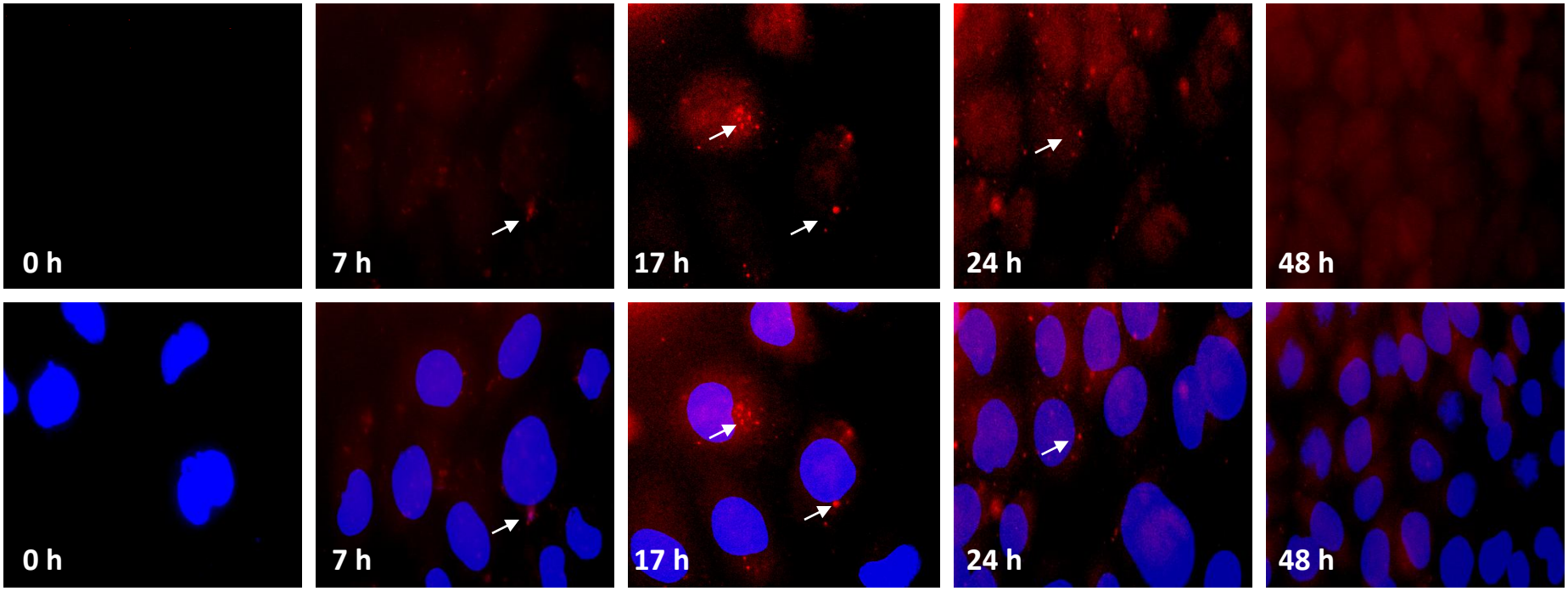
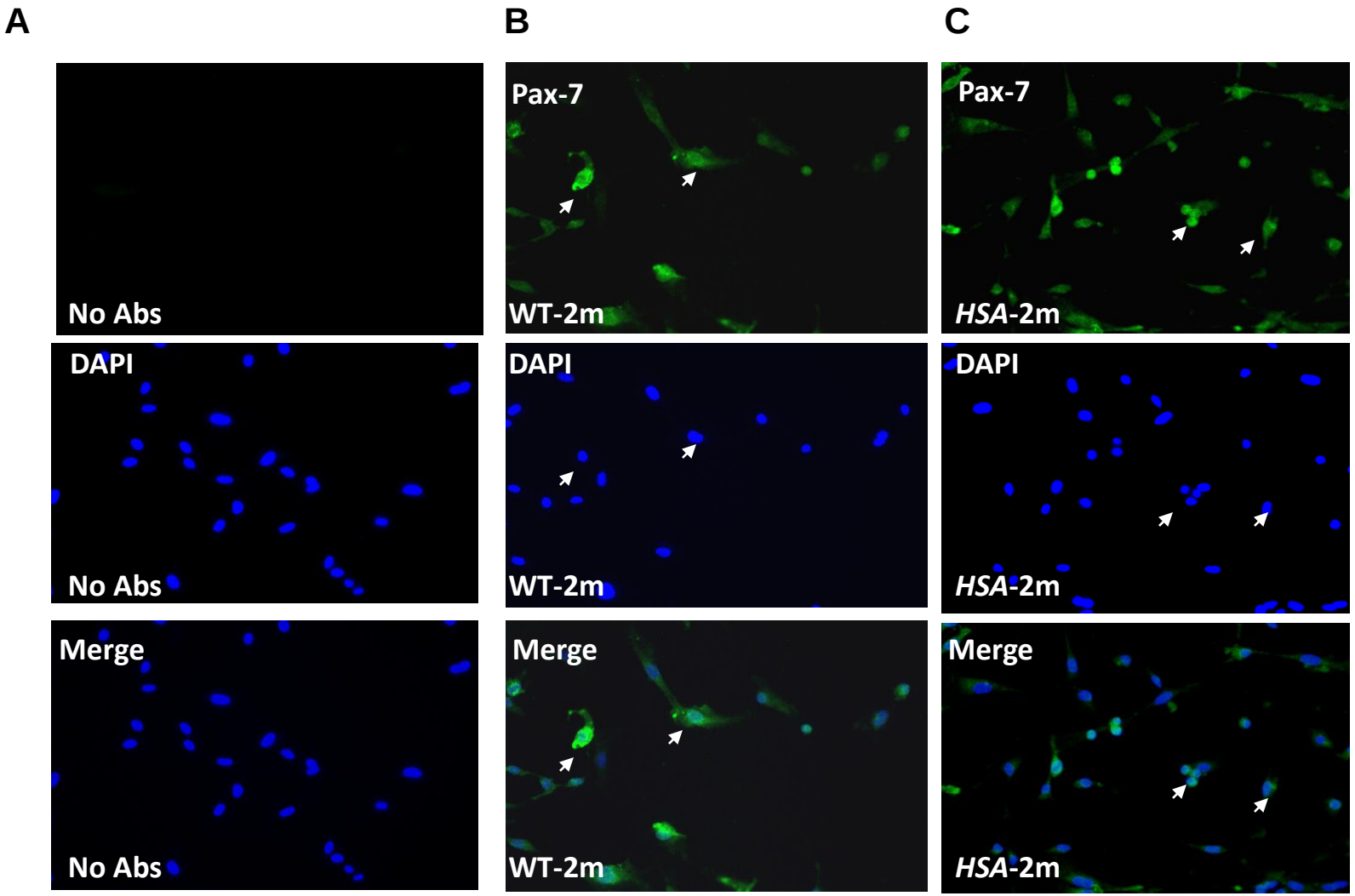
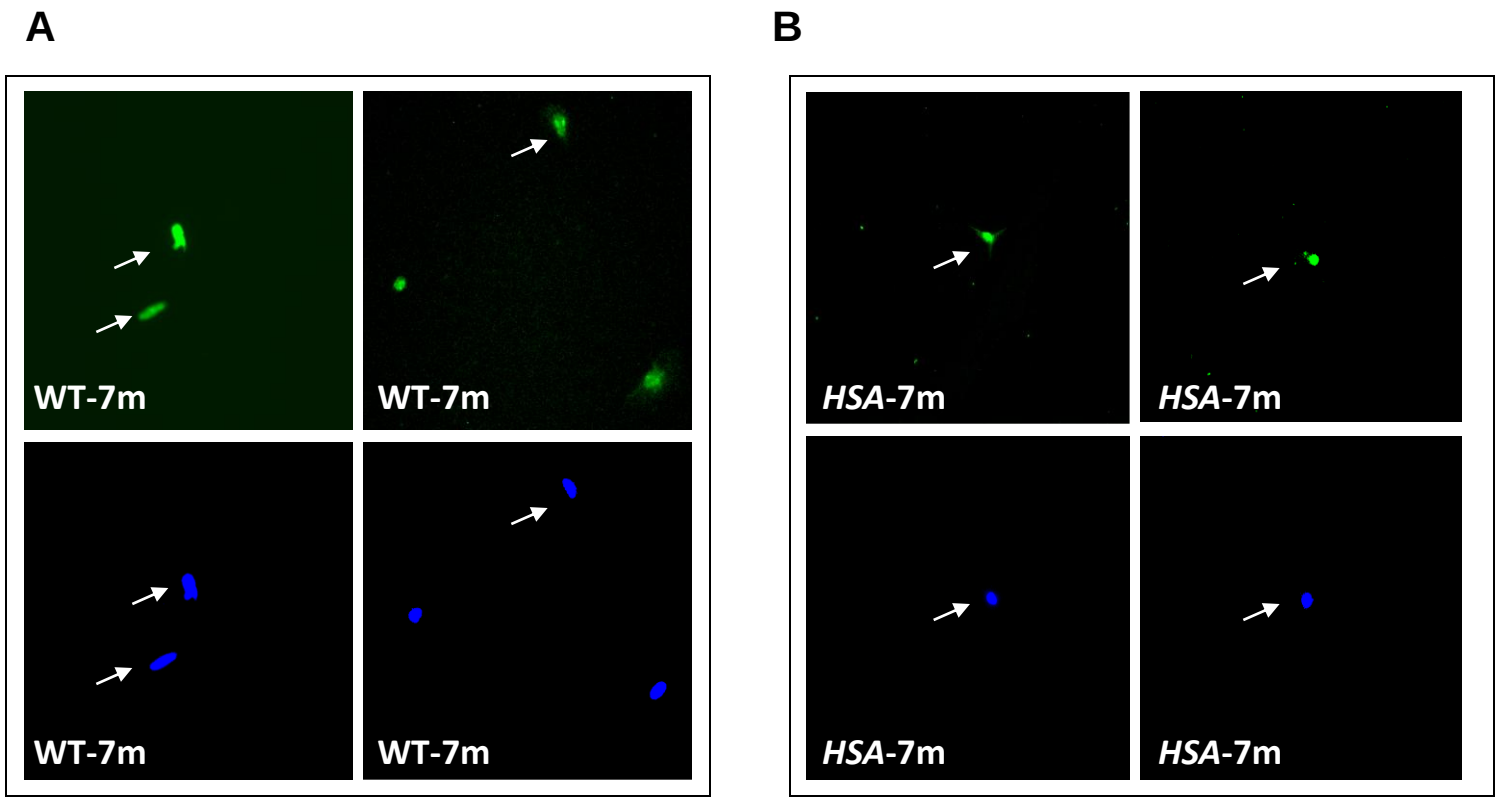
Supplementary Figures



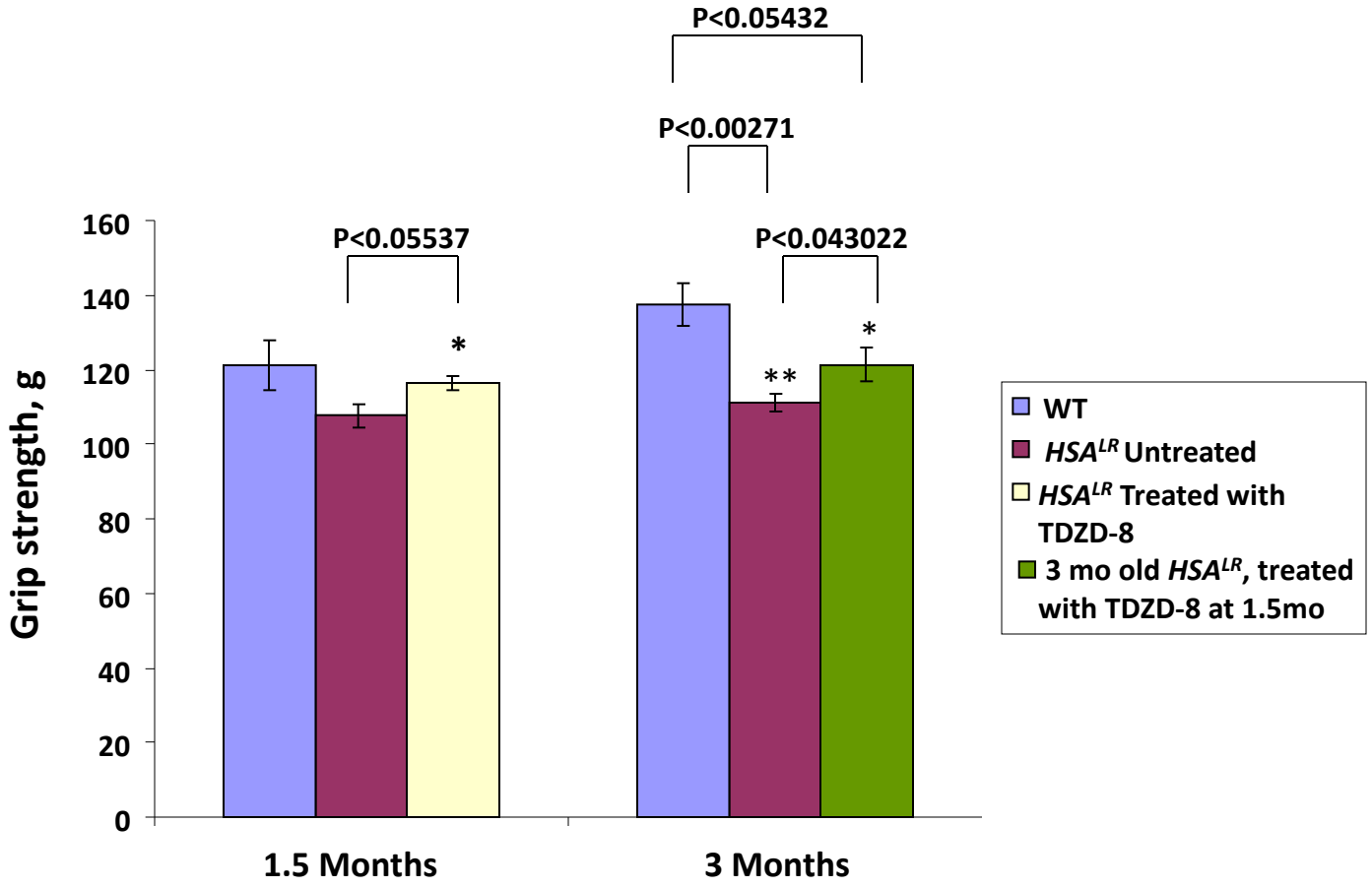
Supplementary Figure 1. Induction of CUG_{914} repeats in the double-stable monoclonal CHO cell model after Dox addition. **Upper panel**: CUG_{914} RNA was identified by FISH hybridization with CAG probe labeled with Alexa555 (red signal). Nuclei were stained with DAPI. Original magnification, x60. Images on the lower panel show merge of the CUG signal with DAPI. Numbers show the time points (hours) after