

**Table S1**

Study-specific Designation	Common Name	Jax #	SMN2 Copy #	SMA genotype	Breeding Strategy	Median survival untreated SMA	Primary outcome measures	Secondary outcome measures	Reference	% SMA pups/litter
5058-Hemi-FVB/N	Taiwanese Hemizygous	#005058	2	<i>Tg(SMN2)2Hung<sup>tg/0</sup>;Smn<sup>tm1Hung/tm1Hung</sup></i>	♂: Smn <sup>tm1Hung/Wt</sup> (FVB/N) X ♀: Tg(SMN2)2Hung <sup>tg/tg</sup> ;Smn <sup>tm1Hung/tm1Hung</sup>	9 days	Survival, righting reflex	Weight	Hsieh-Li et al., 2000; Gogliotti et al., 2010	50%
5058-Hemi-Hybrid	N/A	#006146 #005058	2	<i>Tg(SMN2)2Hung<sup>tg/0</sup>;Smn<sup>tm1Hung/tm1Jme</sup></i>	♂: Smn <sup>tm1.1Jme/Wt</sup> (C57BL/6J-congenic) X ♀: Tg(SMN2)2Hung <sup>tg/tg</sup> ;Smn <sup>tm1Hung/tm1Hung</sup>	22 days	Survival, Pedal edema	Weight, ECG, NMJ Pathology	Current Study	50%
5058-Homozygous	Taiwanese Homozygous	#005058	4	<i>Tg(SMN2)2Hung<sup>tg/0</sup>;Tg(SMN2)2Hung<sup>tg/0</sup>;Smn<sup>tm1Hung/tm1Hung</sup></i>	♂: Tg(SMN2)2Hung <sup>tg/tg</sup> ;Smn <sup>tm1Hung/tm1Hung</sup> X ♀: Tg(SMN2)2Hung <sup>tg/tg</sup> ;Smn <sup>tm1Hung/tm1Hung</sup>	WT survival	Protein and RNA Analyses	N/A	Hsieh-Li et al., 2000; Gogliotti et al., 2010	100%

**Table S1** SMA models utilized in this study. The table outlines the 3 SMA mouse models used in the present study, including the various nomenclatures, genotype, breeding strategy, median survival, and outcome measures of each mouse strain.