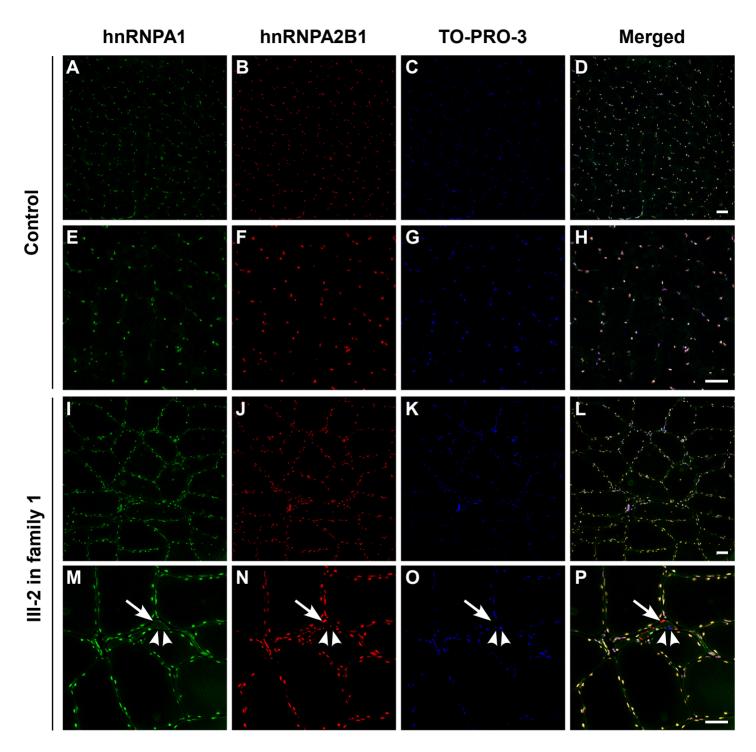
## **Supplementary Figure e-1**

Multiple immunohistochemistry for hnRNPA1 and A2B1



Representative microphotographs of transverse cryosections from the specimens of patient III-2 in family 1 and a control. A–D and E–H: In the control sections, hnRNPA1-like immunoreactivity was found exclusively in myonuclei, and it was colocalized with A2B1. I–L and M–P: In almost all of the fibers of the patients, myonuclei retained both hnRNPA1 and A2B1; however, nuclear depletion (arrowheads) and differential aggregation of the hnRNPs (arrows indicate A2B1) were rarely observed in exceedingly atrophied fibers (M–P). Bars = 50  $\mu$ m