

These mice contain a global knock-out of the sulfonylurea receptor (Sur1), which is part of the ATP-dependent potassium channel. In humans, mutations in Sur1 cause persistent hyperinsulinemic hypoglycemia of infancy (PHHI).

Keywords: [Sur1](#) [Sur1<sup>neo</sup>](#) [mouse](#) [Abcc8](#)

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## Mouse Information

Common Name	Sur1 <sup>neo</sup>
Research Applications	<i>Not provided</i>
MMRRC ID	<a href="#">011968-UNC</a>
Jackson Laboratories Stock No	<i>Not provided</i>
VCMR ID	CO, GA
Additional Strain Information	<i>Not provided</i>

## Genetic Alteration

### Mutation #1: Targeted Mutagenesis

Type of Allele	Global Null
Targeted Gene	Name: ATP-binding cassette, sub-family C (CFTR/MRP), member 8 Symbol: <a href="#">Abcc8</a> NCBI: <a href="#">20927</a>
Allele	Name: targeted mutation 1.1Mgn Symbol: <a href="#">Sur1<sup>neotm1.1Mgn</sup></a> MGI: <a href="#">MGI:2388392</a>

<b>Description of Targeting Vector</b>	A gene targeting strategy that involved the use of Cre/loxP was used generate mice that globally lack the proximal promoter and exon 1 of the sulfonylurea receptor type 1 (Sur1) gene. A pgk-neomycin resistance gene remains in the locus. Genotype: DNA PCR utilizing 5'-CAA TTC CTC AAC TGA GGC TCT TAA-3' and 5'-AGC CTC TGT TCC ACA TAC ACT TCA-3' primers amplify a 414 bp Sur1[neo] allele. DNA PCR using 5'-CAA TTC CTC AAC TGA GGC TCT TAA-3' and 5'-TCG CAG AGT GAC CTC ACA GCC TGT-3' primers amplify a 412 bp for the wild type allele. Homozygous phenotype: Mice that are homozygous null for Sur1 (Abcc8) are viable, fertile and grossly indistinguishable from their wild type littermates except after 16 wks when they become mildly hypoglycemia. Mice lacking Sur1 (Abcc8) lack functional K-ATP channels in pancreatic beta cells but remain euglycemic despite having beta cells that are constantly depolarized due to the lack of this protein. Heterozygous phenotype: These mice are also viable and do not differ from wild type.
<b>Vector Genbank File</b>	<a href="#">pSUR1.KO1.gb</a>
<b>Allele Map</b>	<i>Not Provided</i>
<b>PCR Genotyping Protocol</b>	<i>Not provided</i>
<b>Citations</b>	<p><b>Publication</b></p> <p><a href="#">Sulfonylurea receptor type 1 knock-out mice have intact feeding-stimulated insulin secretion despite marked impairment in their response to glucose.</a> (2002) <i>J Biol Chem</i> <b>277</b>: 37176-83 (Added 12/10/2013)  PMID: <a href="#">12149271</a></p>

## Background Strain Information

<b>Strain Type</b>	Congenic Strain
<b>Chimera/Founder Genetic Background</b>	129S6/SvEvTac
<b>Current Genetic Background</b>	C57BL/6J
<b>Number of Generations Backcrossed</b>	10
<b>Strain Description</b>	After achieving germline transmission mice carrying the Sur1[neo] allele were backcrossed for ten generations.

## Publications / Citations

1. [Differential structure of atrial and ventricular KATP: atrial KATP channels require SUR1.](#) Flagg TP, Kurata HT, Masia R, Caputa G, Magnuson MA, Lefer DJ, Coetzee WA, Nichols CG (2008) *Circ Res* **103**(12): 1458-65  
> Primary publication · [18974387](#) (PubMed) · [PMC2768594](#) (PubMed Central) · Added on 2/23/2011
2. [Exendin-\(9-39\) corrects fasting hypoglycemia in SUR-1<sup>-/-</sup> mice by lowering cAMP in pancreatic beta-cells and inhibiting insulin](#)

