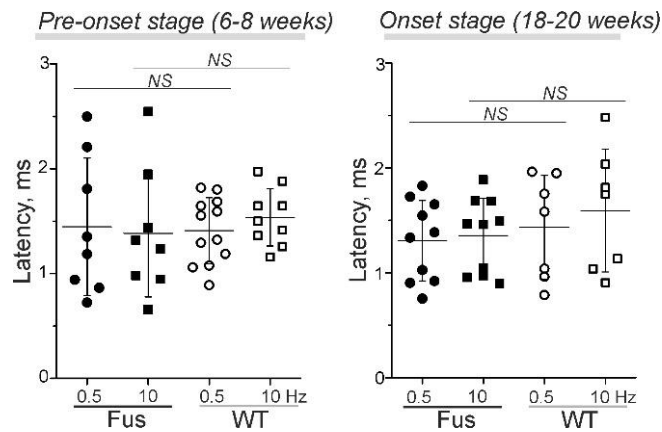
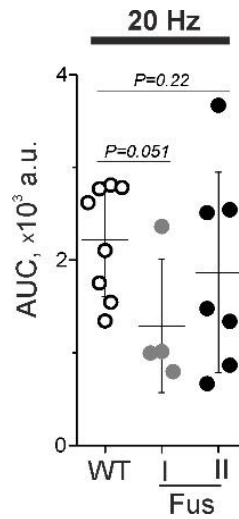


**Supplementary Figure S1.** A-Measurements of nAChR area (in  $\mu\text{m}^2$ ), assessed as  $\alpha\text{-Btx}$  positive regions. Shown the values of individual NMJs from 6 mice *per* group. \*\*\* $P < 0.001$  by Kolmogorov-Smirnov test between NMJs from FUS and WT animals. B – typical fluorescent images of NMJs labeled with  $\alpha\text{Btx}$  and anti-synaptophysin (Sph) antibody. Scale bars – 20  $\mu\text{m}$ . C – Quantification of synaptophysin fluorescence in motor nerve terminals.  $n = 5$  mice per group (13-15 NMJs from each muscle). \* $P < 0.05$  by Mann-Whitney U test compared to WT. 6-8-week-old and 18-20-week-old mice with  $\Delta\text{FUS}(1-359)$  represent animals at pre-onset and onset stages.



**Supplementary Figure S2.** Quantification of latency (interval from stimulus artefact to presynaptic action potential) in the motor axon of  $\Delta\text{FUS}(1-359)$  mice *vs* WT mice. Shown comparison of the latency at 0.5 and 10 Hz nerve stimulation in  $\Delta\text{FUS}(1-359)$  mice at pre-onset and onset stages as compared to age-matched WT animals.  $n = 8-12$  muscles from individual mice *per* group; 300 action potentials were analyzed in each muscle.



**Supplementary Figure S3.** Area under curve of intraterminal  $\text{Ca}^{2+}$  elevation upon one-minute stimulation at 20 Hz. Shown data from  $\Delta\text{FUS}(1-359)$  mice at pre-onset (I; 6-8 week-old) and onset (II; 18-20 week-old) stages as compared to WT (6-8 + 18-20 week-old) mice. Nerve terminal  $[\text{Ca}^{2+}]_{\text{in}}$  dynamics are shown in Fig. 7B.  $n=4-8$  muscles from individual mice per group (WT: 36 NMJs of 8 mice; FUS I: 18 NMJ of 4 mice; 22 NMJs of 7 mice).  $P_{\text{asympt.}}=0.051$  ( $P_{\text{exact}}=0.049$ ),  $P_{\text{asympt.}}=0.224$  by Mann-Whitney  $U$  test.