Table S1. List of gRNAs and primers for *NEAT1* knockdown HEK293 cells.

NEAT1 gRNA up fwd	CACCGCGAAAGTCACGCGCGCCTCC
NEAT1 gRNA up rev	AAACGGAGGCGCGCGTGACTTTCGC
NEAT1 gRNA down fwd	CACCGCCAGACCTGGACGCTCCACC
NEAT1 gRNA down rev	AAACGGTGGAGCGTCCAGGTCTGGC
NEAT1 det primer fwd	GTTGTCACCCACTAGCTCCT
NEAT1 det primer rev	AAGTCCAAAAGGAGCACTGC

Table S2. shSFPQ and shScramble hairpin sequences.

shScramble	CCTAAGGTTAAGTCGCCCTCGCTCGAGCGAGGGCGACTTAACCTTAGG	Addgene	#1864
shSFPQ	CCGGCGGTTGTTTGTTGGGAATCTACTCGAGTAGATTCCCAACAAACAACCGTTTTT	Sigma Aldrich	NM_005066.x-977s1c1

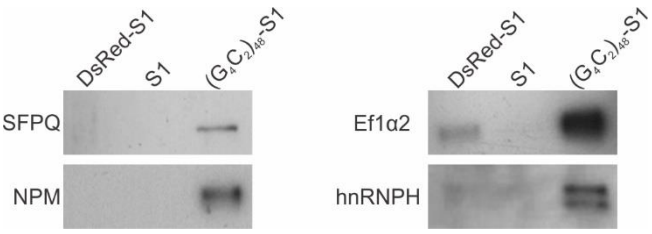

Fig. S1. Immunoblots confirming SFPQ, NPM1, EF1α2 and hnRNPH specifically co-precipitate with $(G4C2)_{48}$-S1 RNA in rat cerebellar nuclear extracts.

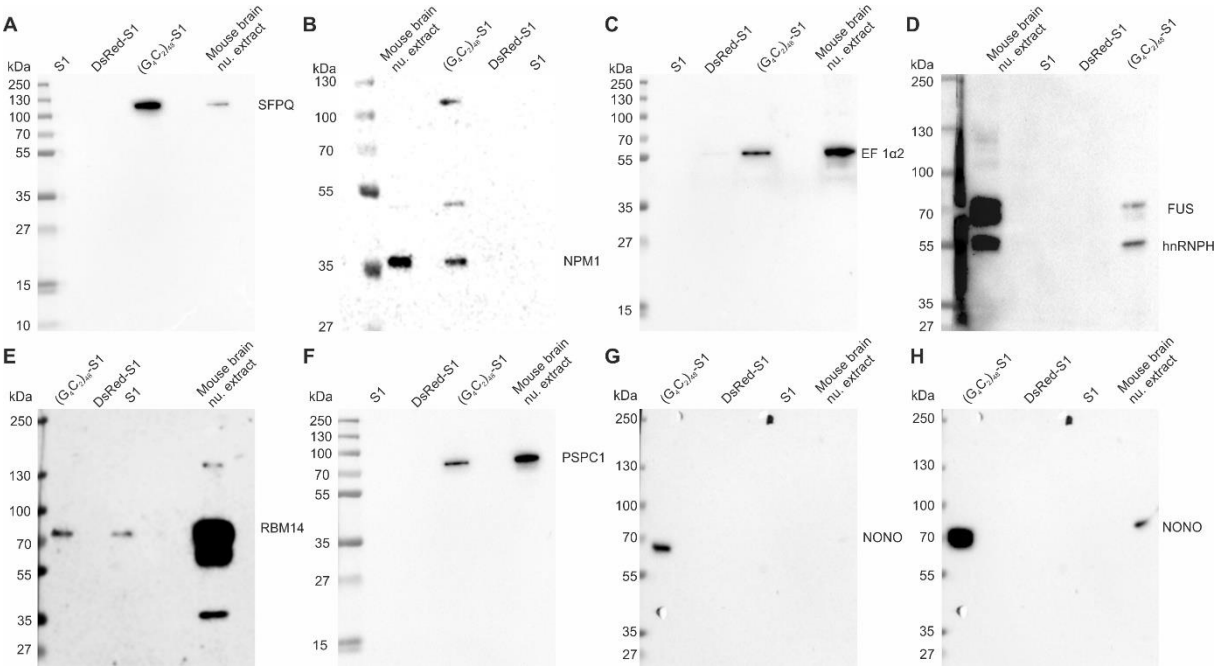

Fig. S2. Full length blots from Fig. 1.

