**Niclosamide targets inflammatory and profibrotic pathways in**

**amyotrophic lateral sclerosis**

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**Running title**

Niclosamide in ALS

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**Supplementary Material**

**Figure S1. FUS and TARDBP associated mutations induce an upregulation of S100A4**

Protein lysates of fibroblasts from controls (n=3/group), and from a patient carrying the *FUS* p.R521C pathogenic variant (n=3 replicates) (**a**) or from a patient with the *TARDBP* p. Q303H and a patient with the *TARDBP* p.A382T mutations (**b**) were analysed by western blotting using anti-S100A4. GAPDH was used to normalize samples. The expression levels were calculated by densitometric analyses. Data represent mean ± SEM. Statistical significance was calculated by student’s t-test and values significantly different from controls are indicated with an asterisk when p≤0.05.

**Figure S2. S100A4 is increased in the spinal cord of hFUS mice**

(**a**) Protein from lumbar spinal cord lysates of non-transgenic (Non-Tg) and hFUS mice at end stage (n=5/group) were assayed by Western blot with anti-S100A4. GAPDH levels served as loading control. Relative densitometric values are reported on the right. Data represent mean ± SEM. Statistical significance was calculated by student’s *t*-test and significantly different values from Non-Tg are indicated with an asterisk when p≤0.05. (**b**) Representative fluorescence images of S100A4 (green) and GFAP (purple) in grey and white matter of the lumbar spinal cord from non-transgenic (Non-Tg) (~ 40 days) and end stage hFUS mice. WM = white matter. GM = grey matter. Scale bars: 50 µm.



