

Supplementary Table 2

Case Number	ALS associated mutation or Control	aa131-153 Ra-ab	aa57-72 Ra-ab
1	<i>C9orf72HRE</i>	++	++
2	<i>C9orf72HRE</i>	+	++
3	<i>C9orf72HRE</i>	++	++
4	<i>C9orf72HRE</i>	++	+++
5	<i>C9orf72HRE</i>	+++	+++
6	<i>C9orf72HRE</i>	++	++, +++
7	<i>C9orf72HRE</i>	+, ++	++
8	<i>C9orf72HRE</i>	++, +++	++
9	<i>C9orf72HRE</i>	++	++
11	<i>C9orf72HRE</i>	++, +++	+++
13	<i>C9orf72HRE</i>	++	++
14	<i>C9orf72HRE</i>	+	++
15	<i>C9orf72HRE</i>	+	+
16	<i>C9orf72HRE</i> + <i>KIF5A</i>	++, +++	+++
17	<i>C9orf72HRE</i> + <i>KIF5A</i>	+, ++	++
18	<i>C9orf72HRE</i> + <i>KIF5A</i>	++	++
19	<i>KIF5A</i>	++	++
21	<i>KIF5A</i>	+, ++	+
22	<i>KIF5A</i>	++	+, ++
23	<i>KIF5A</i>	+	++
24	<i>KIF5A</i>	++	++
25	<i>NEK1 R812X</i>	++, +++	++
26	<i>VAPB S160delS</i>	++	++
27	<i>FUS R495X</i>	++	++
28	<i>FUS Q23L</i>	++, +++	++, +++
29	<i>ALSIN</i> + <i>KIF5A</i>	++	++
31	<i>SOD1 A4V</i> + <i>KIF5A</i>	++	++
32	<i>SOD1 G127X</i>	n.a.	+
36	<i>SOD1 G114A</i>	++, +++	++
37	<i>SOD1 D76Y</i>	++	++
38	<i>SOD1 D83G</i>	+	+, ++
39	<i>SOD1 D101G</i>	++	+, ++
41	<i>SOD1 D90A</i>	++	++
42	<i>SOD1 D90A</i>	++	+++
43	<i>SOD1 D90A</i>	++, +++	++, +++
44	<i>SOD1 D90A</i>	++	++
45	<i>SOD1 D90A</i>	++	++, +++
46	<i>SOD1 D90A</i>	++, +	++
47	N.D. Control	-	-
48	N.D. Control	+	+
49	N.D. Control	+	+
50	N.D. Control	++	++

51	N.D. Control	-	-
52	N.D. Control	+	+
53	N.D. Control	-	-
54	N.D. Control	-	-
55	N.D. Control	-	-
56	N.D. Control	-	-
57	Non-neurol. control	-	-
58	Non-neurol. control	-	-
59	Non-neurol. control	-	-
60	Non-neurol. control	+	+
61	Non-neurol. control	-	-
62	Non-neurol. control	-	-
63	Non-neurol. control	-	-
64	Non-neurol. control	-	-
65	Non-neurol. control	-	-
66	Non-neurol. control	0+, +	+
67	Non-neurol. control	-	-
68	Non-neurol. control	-	-
69	Non-neurol. control	-	-
70	Non-neurol. control	-	-
71	Non-neurol. control	-	-
72	Non-neurol. control	-	-
73	Non-neurol. control	-	-
74	Non-neurol. control	-	-
75	Non-neurol. control	-	-
76	Non-neurol. control	-	-

Supplementary Table 2: Comparative analysis of ALS/FTD and controls using the anti-SOD1 peptide antibodies aa131-153 and aa57-72 Ra-ab stained in parallel and examined independently by two neuropathologists (KF, TB) in a blinded set-up fashion. In cases when the two examiners assessed the sections differently, both ratings are shown.