(A, B, C, D, E, F, G, H) Full length immunoblots with positive control confirming that proteins SFPQ, NPM1, EF1α2, hnRNPH, FUS, RBM14, PSPC1 and NONO from mouse brain nuclear extracts specifically co-precipitate with $(G_4C_2)_{48}$-S1 RNA.

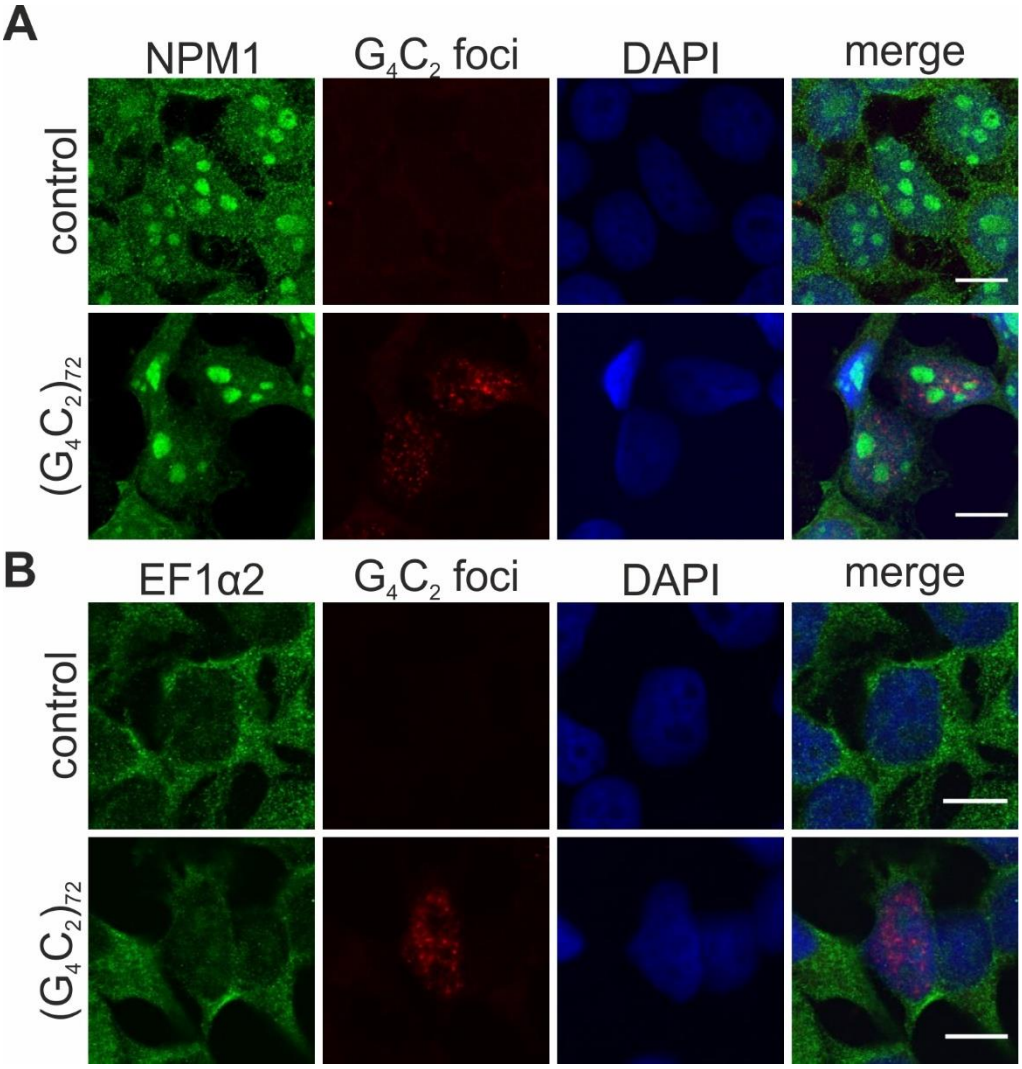

Fig. S3. NPM1 and EF1α2 do not colocalize with $(G_4C_2)_{72}$ nuclear foci.

HEK293T cells transfected with a plasmid expressing $(G_4C_2)_{72}$ repeats and probed for $(G_4C_2)_{72}$ and NPM1 (A) or EF1α2 (B). Scale bars: 10 µm.

