Supplemental Data

Frontotemporal dementia nonsense mutation progranulin rescued by aminoglycosides

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Supplemental Figure S1. Gentamicin did not induce readthrough of progranulin PTC mutations Q125X or Y229X or FUS PTC mutation R495X. (A) Gentamicin had no readthrough effect on two other FTD mutations Q125X and Y229X. N2A cells were transfected with Q125X or Y229X progranulin, allowed to recover, and treated with two different concentrations of each compound for 24 hours. Cell lysates were generated, separated by SDS-PAGE, and analyzed by Western analysis using anti-FLAG (top), anti-progranulin (middle), and anti-actin (lower) antibodies. In the anti-progranulin blot, the higher band is WT progranulin and the lower band is the Q229Y truncated protein. (B) Gentamcin had no readthrough effect on the FUS R495X mutation responsible for familial ALS. N2A cells were transfected with WT or R495X FUS, allowed to recover, and treated with three concentrations of G418 for 24 hours. Cells were harvested and cell lysates were subjected to SDS-PAGE and Western analysis using anti-
FLAG (top) and anti-actin (lower) antibodies. The slightly higher band is full-length WT FUS and the lower band is R495X truncated FUS. Gentamicin did not induce readthrough of R495X FUS.