Dox addition. The CUG RNA aggregates are shown by arrows. Note that the expression of CUG RNA at 7 hours after Dox addition is relatively weak. At 48 hours after Dox addition CUG RNA is partially degrading.



Supplementary Figure 2. Representative IF analysis images of the active satellite cells (shown by arrows) isolated from gastroc of 2 m old WT (**B**) and *HSA*^{LR} (**C**) mice with antibodies to Pax-7 (green). Negative control of IF analysis without antibodies to Pax-7 is shown on the panel **A**. Nuclei were stained with DAPI (blue). Original magnification, x20.



Supplementary Figure 3. Representative images of the IF analysis of the active satellite cells isolated from gastroc of 7 m old WT (**A**) and *HSA*^{LR} (**B**) mice with antibodies to Pax-7 (green). Two images for each mouse are shown. Arrows point to Pax-7-positive cells. Nuclei were stained with DAPI. Original magnification, x20.



Supplementary Figure 4. Adult (3 mo) *HSA*^{LR} mice, treated with TDZD-8 at young age (1.5 months), show improvement of muscle strength. *HSA*^{LR} mice at 1.5 months were treated with TDZD-8 (10 mg/kg) three times a week for 1 week. Grip strength was measured before the treatment, immediately after the treatment and in six weeks after the treatment. Untreated *HSA*^{LR} mice at 1.5 months show slight and not significant reduction of muscle strength. Treatment with TDZD-8 increased grip strength in young (1.5 mo) *HSA*^{LR} mice ($P<0.05537$). Un-treated *HSA*^{LR} mice at 3 mo of age show a reduction of grip strength ($P<0.00271$); whereas *HSA*^{LR} mice, which were treated at 1.5 mo of age and analyzed at 3 mo of age show an improvement of grip strength ($P<0.05432$). Six mice per group were used. The standard errors of the means (SEM) are shown.