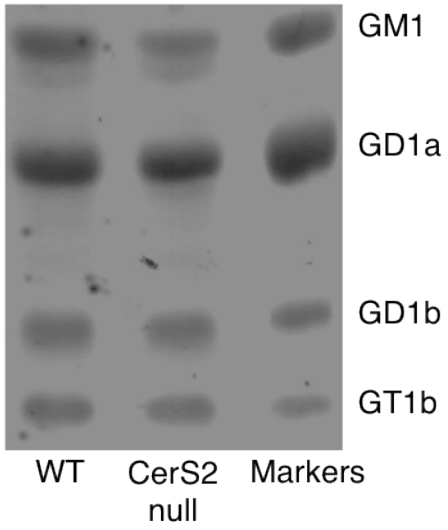
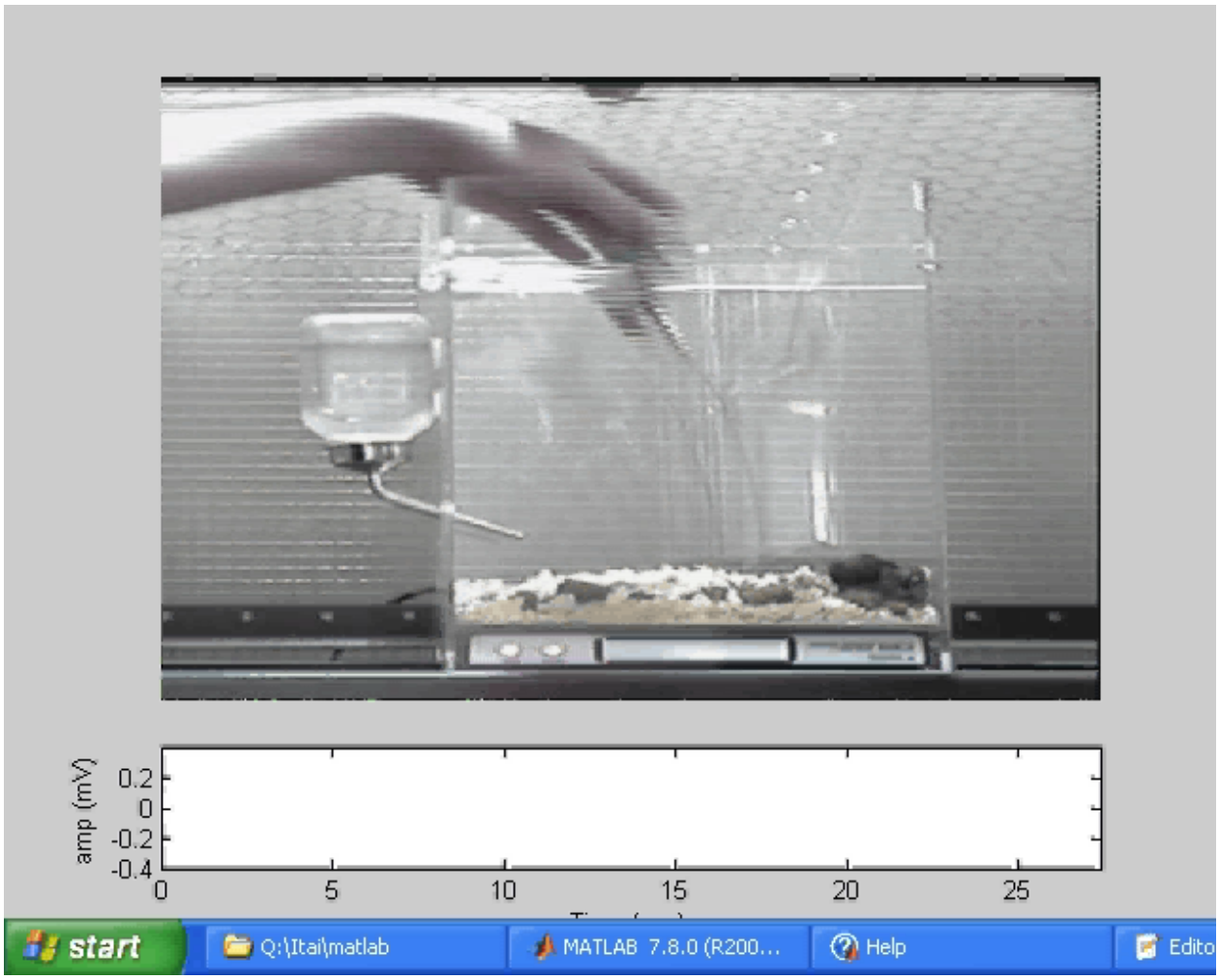
Supplementary Data

Suppl. Fig. 1. Ganglioside levels in 4 month-old mice. One of three representative thin layer chromatography plates is shown; in 2 of the 3 analyses there was no change in GM1 levels, but a small decrease in GM1 levels is observed in the thin layer chromatography plate shown in the figure. The *right-hand* lane shows pure ganglioside standards.



