

Table S1. Mean axonal caliber and relative frequency of small (axonal diameters  $\leq 6$   $\mu\text{m}$ ) and large (axonal diameters  $> 6$   $\mu\text{m}$ ) myelinated fibers in the dorsal and ventral roots and sciatic nerve.

	MOCA +/+	MOCA -/-	P value
Dorsal root			
MAD ( $\mu\text{m}$ )	5.60 $\pm$ 0.26	6.00 $\pm$ 0.39	NS
$\leq 6$ (%)	63.0 $\pm$ 5.3	53.1 $\pm$ 8.5	NS
$> 6$ (%)	37.0 $\pm$ 5.4	46.9 $\pm$ 8.5	NS
Ventral root			
MAD ( $\mu\text{m}$ )	7.41 $\pm$ 0.39	8.42 $\pm$ 0.45	NS
$\leq 6$ (%)	32.9 $\pm$ 3.6	20.1 $\pm$ 4.3	NS
$> 6$ (%)	63.2 $\pm$ 3.5	79.9 $\pm$ 4.3	$< 0.03$
Sciatic nerve			
MAD ( $\mu\text{m}$ )	6.02 $\pm$ 0.32	5.62 $\pm$ 0.19	NS
$\leq 6$ (%)	58.6 $\pm$ 4.7	64.7 $\pm$ 4.3	NS
$> 6$ (%)	41.1 $\pm$ 4.7	35.2 $\pm$ 4.2	NS

Data are presented as mean  $\pm$  SEM (N = 3-4 per group for dorsal and ventral roots; N = 8 per group for sciatic nerve) and were analyzed with an unpaired, two-tailed t-test. MAD – mean axonal diameter.

Table S2. Myelinated fiber number, fascicular area and myelinated fiber density in the dorsal and ventral roots.

	MOCA +/+	MOCA -/-	P value
Dorsal root			
# myelinated fibers	1,819±146	1,692±333	NS
Fascicular area (mm <sup>2</sup> )	0.076±0.007	0.070±0.012	NS
Myelinated fiber density (#/mm <sup>2</sup> )	25,252±733	23,874±909	NS
Ventral root			
# myelinated fibers	885±101	967±81	NS
Fascicular area (mm <sup>2</sup> )	0.069±0.007	0.072±0.004	NS
Myelinated fiber density (#/mm <sup>2</sup> )	12,725±395	13,506±757	NS

Data are presented as mean ± SEM (N = 3-4 per group) and were analyzed with an unpaired, two-tailed t-test.

**Figure S1.** Construction of *moca*<sup>-/-</sup> mice. (A) The targeting construct for MOCA knockout. (B-C) Southern hybridization of mutant ES clones. The wild type shows only one 21kb fragment while the chimeric clones show two bands containing 21 and 18kb fragments, respectively, which represents the 5' arm integration (B). (C) The 3' arm integration patterns (two fragments of 8 and 7kb for the chimeric clones while one fragment of 8kb for the wild type). (D) Genotyping of MOCA gene-knockout mice by PCR. The primer set 1 has two primers both located on the downstream of MOCA gene exon 2 (lane 1). The primer set 2 contains one primer that is the same as the above 5'-end primer and the other that is localized within the *neo* gene in the targeting vector (lane 2). The primer set 1 amplifies the wild type sequence while the primer set 2 only amplifies the targeting construct sequence.

**Figure S2.** Muscle atrophy and sciatic nerve abnormalities in *moca*<sup>-/-</sup> mice. (A-B) Hematoxylin and eosin staining of the gastrocnemius muscle of control (A) and *moca*<sup>-/-</sup> (B) mice at the age of 20 months. (C-D) ATPase staining of the gastrocnemius muscle of control (C) and *moca*<sup>-/-</sup> mice (D). (E-H) Toluidine blue staining of sciatic nerve of control (E, G) and *moca*<sup>-/-</sup> mice (F, H).

**Figure S3.** The size-frequency distribution of nerve fibers derived from L5 dorsal (A) and ventral (B) roots of *moca*<sup>+/+</sup> and *moca*<sup>-/-</sup> mice.

**Figure S4.** Abnormal axonal spheroids become apparent after the embryonic development. (A-B) No NF aggregate stained by a NF-68 antibody is seen in the spinal cord of mutant or control mice on postnatal day one. (C-D) Abnormal axonal spheroids stained by a NF-68

antibody are present in the spinal cord of *moca*<sup>-/-</sup> mice at the age of 2 months (D) but not in the age-matched controls (C). (E-F) Activated microglia are present in the spinal cord of *moca*<sup>-/-</sup> (F) but not in control (E) mice at the age of 2 months. Bar = 50µm.

