

Resource Summary Report

Generated by [RRID](#) on Jul 30, 2026

B6;129-Gaa^{tm1Rabn/J}

RRID:IMSR_JAX:004154

Type: Organism

Proper Citation

RRID:IMSR_JAX:004154

Organism Information

URL: <https://www.jax.org/strain/004154>

Proper Citation: RRID:IMSR_JAX:004154

Description: Mus musculus with name B6;129-Gaa^{tm1Rabn/J} from IMSR.

Species: Mus musculus

Notes: gene symbol note: glucosidase; alpha; acid; mutant stock: Gaa

Affected Gene: glucosidase; alpha; acid

Genomic Alteration: targeted mutation 1; Nina Raben

Catalog Number: JAX:004154

Database: JAX Mice and Services

Database Abbreviation: JAX

Availability: live

Organism Name: B6;129-Gaa^{tm1Rabn/J}

Record Creation Time: 20250513T053643+0000

Record Last Update: 20260725T044352+0000

Ratings and Alerts

No rating or validation information has been found for B6;129-Gaa^{tm1Rabn/J}.

No alerts have been found for B6;129-Gaa^{tm1Rabn/J}.

Data and Source Information

Source: [Integrated Animals](#)

Source Database: JAX Mice and Services

Usage and Citation Metrics

We found 11 mentions in open access literature.

Listed below are recent publications. The full list is available at [RRID](#).

Marcó S, et al. (2025) Rat models of musculoskeletal lysosomal storage disorders and their role in pre-clinical evaluation of gene therapy approaches. Mammalian genome : official journal of the International Mammalian Genome Society, 36(2), 488.

Rohm M, et al. (2023) Dysregulation of Metabolism and Proteostasis in Skeletal Muscle of a Presymptomatic Pompe Mouse Model. Cells, 12(12).

Nagy JA, et al. (2022) Altered electrical properties in skeletal muscle of mice with glycogen storage disease type II. Scientific reports, 12(1), 5327.

Aguilar-González A, et al. (2022) Isogenic GAA-KO Murine Muscle Cell Lines Mimicking Severe Pompe Mutations as Preclinical Models for the Screening of Potential Gene Therapy Strategies. International journal of molecular sciences, 23(11).

Costa-Verdera H, et al. (2021) Hepatic expression of GAA results in enhanced enzyme bioavailability in mice and non-human primates. Nature communications, 12(1), 6393.

Piras G, et al. (2020) Lentiviral Hematopoietic Stem Cell Gene Therapy Rescues Clinical Phenotypes in a Murine Model of Pompe Disease. Molecular therapy. Methods & clinical development, 18, 558.

Huang JY, et al. (2020) CRISPR-Cas9 generated Pompe knock-in murine model exhibits early-onset hypertrophic cardiomyopathy and skeletal muscle weakness. Scientific reports, 10(1), 10321.

Colella P, et al. (2020) Gene therapy with secreted acid alpha-glucosidase rescues Pompe disease in a novel mouse model with early-onset spinal cord and respiratory defects. EBioMedicine, 61, 103052.

Colella P, et al. (2019) AAV Gene Transfer with Tandem Promoter Design Prevents Anti-transgene Immunity and Provides Persistent Efficacy in Neonate Pompe Mice. Molecular therapy. Methods & clinical development, 12, 85.

Doyle BM, et al. (2019) AAV Gene Therapy Utilizing Glycosylation-Independent Lysosomal Targeting Tagged GAA in the Hypoglossal Motor System of Pompe Mice. Molecular therapy. Methods & clinical development, 15, 194.

Yambire KF, et al. (2019) Impaired lysosomal acidification triggers iron deficiency and inflammation in vivo. eLife, 8.