Supplemental information figure legends

Figure S1. Gut motility assay on SOD1 G93A mice. Intestinal transit time was assessed by time in hours required for the appearance of the first blue pellet after the administration of 0.3ml of blue dye via oral gavage in male (left; *n*=10 *SOD1*, 6 *WT*) and female (right; *n*=11 *SOD1*, 6 *WT*) mice. No significant differences were detected. Values represent the mean ± SEM.

Figure S2. Gait analysis on TDP-43 mice. The hindlimbs of male (left panels; *n*=20 *TDP-43*, 10 *WT*) and female (right panels; *n*=19 *TDP-43*, 10 *WT*) mice were dipped in non-toxic black India ink and then the mice were inserted inside a tunnel-like structure, 4 cm in diameter and 30 cm in length, positioned over a white sheet of paper. The mice quickly ran through the tunnel leaving black footprints behind. Analysis was performed on the three most consistent consecutive footprints. The length between footprints of the same side (toe to toe) was taken as the stride length (top panels). The width between footprints from opposite sides was taken as the stance width (bottom panels). No significant differences were detected. Values represent the mean ± SEM.

Figure S3. Superior mesenteric ganglion. Expression of the TDP-43 transgene was assayed by anti-Flag imunocytochemistry in the SMG (green: anti-Flag, blue: DAPI) of 90-150 day old WT (A) and TDP-43 (B) mice. Hematoxylin-eosin staining of SMG paraffin sections in *WT* (C) and *TDP-43* (D), 90-150 day old male mice. Representative pyknotic nuclei in TDP-43 are indicated by arrows (D). Scale bar: 50 uM. Quantification of the pyknotic nuclei expressed as a % of total neurons (E; *n*=3 mice per genotype, Student's t-test, p=0069).

Figure S4. Nerve histology. Representative sections of the motor branch of the femoral nerve of representative 3-month old control (A) and TDP-43 (B) mice. Scale bar: 50 um. Axon number (C,E) and mean diameter (D,F) was quantified in the motor (C,D) and sensory (E,F) branches of the femoral nerve. *n*=7 mice per genotype. Genotype effect is shown by brackets, two-way ANOVA, $p < 0.05$. Post-hoc test found no significant difference by multiple comparisons.

Figure S5. Neuromuscular junction phenotype. Representative neuromuscular junctions of the tibialis anterior stained with alpha-bungarotoxin in a 3 month old male control (A) and TDP-43 (B). Scale bar: 50 um. Quantification of the NMJ area in the tibialis anterior of 3-5 month old male mice (C). *n*=7 mice per genotype. Student's t-test <0.001.

Figure S1

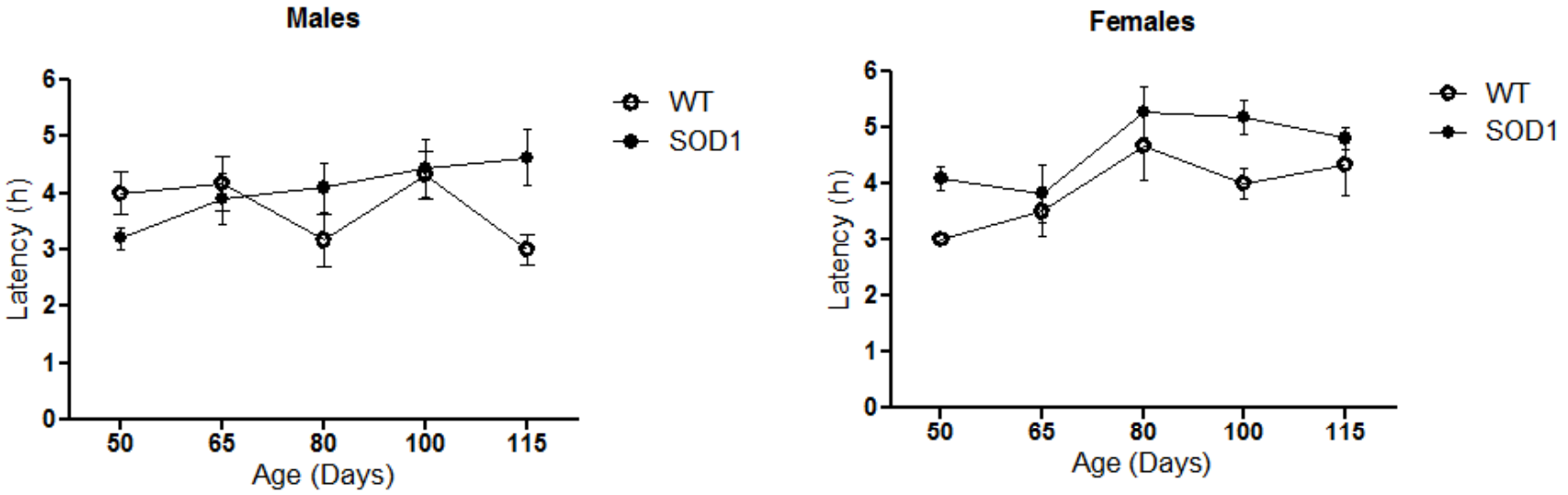
Males
Latency (h)
0
1
2
3
4
5
6
50
65
80
100
115
Age (Days)
WT
SOD1
Females
Latency (h)
0
1
2
3
4
5
6
50
65
80
100
115
Age (Days)
WT
SOD1

