Fig. S1. Progressive impairment of motor functions in lister mutant mice. (A) Progression of hind-limb extension reflex measured by the time to develop clenching. Score 0 — no clenching in 30 sec, score 1 — clenching in 21–30 sec, score 2 — clenching in 11–20 sec, score 3 — clenching in 6–10 sec, score 4 — clenching in 5 sec, score 5 — clenching in 5 sec and difficulty in recovering. lister/lister (color lines) and wild-type (gray line) mice. More than 20 of +/+ and lister/+ tested, all showing 0 in this assay. (B) Progression of the righting reflex ability in lister/lister mutants within 60 sec. Each trace represents an individual lister/lister mouse.
Fig. S2. Localized gliosis in brainstem. (A–C) GFAP staining (arrow) of brainstem (A), cerebellum (B), and olfactory bulb (C) from +/+ and lister/lister mice.
Fig. S3. Ultrastructural abnormalities in gray and white matter of *lister* mutant mice. (A and B) TEM images of neurons (arrows) in the ventral side of lumbar spinal cord from wild-type and *lister/lister*, respectively. Arrowheads indicate abnormal axons in B. (C and D) TEM images of axons in white matter of the ventral lateral side of lumbar spinal cord from wild-type and *lister/lister*, respectively. Arrowheads indicate two of the abnormal/degenerating axons. (E and F) Vacuolated mitochondria (M) are found in postsynaptic profiles (+), likely dendrites of mutant animal (F). Compare this to the wild-type animal (E). Note Synapses or presynaptic profiles appear to be normal. Arrowhead indicates a damaged axoplasm surrounded by myelin sheath.
Fig. S4. Axonal and neuronal analysis in wild-type and lister mutant mice. (A) Distribution of axon diameters from the entire motor and sensory L5 roots from lister/lister (n = 2) and wild-type (n = 4) animals at symptom onset. (B and C) Toluidine blue staining of distal femoral nerve branches of wild-type and lister mutants, respectively. Note two of the four branches in the mutant have abnormal axons. Arrows indicate degenerating axons.
Fig. S5.  *lister* is expressed in the brain and spinal cord. (A) Detection of *lister* mRNA expression, using antisense probe or sense probe as negative control, in adult wild-type brain and spinal cord sections.
Fig. S6. Gene trap lister allele-RRR322 and phenotypic analysis of lister/RRR322 mice. (A) A schematic representation of the β-geo insertion in line RRR322 targeting intron 25 of lister gene. (B) Motor dysfunction revealed by accelerated rotarod test in compound heterozygous lister/RRR322 (n = 2), but not +/lister (n = 2), RRR322/+ (n = 1) or +/+ mice (n = 6). +/+ versus lister/RRR322 mice are significant at 7 weeks of age (P < 0.005) and at 14 weeks of age (P < 0.001).
Movie S1. A homozygous lister animal at 2 months of age.
Movie S2. A compound heterozygous lister/RRR322 mouse at 4.5 months of age.