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TRM: Mouse Models of Cryopyrin Associated Periodic Syndromes (Nlrp3tm1Hhf, Nlrp3tm2Hhf, Nlrp3tm3Hhf)

Tech ID: 19891 / UC Case 2010-090-0

BRIEF DESCRIPTION

Cryopyrin (NALP3) mediates formation of the inflammasome, a protein complex responsible for cleavage of pro-IL-1 β to its active form. Mutations in the cryopyrin gene, *NLRP3*, cause the autoinflammatory disease spectrum: cryopyrin-associated periodic syndromes (CAPS). The central role of IL-1 β in CAPS is supported by the remarkable response to IL-1 targeted therapy.

TECHNOLOGY DESCRIPTION

Researchers from UC San Diego describe three different knock-in/out mice, each with a specific mutation associated with different disease phenotypes relating to cryopyrin associated periodic syndroms: familial cold autoinflammatory syndrome, Muckle Wells Syndrome and neonatal onset multisystem inflammatory disease.

These knock-in mice NIrp3tm1Hhf (also known as NIrp3^{A350VneoR}) contain a floxed neomycin cassette (neoR) in opposite orientation to a mutated *NIrp3* gene resembling the human mutation associated with Muckle-Wells syndrome. When bred to mice that express Cre recombinase to delete the floxed-neoR, the mutant gene is expressed in *cre*-expressing tissues of the offspring. These mice may be useful in studying the role of cryopyrin in the regulation of autoinflammatory diseases. (Jackson Lab Cat. No. 017969)

These NIrp3tm2Hhf (also known as *NIrp3^{L351PneoR}*) mice serve as a constitutive knockout of the *NIrp3* gene. In the presence of cre recombinase however, a transcript encoding a L351P mutation is produced. This line may be useful in studying the role of cryopyrin in the regulation of autoinflammatory diseases. (Jackson Lab Cat. No. 017970)

These *NIrp3*^{tm3Hhf} (or *NIrp3*^{D301NneoR}) knock-in mice serve as a constitutive knock-out of the *NIrp3* gene. This line may be useful in studying the role of cryopyrin in the regulation of autoinflammatory diseases. (Jackson Lab Cat. No. 017971)

STATE OF DEVELOPMENT

The mice are designated Tangible Research Material (TRM). A complete description, including genotyping, phenotyping, etc is found at :The Jackson Lab cat. No. 017969 <u>https://www.jax.org/strain/017969; https://www.jax.org/strain/017970;</u> https://www.jax.org/strain/017971

SUGGESTED USES

These mouse models allow genetic studies of the human disease as well as to study pharmacologic compounds that may be useful for treatment.

Academic and non-profit institutions please order directly from The Jackson Laboratory. Commercial entities require a license from

UC San Diego contact (https://innovation.ucsd.edu/contact/).

RELATED MATERIALS

CONTACT

University of California, San Diego Office of Innovation and Commercialization innovation@ucsd.edu tel: 858.534.5815.



OTHER INFORMATION

KEYWORDS Mice, NLRP3 Inflammasome, cryopyrin, innate immunity, PAMP, caspase-1

CATEGORIZED AS

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RELATED CASES 2010-090-0

Brydges SD, Mueller JL, McGeough MD, Pena CA, Misaghi A, Gandhi C, Putnam CD, Boyle DL, Firestein GS, Horner AA, Soroosh P, Watford WT, O'Shea JJ, Kastner DL, Hoffman HM. Inflammasome-mediated disease animal models reveal roles for innate but not adaptive immunity. Immunity. 2009 Jun 19;30(6):875-87. doi: 10.1016/j.immuni.2009.05.005. Epub 2009 Jun 4. - 06/19/2009

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