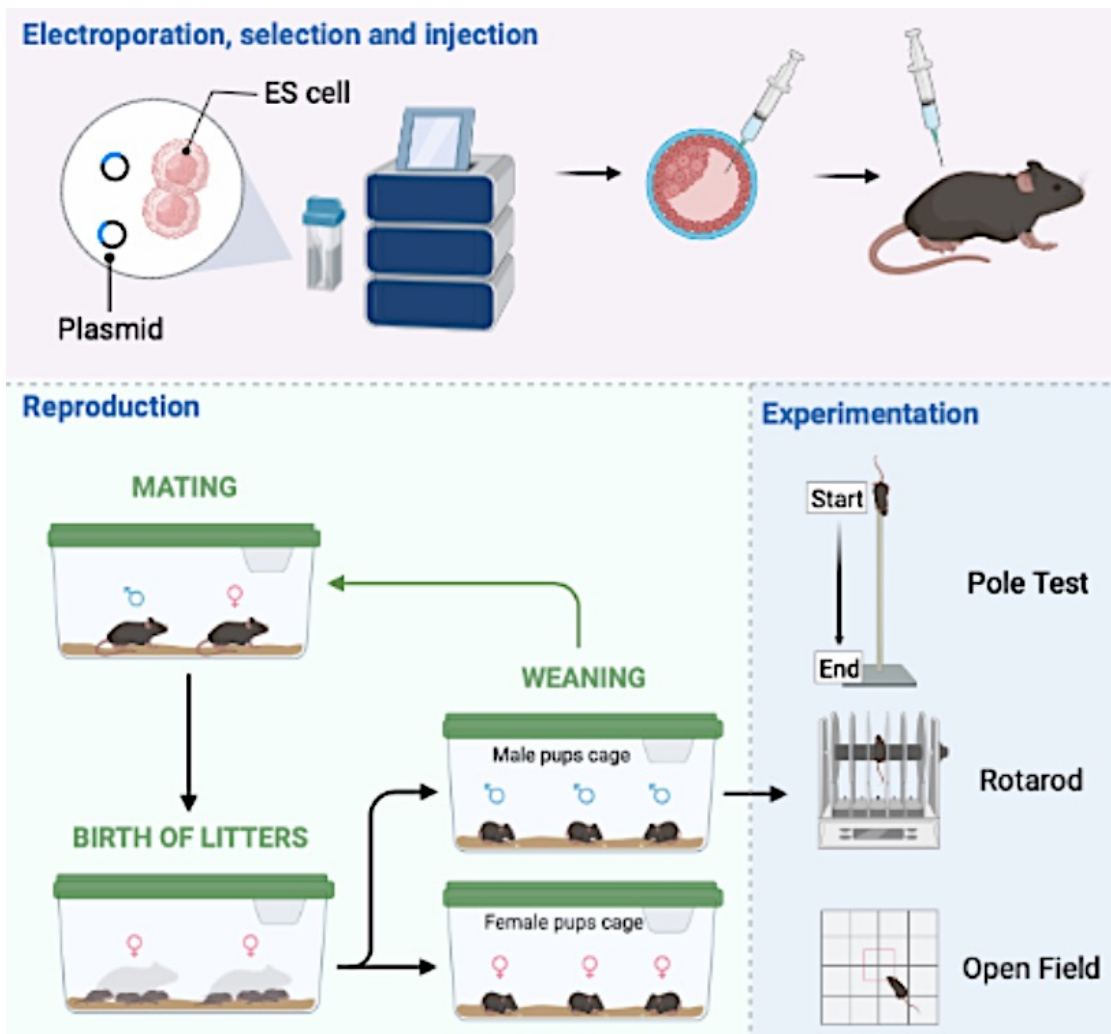


Figure S1. Generation of CHCHD2 p.T61I mutant mice.

