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Overexpression of human wild-type FUS causes progressive motor neuron degeneration in an age and dose-dependent fashion

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Supporting Information

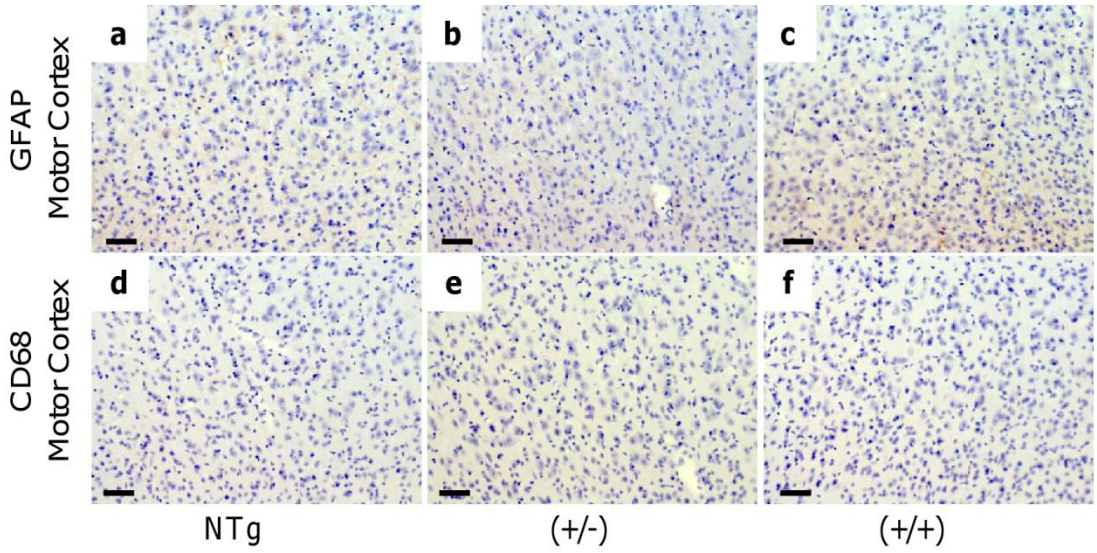

Figure S1 hFUS overexpression does not alter neuroinflammation in the cortex. (a-c) No changes in astrogliosis, detected using GFAP immunohistochemistry (d-f) or microgliosis, detected using CD68 immunohistochemistry, were detected in hFUS (+/-) or (+/+) animals (scale bar: 20µm)