Suppl. Movie 1. EEG recording of the CerS2 null mice during an audible induced stimulus, inflicted by key jerking.

Recording of a CerS2 null mouse during an audible-induced stimulus response inflicted by shaking a key bundle above the transgenic mouse for 5 secs, which induced a myoclonic jerk in the CerS2 null mouse. EEG recording during the induced response is shown. No change in cortical EEG was observed indicating a sub-cortical origin of the motor dysfunction.



Supplementary Table 1. HexCer composition of WT and CerS2 null mouse brains. For details, see Fig. 1.

Age (*days*)	HexCer acyl chain length (*pmol/mg of tissue*) (WT)						
	C16	C18	C18:1	C20	C22	C24:1	C24
0	1.2 ± 0.04	8.5 ± 6.6	0.2 ± 0.02	0.3 ± 0.1	1.3 ± 0.6	0.4 ± 0.3	0.3 ± 0.1
30	1.2 ± 0.5	36.9 ± 7.7	0.1 ± 0.1	3.3 ± 0.8	16 ± 1.2	122.1 ± 5.2	39.8 ± 4.1
120	2.2 ± 0.4	51.5 ± 11.2	0.4 ± 0.1	17 ± 5.5	112.6 ± 22.2	838 ± 80	281 ± 52.1

Age (*days*)	HexCer acyl chain length (*pmol/mg of tissue*) (CerS2 null)						
	C16	C18	C18:1	C20	C22	C24:1	C24
0	1.3 ± 0.5	5.5 ± 1.6	0.3 ± 0.1	0.4 ± 0.2	0.7 ± 0.2	0.3 ± 0.1	0.2 ± 0.005
30	3.9 ± 0.3 [a]	214 ± 12.2 [a]	0.0 ± 0.0 [a]	2.1 ± 0.2 [a]	0.8 ± 0.2 [a]	0.4 ± 0.2 [a]	0.4 ± 0.1 [a]
120	3.2 ± 0.9 [a]	106 ± 38 [a]	0.9 ± 0.5 [a]	9 ± 0.6 [a]	3.9 ± 0.3 [a]	4.1 ± 1.5 [a]	4.2 ± 0.6 [a]

[a] $p<0.05$ (CerS2 null versus WT)

Supplementary Table 2. SL composition of purified myelin from WT and CerS2 null mice. For details, see Fig. 3.

	SL acyl chain length (*pmol/mg of tissue*) (WT)						
Lipid	C16	C18	C18:1	C20	C22	C24:1	C24
Ceramide	22.3 ± 5.1	549 ± 75	4.7 ± 1.3	14.3 ± 1.8	9.6 ± 2.3	258 ± 91	23.5 ± 5.2
SM	46.8 ± 6	452 ± 20.6	120 ± 20	34.3 ± 4.1	34.4 ± 4.2	447 ± 60	82.5 ± 6.8
GlcCer	0.4 ± 0.3	2.3 ± 0.4	1 ± 0.2	0.2 ± 0.04	0.5 ± 0.04	7.4 ± 1.9	1.5 ± 0.5
GalCer	6.2 ± 3.5	79 ± 33	1.7 ± 1.2	26.6 ± 13.5	147 ± 67	1963 ± 1158	417 ± 200
LacCer	1.2 ± 0.3	16.4 ± 1.6	0.3 ± 0.04	0.5 ± 0.2	0.5 ± 0.1	4.2 ± 1.1	0.9 ± 0.3

	SL acyl chain length (*pmol/mg of tissue*) (CerS2 null)						
Lipid	C16	C18	C18:1	C20	C22	C24:1	C24
Ceramide	34.1 ± 29.8	652 ± 515	24 ± 17.3[a]	17.2 ± 15.3[a]	0.4 ± 0.3[a]	0.6 ± 0.1[a]	0.5 ± 0.1[a]
SM	66.2 ± 62.2	711 ± 586	56.2 ± 54.4	45.8 ± 40.5	0.8 ± 0.4[a]	0.6 ± 0.3[a]	0.4 ± 0.1[a]
GlcCer	0.2 ± 0.05[a]	5.3 ± 1.3	5.2 ± 2.6[a]	0.6 ± 0.5	0.3 ± 0.2[a]	0.9 ± 0.3[a]	0.8 ± 0.6[a]
GalCer	1.6 ± 1.3	336 ± 239	3 ± 2.4	2.9 ± 2.7	2.5 ± 1[a]	8.9 ± 2.9[a]	3.4 ± 1.2[a]
LacCer	0.6 ± 0.2	23.3 ± 17.3	0.3 ± 0.1	1.0 ± 0.5	0.2 ± 0.1[a]	0.1 ± 0.1[a]	0.9 ± 0.04[a]

[a] $p<0.05$ (CerS2 null versus WT)