Expanded View Figures

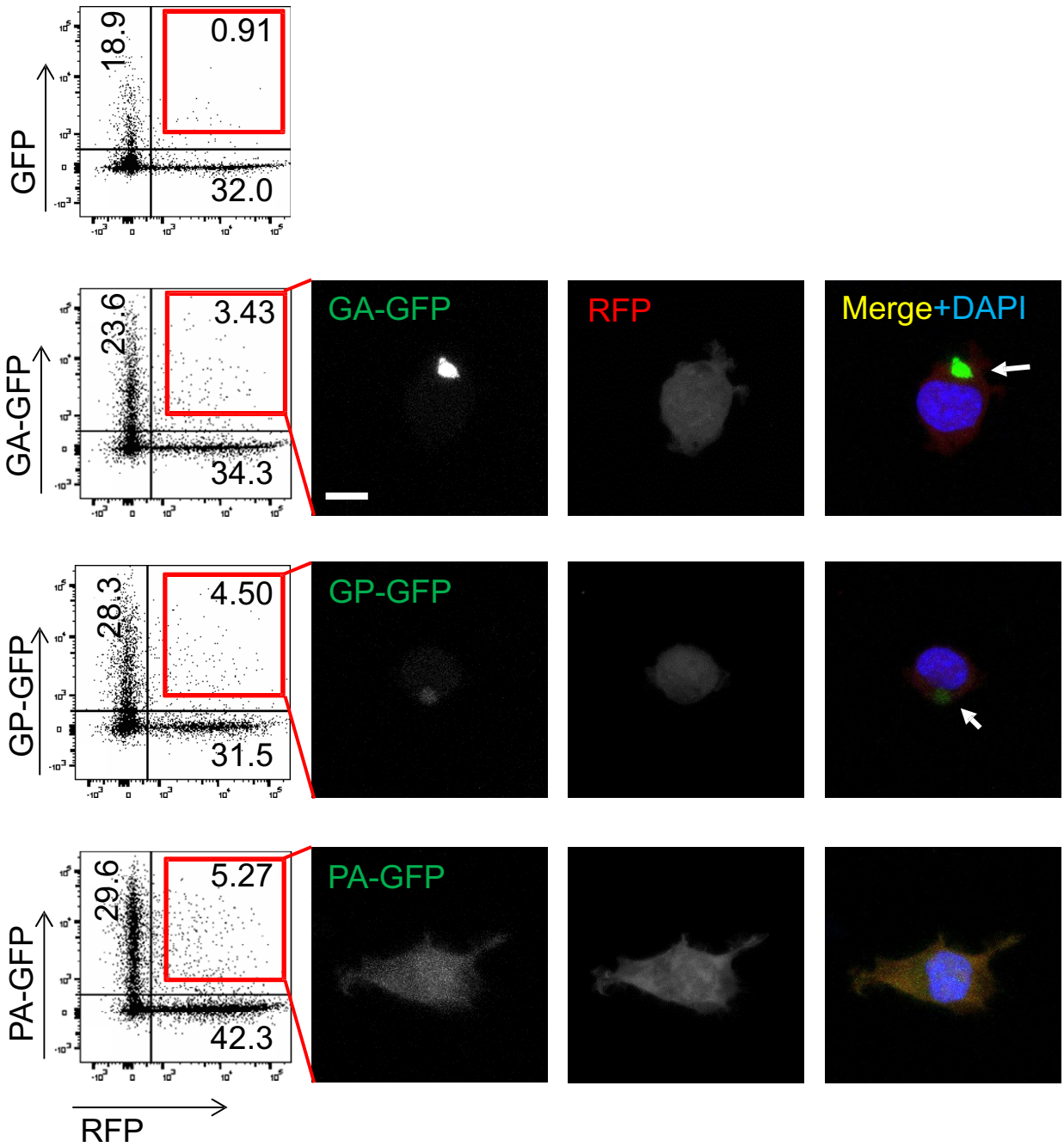

Figure EV1. Transmission of hydrophobic DPR proteins in a co-culture assay.
HEK293 cells were transfected with RFP, GFP, or DPR-GFP for 24 h and mixed in the indicated combination for additional 24 h as in Fig 2 before cell sorting by flow cytometry. Gating was performed on RFP-expressing cells vs. mixture of all green fluorescent cells. The fraction of double-positive cells is indicated in percent. Double-positive cells were sorted and plated on poly-D-lysine-coated coverslips and imaged 17 h later. Images show uptake of DPR-GFP into RFP-positive cells. Arrows indicate co-localization of GA_{175}-RFP aggregates with GA_{175}-GFP and GP_{47}-GFP. Scale bar 10 μm.

