



**Figure S2.** hIF1 is localized in mitochondria. Upper panels, HeLa cells were transfected with the pCMV-SPOR6-H49K plasmid and processed for immunofluorescence microscopy for hIF1 (red) and  $\beta$ -F1-ATPase ( $\beta$ -F1, green). Immunohistochemistry for hIF1 (red) and  $\beta$ -F1-ATPase ( $\beta$ -F1, green) in brain cortex of control (wt, middle panel) and  $H^+/T^+$  (lower panel) mice. Magnification 40x (upper panel) and 20x (middle and lower panel). Note that hIF1 is only expressed in neurons of  $H^+/T^+$  mice whereas  $\beta$ -F1-ATPase is present in glia and neurons of both phenotypes.