Smn1-Flox

Nomenclature	C57BL/6Smoc-*Smn1*$^{tm1(flox)Smoc}$
Cat. NO.	NM-CKO-2102063
Strain State	Embryo cryopreservation

Gene Summary

Gene Symbol Smn1	**Synonyms**	Smn; Gemin1; AI849087
	NCBI ID	20595
	MGI ID	109257
	Ensembl ID	ENSMUSG00000021645
	Human Ortholog	SMN1

Model Description

These mice carry loxP sites flanking target exon 8 of Smn1 gene. When crossed with a Cre recombinase-expressing strain, this strain is useful in eliminating tissue-specific conditional expression of Smn1 gene.
*Literature published using this strain should indicate: Smn1-Flox mice (Cat. NO. NM-CKO-2102063) were purchased from Shanghai Model Organisms Center, Inc..

Disease Connection

Werdnig-Hoffmann Disease	**Phenotype(s)**	MGI:5289775 Note: The expected phenotype(s) may be observed in the above-mentioned mice that bred with Grm7-Flox(NM-CKO-2103861) and Olig2-Cre mice.
	Reference(s)	Park GH, Maeno-Hikichi Y, Awano T, Landmesser LT, Monani UR, Reduced survival of motor neuron (SMN) protein in motor neuronal progenitors functions cell autonomously to cause spinal muscular atrophy in model mice expressing the human centromeric (SMN2) gene. J Neurosci. 2010 Sep 8;30(36):12005-19

Werdnig-Hoffmann disease	Phenotype(s)	MGI:5318858 Note: The expected phenotype(s) may be observed in the above-mentioned mice that bred with Grm7-Flox(NM-CKO-2103861) and Mnx1-Cre mice.
	Reference(s)	Gogliotti RG, Quinlan KA, Barlow CB, Heier CR, Heckman CJ, Didonato CJ, Motor neuron rescue in spinal muscular atrophy mice demonstrates that sensory-motor defects are a consequence, not a cause, of motor neuron dysfunction. J Neurosci. 2012 Mar 14;32(11):3818-29
Werdnig-Hoffmann disease	Phenotype(s)	MGI:3721431 Note: The expected phenotype(s) may be observed in the above-mentioned mice that bred with Eno2-cre mice.
	Reference(s)	Frugier T, Tiziano FD, Cifuentes-Diaz C, Miniou P, Roblot N, Dierich A, Le Meur M, Melki J, Nuclear targeting defect of SMN lacking the C-terminus in a mouse model of spinal muscular atrophy. Hum Mol Genet. 2000 Mar 22;9(5):849-58
Werdnig-Hoffmann disease	Phenotype(s)	MGI:5287852 Note: The expected phenotype(s) may be observed in the above-mentioned mice that bred with Eno2-cre mice.
	Reference(s)	Ferri A, Melki J, Kato AC, Progressive and selective degeneration of motoneurons in a mouse model of SMA. Neuroreport. 2004 Feb 9;15(2):275-80
Werdnig-Hoffmann disease	Phenotype(s)	MGI:3721894 Note: The expected phenotype(s) may be observed in the above-mentioned mice that bred with ACTA1-cre mice.
	Reference(s)	Cifuentes-Diaz C, Frugier T, Tiziano FD, Lacene E, Roblot N, Joshi V, Moreau MH, Melki J, Deletion of murine smn exon 7 directed to skeletal muscle leads to severe muscular dystrophy. J Cell Biol. 2001 Mar 5;152(5):1107-14

Validation Data

No data