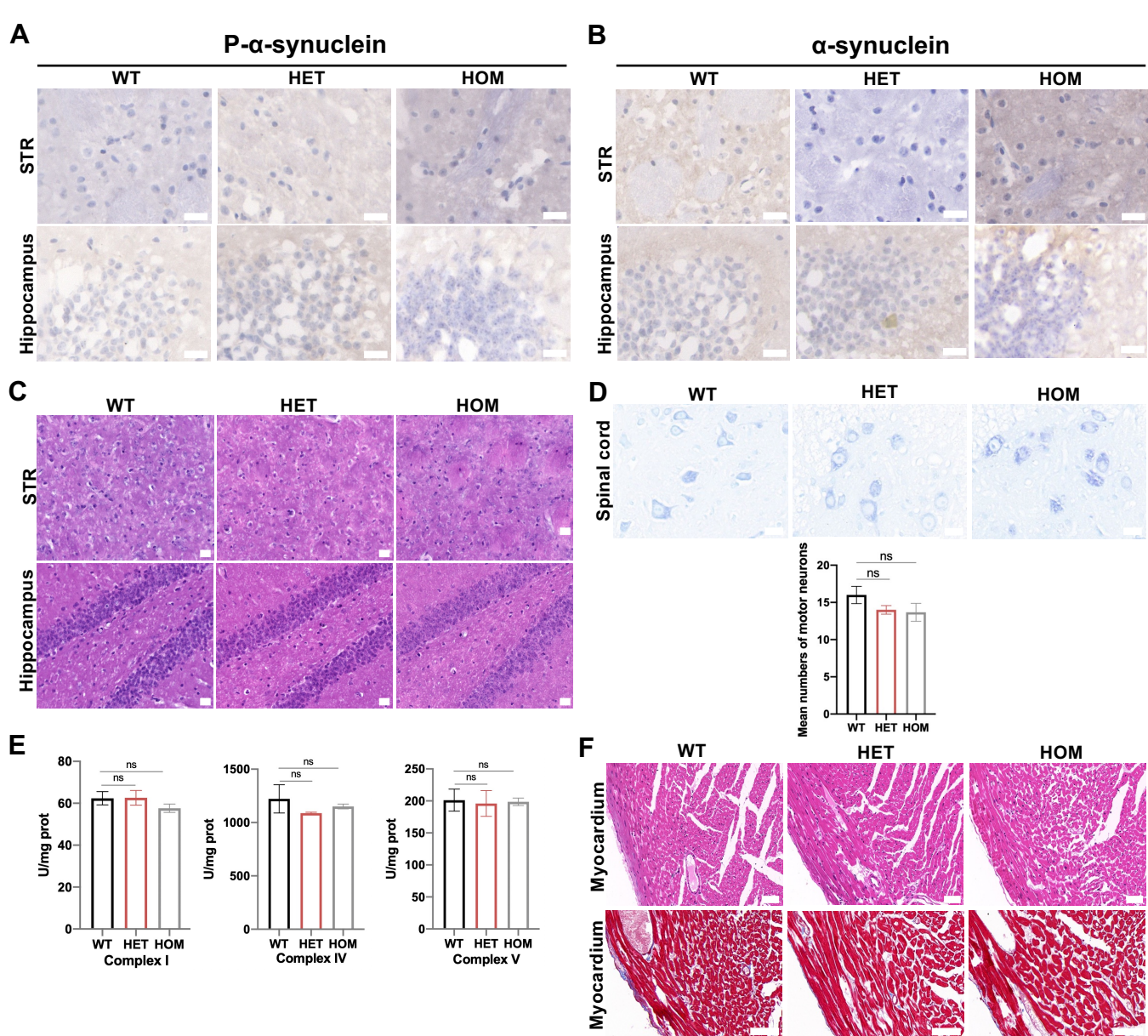


Figure S2. Pathological staining of the brain, spinal cords, and myocardium.