Figure S1

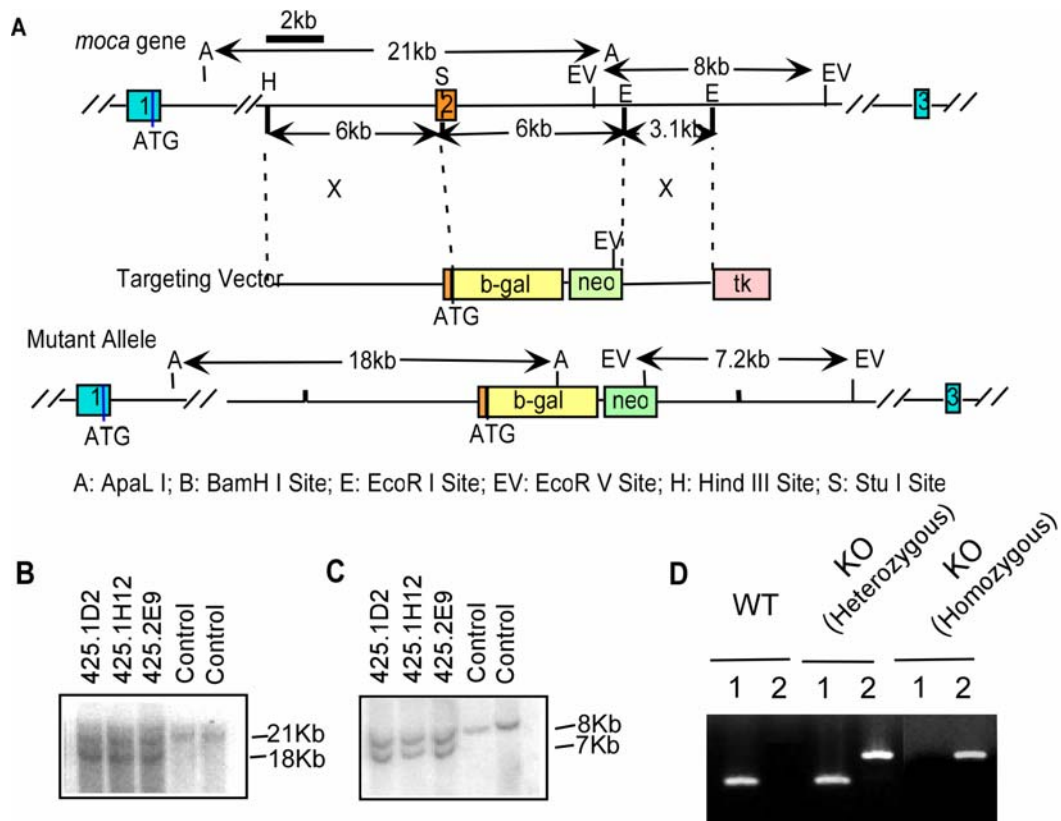


Figure S2

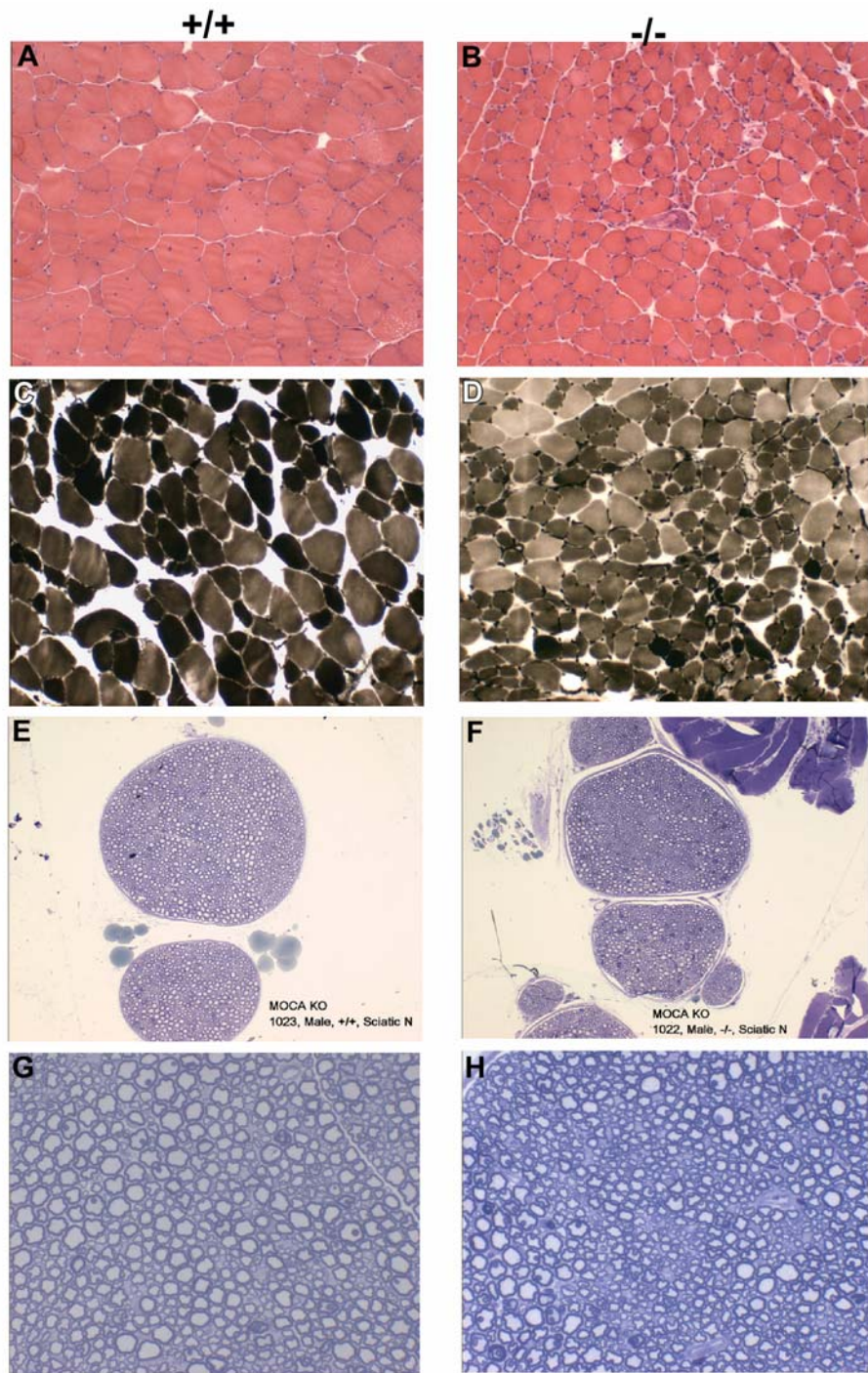