Figure S2

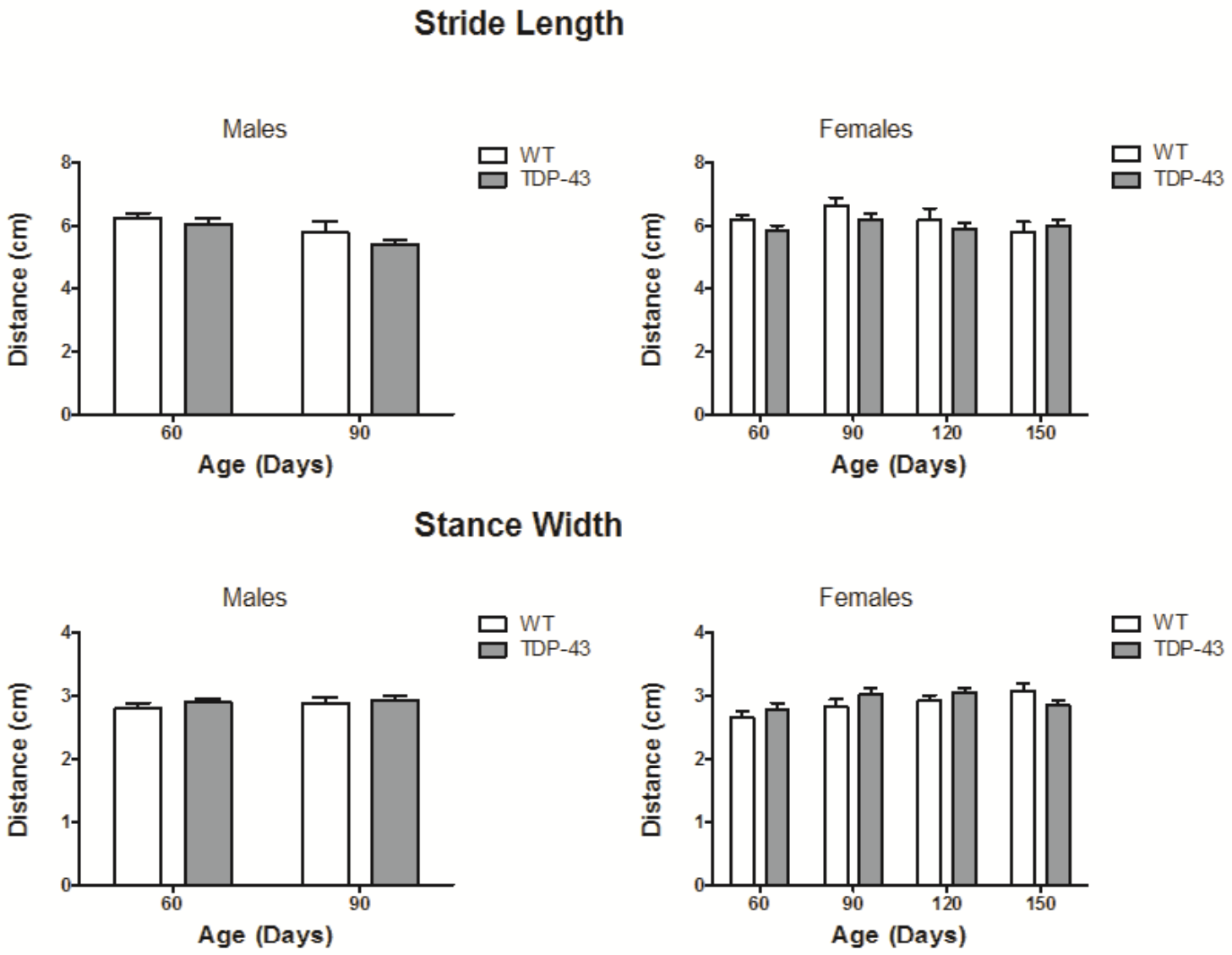
Stride Length
Males
WT
TDP-43
Distance (cm)
0
2
4
6
8
60
90
Age (Days)
Females
WT
TDP-43
Distance (cm)
0
2
4
6
8
60
90
120
150
Age (Days)
Stance Width
Males
WT
TDP-43
Distance (cm)
0
1
2
3
4
60
90
Age (Days)
Females
WT
TDP-43
Distance (cm)
0
1
2
3
4
60
90
120
150
Age (Days)

