Supplemental Data

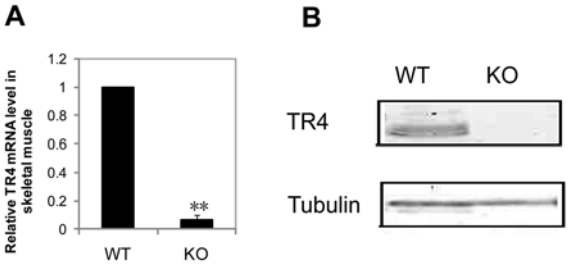

Fig 1. (A) mRNA and (B) protein extracted from skeletal muscle tissues from TR4 KO mice and their WT littermates were subsequently subjected to Q-PCR and Western blot. Modest levels of both mRNA and protein expression of TR4 can be detected in WT skeletal muscle tissues, while they are almost undetectable in TR4KO tissues. Three independent experiments were carried out and the mean ± SD was shown. **$p<0.01$.

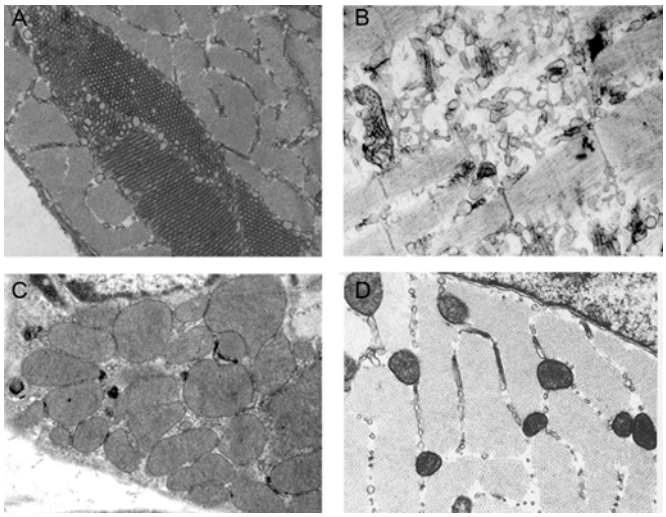

Fig 2. EM examination on TR4$^{-/-}$ soleus muscle revealed abnormal deposits of electron dense granules and tubular aggregates(TAs). (A) Transversely cut myofibers containing a group of dark interconnecting tubules showed ‘honeycomb’ appearance on cross section of TAs. (B) The dilation, segmentation and cystic swelling of the multiple terminal ends of TAs which are the precursors of SR cisternae. (C) Electron dense deposits were found in mitochondria mainly at the terminal cisternae of atrophied myofibers and longitudinal cisternae. Further investigation is needed to confirm the composition of the electron dense deposits. (D) Transversely cut myofiber nearby the A-I junction from soleus muscle of a TR4$^{-/-}$ mouse showed electron dense deposits in mitochondria and TAs.

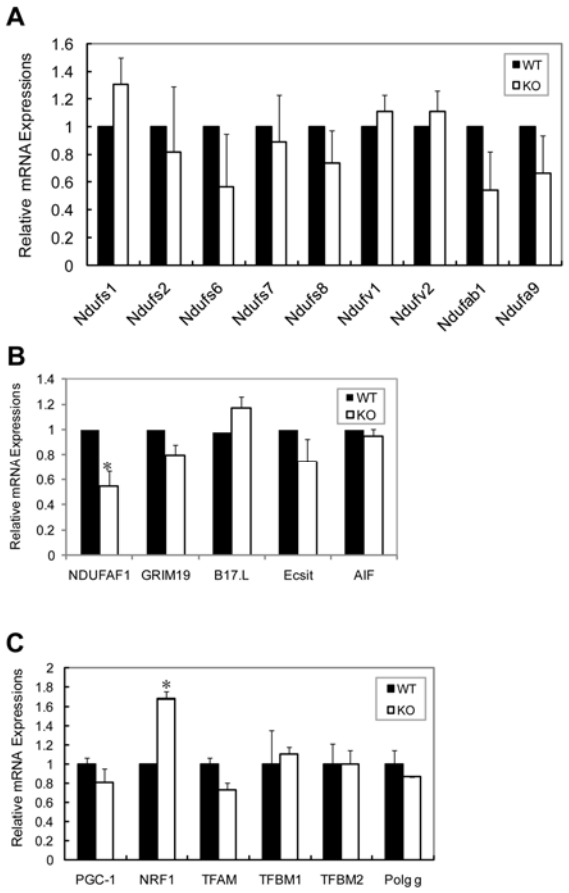

Fig 3. A reduction of complex I assembly factor NDUFAF1 level in *TR4*$^{-/-}$ mouse skeletal muscle tissue. (A) The expression level of complex I structural genes by Q-PCR. (B) The expression level of complex I assembly factors by Q-PCR. (C) The expression level of complex I genes responsible for mtDNA transcription. The mRNA was extracted from skeletal muscle tissues from *TR4*$^{+/+}$ and *TR4*$^{-/-}$ mice and subsequently subjected to Q-PCR. Three independent experiments were carried out and the mean±SD was shown for (A), (B), and (C). * $p < 0.05$.