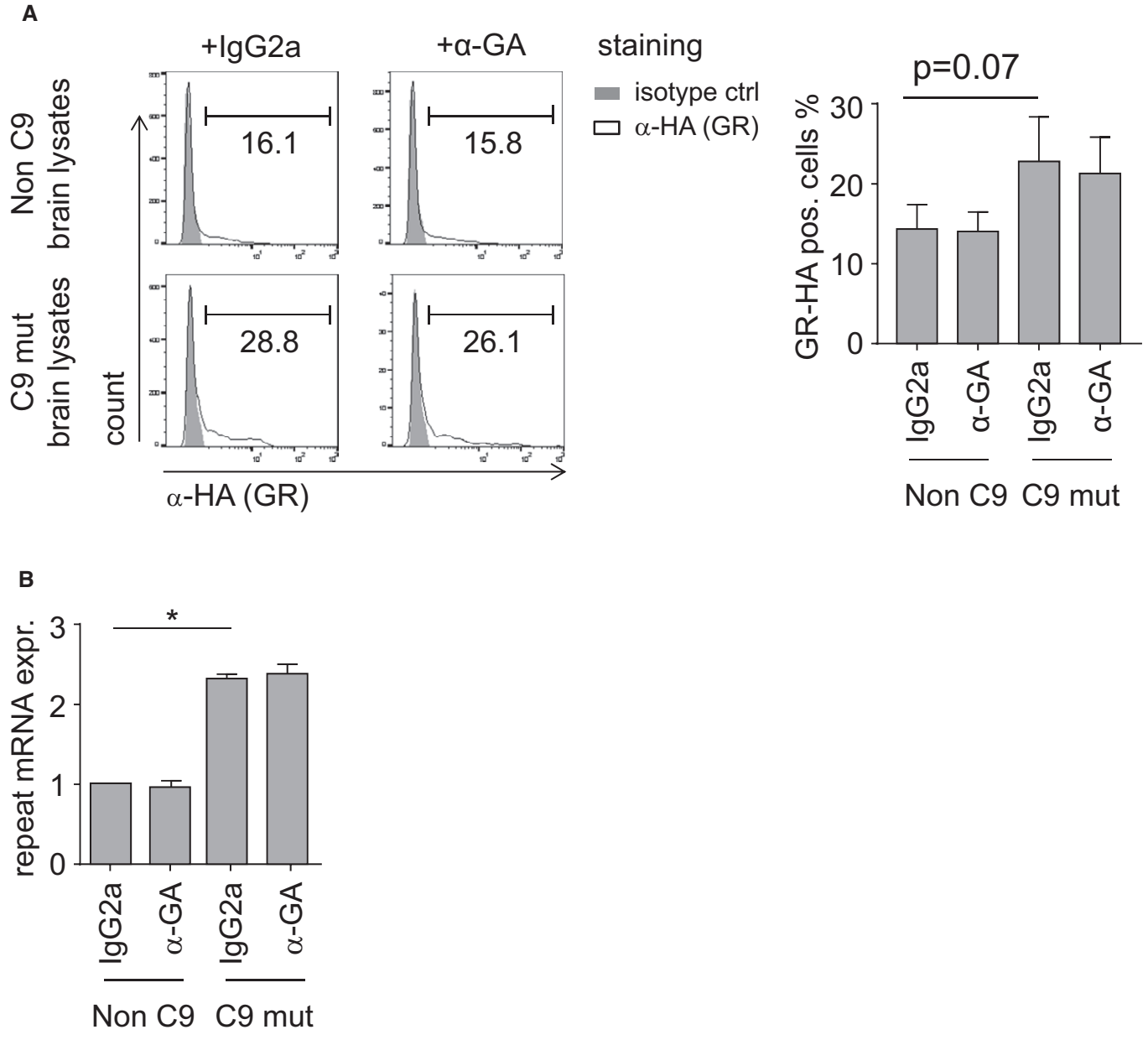

Figure EV2. Anti-GA antibodies do not reduce expression of poly-GR and repeat RNA.

HEK293 cells transfected with $(G4C2)_{80}$ were treated with cerebellar extracts pre-incubated with anti-GA or isotype control.

A The fraction of RAN translation-derived GR_{80}-HA was quantified by flow cytometry. Data indicated the means ± SD of $n = 3$ patients and controls in independent experiments. Statistics were performed by one-way ANOVA with Dunnett's multiple comparisons test.

B Quantitative RT–PCR shows repeat RNA transcripts upon treatment with cerebellar extracts pre-incubated with anti-GA or isotype control. Data are shown as mean ± SD from $n = 3$ patients and controls in independent experiments. Statistics were performed by one-way ANOVA with Dunnett's multiple comparisons test; non-C9 + IgG2a vs. C9 mut + IgG2a $P = 0.0168$; $^*P < 0.05$.