Figure S3

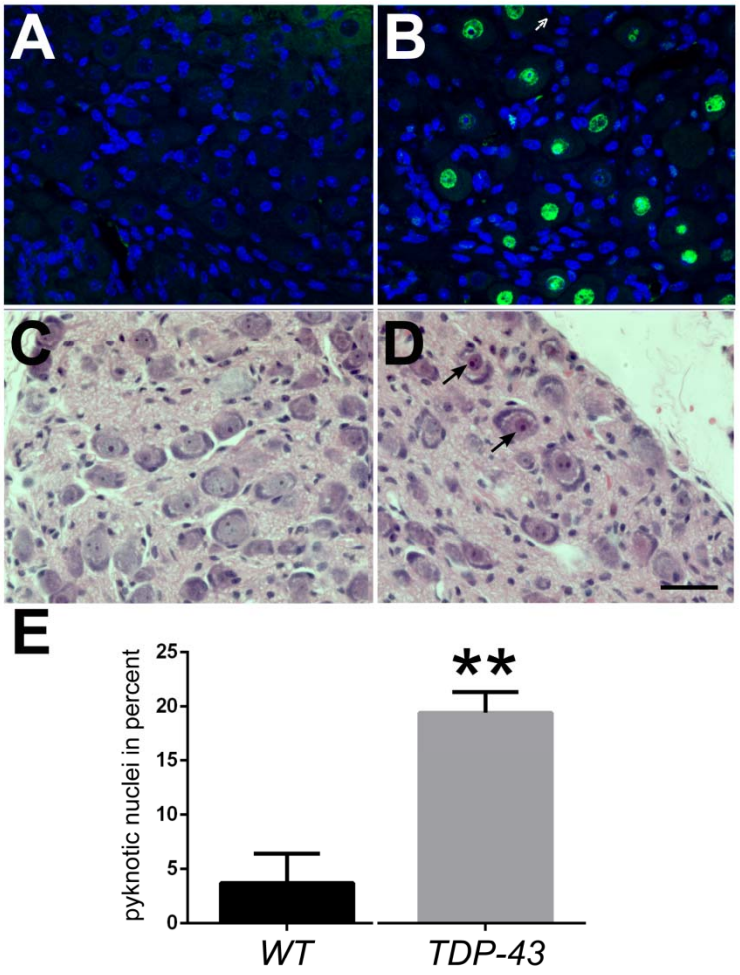

Figure S4

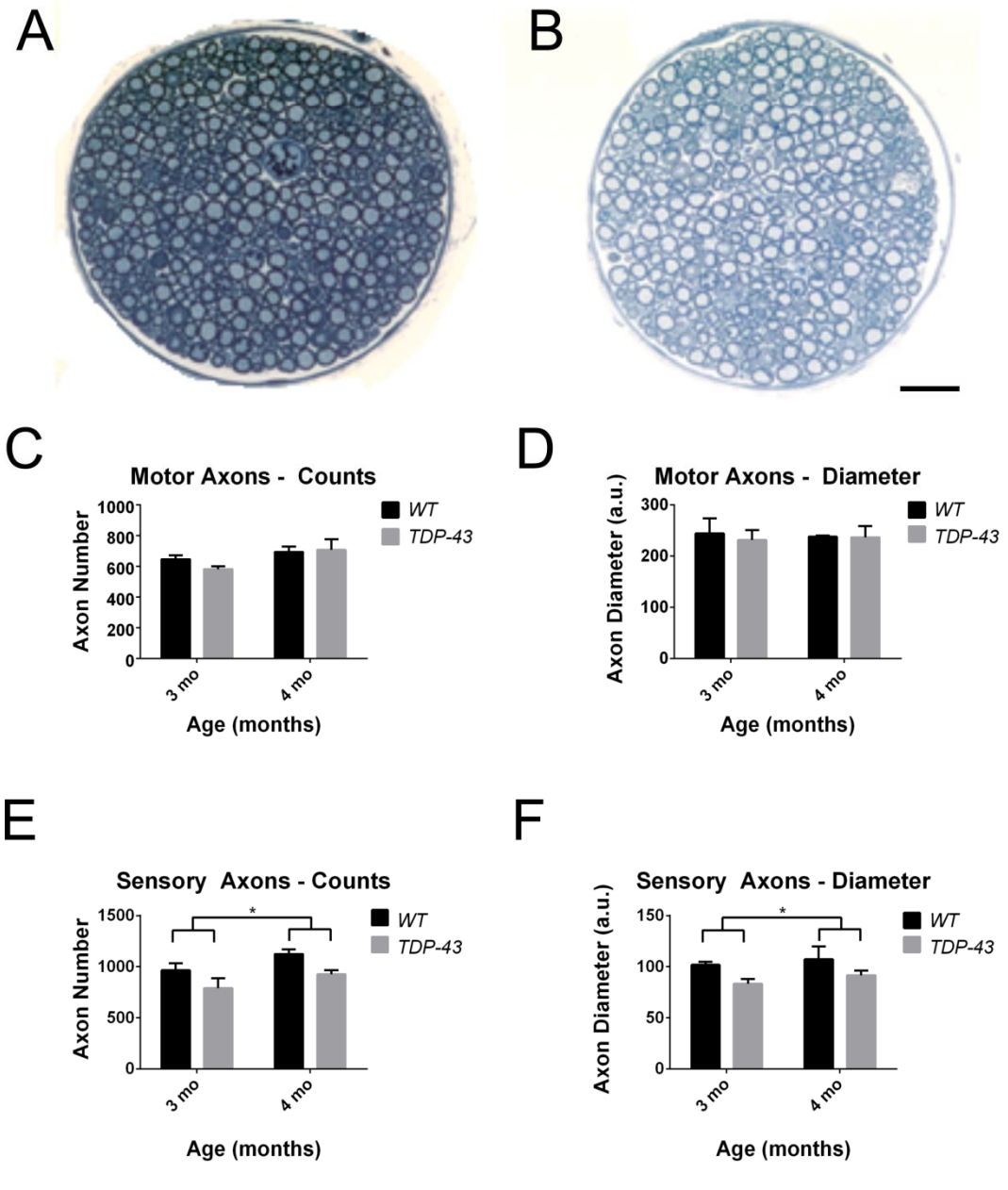
A
B
C
Motor Axons - Counts
Axon Number
1000
800
600
400
200
0
3 mo
4 mo
Age (months)
WT
TDP-43
D
Motor Axons - Diameter
Axon Diameter (a.u.)
300
200
100
0
3 mo
4 mo
Age (months)
WT
TDP-43
E
Sensory Axons - Counts
Axon Number
1500
1000
500
0
*
3 mo
4 mo
Age (months)
WT
TDP-43
F
Sensory Axons - Diameter
Axon Diameter (a.u.)
150
100
50
0
*
3 mo
4 mo
Age (months)
WT
TDP-43

Figure S5