Supplementary Figure 1. Specificity of the hnRNP A1B antibody. (A) HeLa nuclear extracts were immunoblotted with the following antibodies: hnRNP A1 (9H10, Abcam) or hnRNP A1B (custom) and Lamin B1. The custom antibody has an ~8-fold preference for hnRNP A1B over hnRNP A1. (B) Lysates from CB3 cells expressing hnRNP A1 or hnRNP A1B were immunoprecipitated with hnRNP A1B (custom) antibody and immunoblotted for hnRNP A1B (custom), hnRNP A1 (9H10). Lysates were also blotted with actin as loading control. (C) HeLa cells were transfected with Myc-hnRNP A1 or A1B cDNA. Cells were co-labeled for Myc and hnRNP A1B. Scale bar, 25  $\mu$ m. (D) Low magnification images of hnRNP A1B in ALS patient motor neurons, detected with antibody hnRNP A1B (*left*) or hnRNP A1B plus blocking peptide (*right*). Low magnification scale bar, 100  $\mu$ m. Higher magnification image is from boxed region in lower magnification image. Scale bar, 10  $\mu$ m.



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**Supplementary Figure 2.** *In silico* analysis of fibrillization propensity of hnRNP A1 and hnRNP A1B. *In silico* analysis using ZipperDB. The amino acid residues are represented on the x-axis. For clarity, the analysis was trimmed to start at residue 208. Each bar of the histogram represents a hexapeptide and is colored according to its Rosetta energy. All bars in orange and red exhibit Rosetta energies less than -23 kcal /mol, the point at which they are prone to form fibrils. Residues encoded by exon 7B are shaded in red.





## Supplementary Table 1: Details of ALS and control cases.

Case	Sex	Age	Known mutation
number			
sALS-1	F	85	none
sALS-2	М	66	none
sALS-3	М	39	none
sALS-4	М	78	ATXN2 (32 repeats)
sALS-5	F	65	C9 expansion
sALS-6	М	55	none
fALS-1	М	61	FUS
fALS-2	М	58	C9 expansion & RGNEF
fALS-3	F	64	C9 expansion
Ctrl-1	М	55	
Ctrl-2	М	61	
Ctrl-3	М	33	
Ctrl-4	М	67	
Ctrl-5	М	61	