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

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BRIEF DESCRIPTION

Cryopyrin (NALP3) mediates formation of the inflammasome, a protein complex responsible for cleavage of pro-IL-1β 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-1β 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 Nlrp3^{A350VneoR}) contain a floxed neomycin cassette (neoR) in opposite orientation to a mutated *Nlrp3* 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 Nlrp3tm2Hhf (also known as *Nlrp3*^{L351PneoR}) mice serve as a constitutive knockout of the *Nlrp3* 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 *Nlrp3*^{tm3Hhf} (or *Nlrp3*^{D301NneoR}) knock-in mice serve as a constitutive knock-out of the *Nlrp3* 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 https://www.jax.org/strain/017969; https://www.jax.org/strain/017970; 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

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OTHER INFORMATION

KEYWORDS

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

CATEGORIZED AS

- **Medical**
  - Research Tools
- **Research Tools**
  - Animal Models
  - Reagents

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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