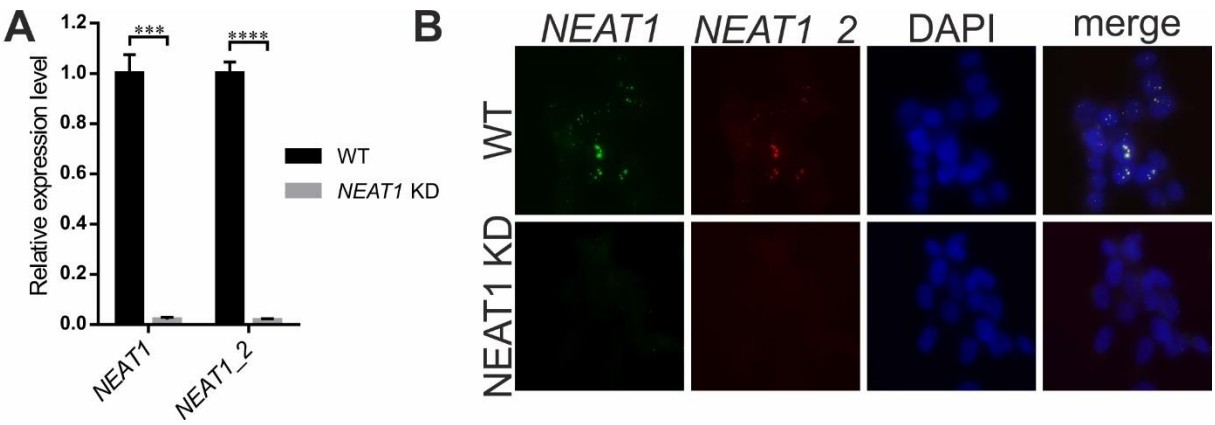

Fig. S4. Characterization of *NEAT1* knockdown HEK293T cells.

(A) Relative expression levels of *NEAT1* and *NEAT1_2* in *NEAT1* knockdown HEK293T cells (*NEAT1* KD) compared to wild-type HEK293T cells (WT) determined by quantitative RT-PCR (t-test, ***p<0.001 for *NEAT1* and ****p<0.0001 for *NEAT1_2*; three experiments were performed; data are presented as mean ± s.e.m). (B) *NEAT1* knockdown HEK293T and wild-type HEK293T cells probed for *NEAT1* and *NEAT1_2*.

(B)

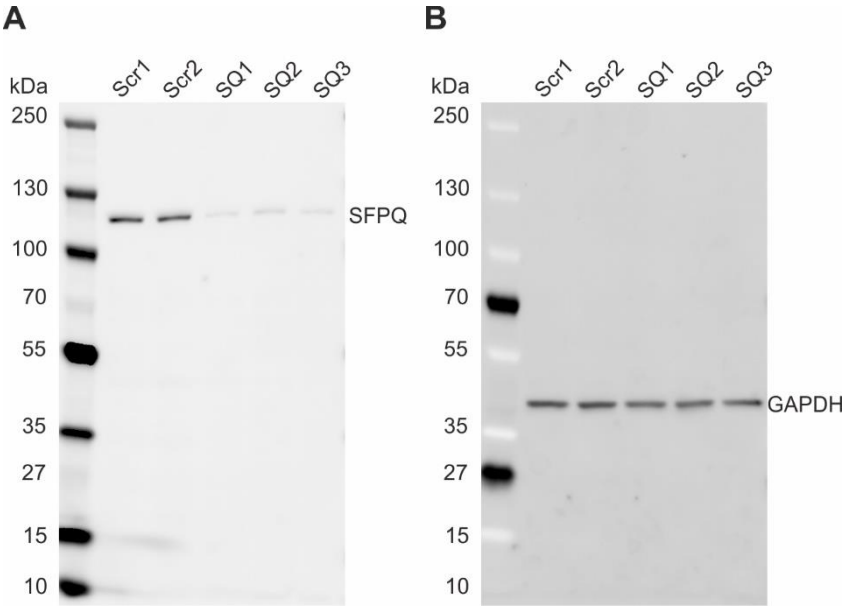

Fig. S5. Full length blot from Fig 5.

(A) Full length blot from Fig. 5 for quantification of SFPQ knockdown in *C9ORF72* mutant fibroblasts transduced with lentiviral particles containing sequences for scrambled shRNA or SFPQ shRNA. (B) GAPDH was used as loading control. Signal was detected using dual-colour imaging system.