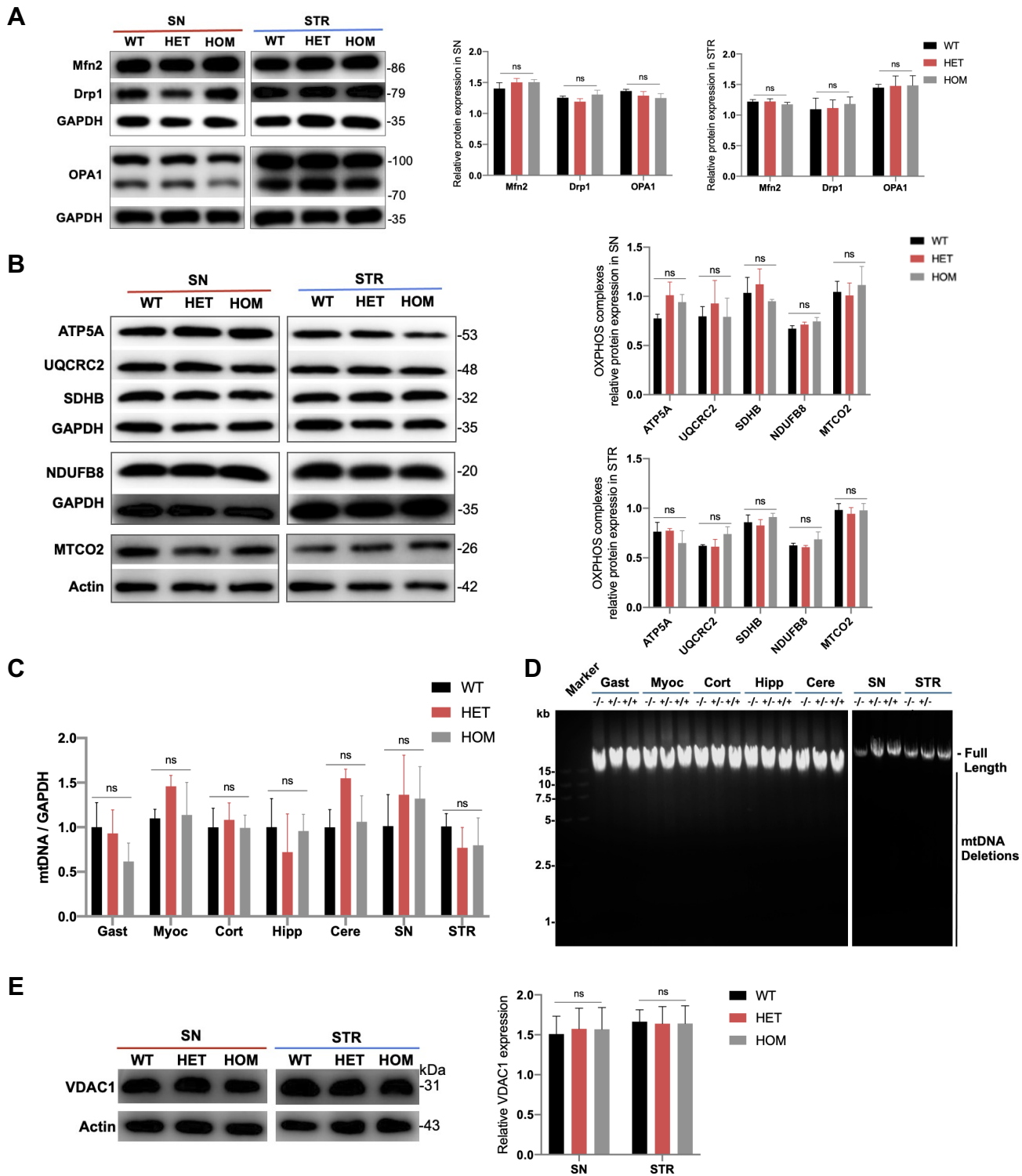
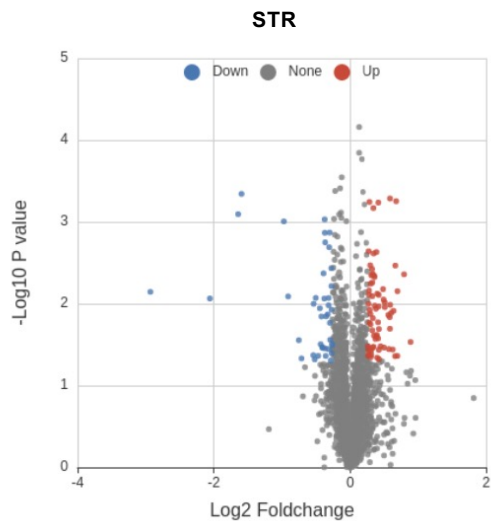
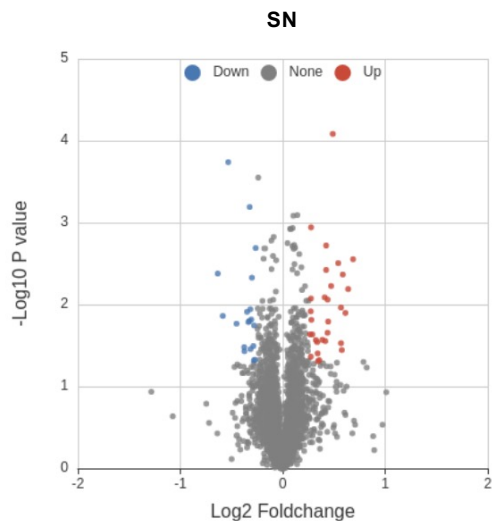
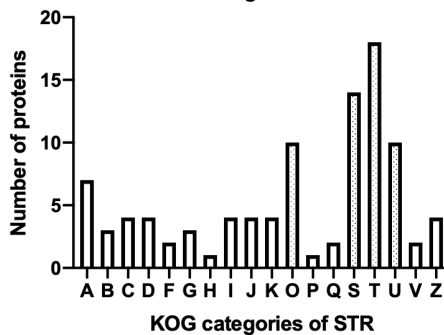
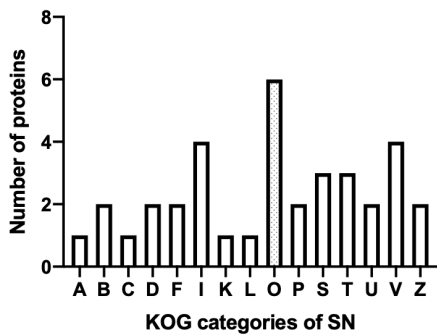


Figure S3. Electron transport chain and mtDNA unaffected by CHCHD2 p.T61I mutation.