Figure S3

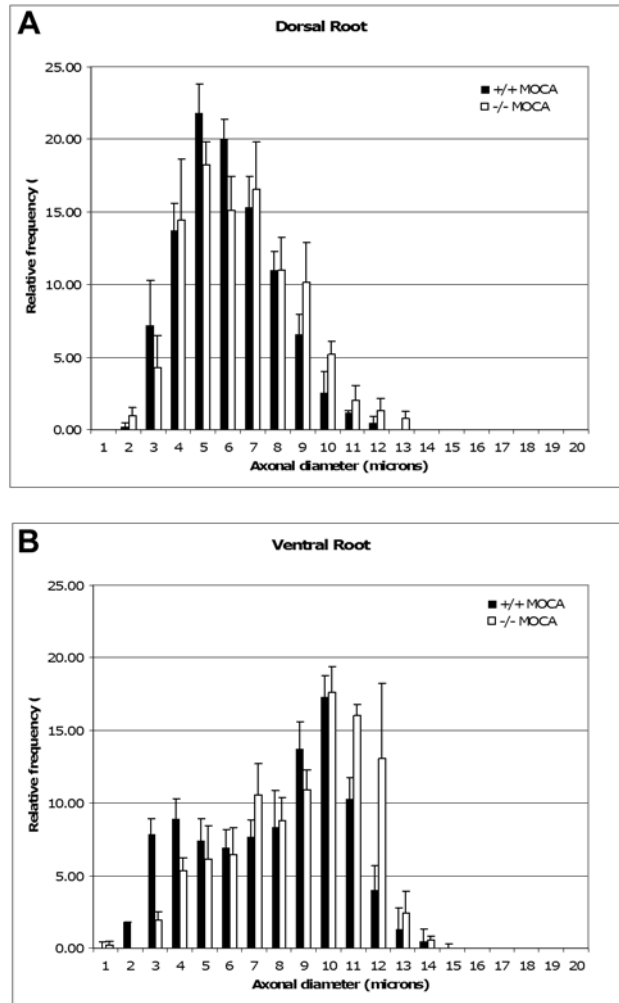


Figure S4