- [secretion](#). De León DD, Li C, Delson MI, Matschinsky FM, Stanley CA, Stoffers DA (2008) *J Biol Chem* **283**(38): 25786-93  
 › Primary publication · [18635551](#) (PubMed) · [PMC3258866](#) (PubMed Central) · Added on 12/21/2016
3. [Role of sulfonylurea receptor type 1 subunits of ATP-sensitive potassium channels in myocardial ischemia/reperfusion injury](#). Elrod JW, Harrell M, Flagg TP, Gundewar S, Magnuson MA, Nichols CG, Coetzee WA, Lefer DJ (2008) *Circulation* **117**(11): 1405-13  
 › Primary publication · [18316485](#) (PubMed) · Added on 2/23/2011
4. [Hyperinsulinism in mice with heterozygous loss of K\(ATP\) channels](#). Remedi MS, Rocheleau JV, Tong A, Patton BL, McDaniel ML, Piston DW, Koster JC, Nichols CG (2006) *Diabetologia* **49**(10): 2368-78  
 › Primary publication · [16924481](#) (PubMed) · Added on 12/21/2016
5. [Cholinergic regulation of fuel-induced hormone secretion and respiration of SUR1<sup>-/-</sup> mouse islets](#). Doliba NM, Qin W, Vatamaniuk MZ, Buettger CW, Collins HW, Magnuson MA, Kaestner KH, Wilson DF, Carr RD, Matschinsky FM (2006) *Am J Physiol Endocrinol Metab* **291**(3): E525-35  
 › Primary publication · [16638820](#) (PubMed) · Added on 2/23/2011
6. [Impaired glucagon secretory responses in mice lacking the type 1 sulfonylurea receptor](#). Shiota C, Rocheleau JV, Shiota M, Piston DW, Magnuson MA (2005) *Am J Physiol Endocrinol Metab* **289**(4): E570-7  
 › Primary publication · [15941784](#) (PubMed) · Added on 12/10/2013
7. [Restitution of defective glucose-stimulated insulin release of sulfonylurea type 1 receptor knockout mice by acetylcholine](#). Doliba NM, Qin W, Vatamaniuk MZ, Li C, Zelent D, Najafi H, Buettger CW, Collins HW, Carr RD, Magnuson MA, Matschinsky FM (2004) *Am J Physiol Endocrinol Metab* **286**(5): E834-43  
 › Primary publication · [14736703](#) (PubMed) · Added on 2/23/2011
8. [Sulfonylurea receptor type 1 knock-out mice have intact feeding-stimulated insulin secretion despite marked impairment in their response to glucose](#). Shiota C, Larsson O, Shelton KD, Shiota M, Efanov AM, Hoy M, Lindner J, Kooptiwut S, Juntti-Berggren L, Gromada J, Berggren PO, Magnuson MA (2002) *J Biol Chem* **277**(40): 37176-83  
 › Primary publication · [12149271](#) (PubMed) · Added on 12/10/2013
9. [The imidazoline RX871024 stimulates insulin secretion in pancreatic beta-cells from mice deficient in K\(ATP\) channel function](#). Efanov AM, Høy M, Bränström R, Zaitsev SV, Magnuson MA, Efendic S, Gromada J, Berggren PO (2001) *Biochem Biophys Res Commun* **284**(4): 918-22  
 › Primary publication · [11409880](#) (PubMed) · Added on 5/27/2010

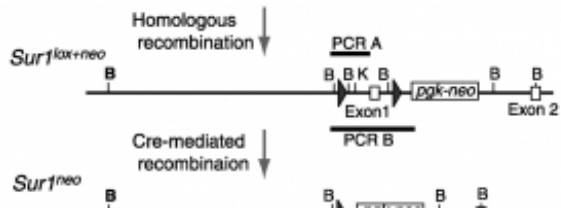
## MeSH Terms


- 1-Methyl-3-isobutylxanthine Acetylcholine Acetylcholinesterase Amino Acids Animals ATP-Binding Cassette Transporters Blood Glucose  
 Calcium Carbachol Cell Respiration Cells, Cultured Cercopithecus aethiops Cholinergic Fibers Cloning, Molecular COS Cells  
 Cyclic AMP Diabetes Complications Eating Exocytosis Female Fibrosis Gene Expression Gene Expression Regulation Genotype  
 Glucagon Glucagon-Like Peptide-1 Receptor Glucagon-Like Peptide 1 Glucose Glucose Clamp Technique Glucose Transporter Type 2  
 Glyburide Heart Atria Heart Ventricles Hormones Hyperinsulinism Hypoglycemia Hypoglycemic Agents Imidazoles Indoles Insulin  
 Insulin-Secreting Cells Insulin Secretion Intracellular Fluid In Vitro Techniques Islets of Langerhans KATP Channels Kinetics  
 Large-Conductance Calcium-Activated Potassium Channel beta Subunits Loss of Heterozygosity Male Membrane Potentials Mice  
 Mice, Inbred C57BL Mice, Inbred Strains Mice, Knockout Mice, Transgenic Models, Biological Molecular Sequence Data  
 Multidrug Resistance-Associated Proteins Myocardial Infarction Myocardial Reperfusion Injury Myocarditis Myocytes, Cardiac Oocytes  
 Oxygen Consumption Patch-Clamp Techniques Peptide Fragments Perfusion Phosphodiesterase Inhibitors Potassium Channel Blockers  
 Potassium Channels Potassium Channels, Inwardly Rectifying Promoter Regions, Genetic Protein Precursors Receptors, Drug  
 Receptors, Glucagon Recombinant Proteins Reference Values RNA, Messenger Second Messenger Systems Signal Transduction  
 Sulfonylurea Compounds Sulfonylurea Receptors Ventricular Dysfunction, Left Xenopus laevis

## Attachments

 [bc4021079001.jpeg](#) - Added on July 27, 2010 at 9:35 AM by Jill Lindner

see [labnodes.vanderbilt.edu/resource/file/cnzpv2mq.jpeg](http://labnodes.vanderbilt.edu/resource/file/cnzpv2mq.jpeg)



 [sur\\_neo\\_wt\\_protocol\\_1\\_w.doc](#) - Added on July 27, 2010 at 2:30 PM by Jill Lindner

PCR protocol for genotyping mice