A**B****C**

- [A] RNA processing and modification
- [B] Chromatin structure and dynamics
- [C] Energy production and conversion
- [D] Cell cycle control, cell division, chromosome partitioning
- [F] Nucleotide transport and metabolism
- [I] Lipid transport and metabolism
- [G] Carbohydrate transport and metabolism
- [H] Coenzyme transport and metabolism
- [J] Translation, ribosomal structure and biogenesis
- [K] Transcription
- [L] Replication, recombination and repair
- [O] Posttranslational modification, protein turnover, chaperones
- [P] Inorganic ion transport and metabolism
- [Q] Secondary metabolites biosynthesis, transport and catabolism
- [S] Function unknown
- [T] Signal transduction mechanisms
- [U] Intracellular trafficking, secretion, and vesicular transport
- [V] Defense mechanisms
- [Z] Cytoskeleton

Figure S4. Proteomics of HET and WT mice.

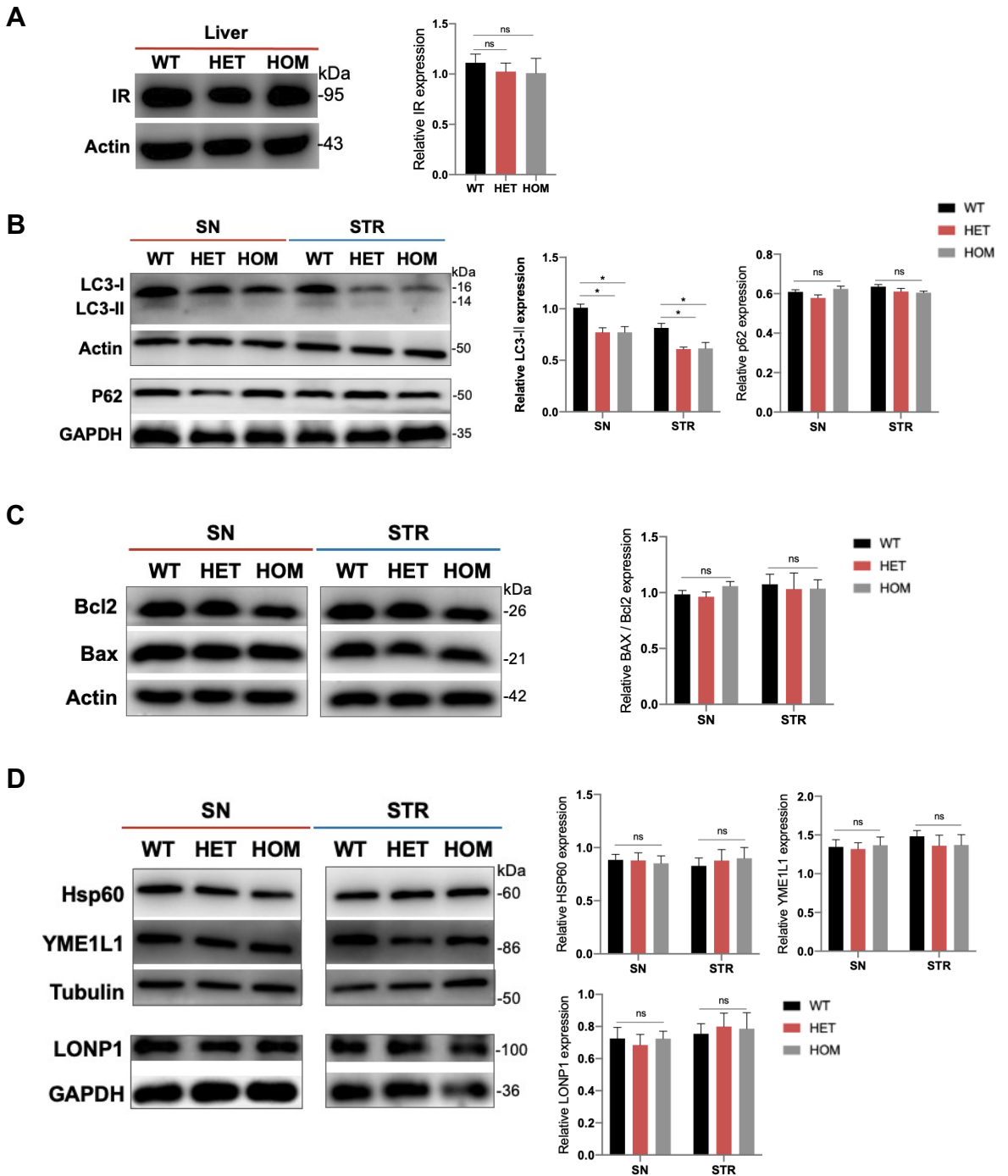


Figure S5. Studies on insulin-metabolism, autophagy, apoptosis, and mtUPR pathways.