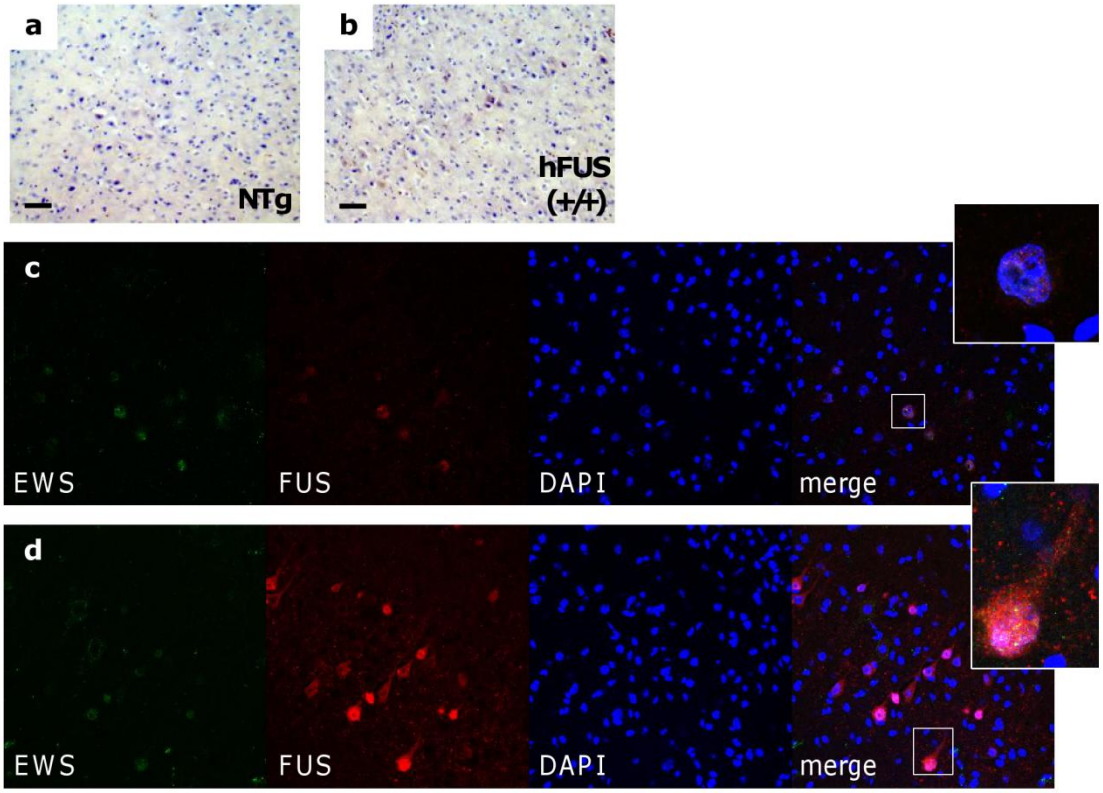

Figure S2 FUS does not colocalise with EWS or TAF15 in the cortex of hFUS (+/+) mice.
(a-b) No significant TAF15 staining was detected in NTg (A) or hFUS (+/+) (b) mice. (c-d) Low level EWS staining was detected in the nucleus of FUS positive cells in both NTg (c) and hFUS (+/+) (d) mice. However, total EWS levels did not appear to differ between NTg and hFUS (+/+) mice, and there was no evidence of FUS and EWS cytoplasmic colocalisation in hFUS (+/+) animals

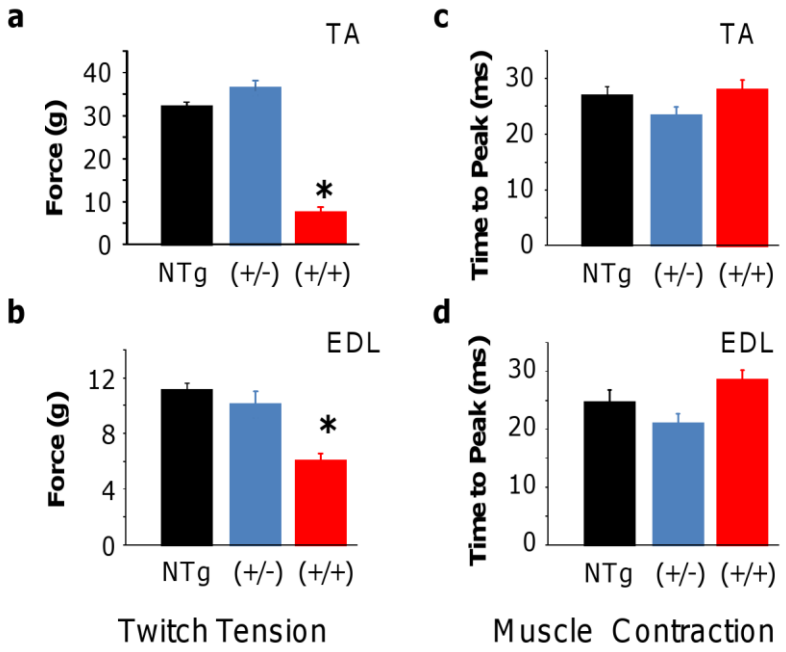

Figure S3 Impaired twitch force but normal muscle contraction in hFUS homozygous mice.

(a-b) Twitch force production of the hindlimb TA (A) and EDL (B) muscles was significantly reduced in hFUS (+/+) mice compared to NTg mice, with a deficit in force of approximately 80% and 20% in the TA and EDL respectively (*****$p<0.001$). (c-d) Muscle contraction measured as time to peak in both muscles was not significantly altered in response to hFUS overexpression