Supplementary Table 1: Pre-application of riluzole increases its effect on I_{ACh} amplitude and decay.

	ε-AChR		γ-AChR	
Pretreatment (s)	Amplitude (%)	τ_{decay} (%)	Amplitude (%)	τ_{decay} (%)
0	100 ± 3 (n=7)	83 ± 4	92±5 (n=7)	97±2
0.5	87, 81	70, 79	78±3 (n=10)*	100±5
1	NA	NA	67±2 (n=10)[#]	81±2[#]
5	88±6 (n=4)	89±3*	54±5 (n=9)[#]	74±3[#]
10	93±7 (n=5)	80±3[#]	55±5 (n=8)*	75±3*
30	86±3 (n=5)*	71±4[#]	72±4 (n=14)[#]	75±3[#]
30, +GDPβS	84±5 (n=5)*	69 ± 4*	70±10* (n=4)	63±2[#]
30, Riluzole 50 μM	62 ± 4 (n=4)[#]	49± 1[#]	41±4 (n=5)[#]	55±6[#]
60	NA	NA	69±5 (n=12)[#]	73±2[#]
120	97 ± 3 (n=6)	79 ± 8 *	69±6 (n=8)[#]	73±3[#]

Data are mean ± SEM values (number of cells tested) of the amplitude and decay time constant (t_{decay}) of whole-cell currents elicited by ACh (100 μM at -60 mV) in HEK cells expressing the indicated AChR type. Values obtained in the continuous presence of riluzole (0.5 μM unless otherwise noted), preapplied for the indicated amount of time, are expressed as percent of the values obtained under control conditions (ACh alone) in each cell. Comparison of data obtained with riluzole *vs.* control values was performed using paired Student's *t* test; *: P<0.05; #: P < 0.005,

Supplementary Table 2: Open channel distribution in cell-attached recordings

	HEK cells		ALS myotubes	
	τ_1 (ms) (a$_1$) (%)	τ_2 (ms) (a$_2$) (%)	τ_1 (ms) (a$_1$) (%)	τ_2 (ms) (a$_2$) (%)
Control	1.4 ± 0.1 (n=6) (33 ± 4)	7.0 ± 0.7 (67 ± 4)	1.2 ± 0.1 (n=5) (34 ± 2)	9.4 ± 0.5 (66 ± 3)
riluzole	1.4 ± 0.2 (n=7) (43 ± 4)	7.6 ± 0.8 (57 ± 4)	2.0 ± 0.5 (n=7) (38 ± 5)	12.2 ± 0.9 (62 ± 5)

τ_1 (a$_1$) and τ_2 (a$_2$): time constants (weight) of the two exponential components best fitting the distribution of channel open durations, in HEK cells transfected with γ-AChR or in myotubes derived from satellite cells of ALS patients. Data are given as mean $\pm$ S.E.M. for the indicated number of patches, each exposed to ACh (100 nM, -80 mV) alone or to ACh plus riluzole (0.5 μM). In both cell types, values obtained with riluzole were not significantly different (P > 0.06) from the corresponding values in control recordings.