

Resource Summary Report

Generated by [RRID](#) on Jul 31, 2026

B6.129S4-Ndufs4^{tm1.1Rpa/J}

RRID:IMSR_JAX:027058

Type: Organism

Proper Citation

RRID:IMSR_JAX:027058

Organism Information

URL: <https://www.jax.org/strain/027058>

Proper Citation: RRID:IMSR_JAX:027058

Description: Mus musculus with name B6.129S4-Ndufs4^{tm1.1Rpa/J} from IMSR.

Species: Mus musculus

Notes: gene symbol note: NADH:ubiquinone oxidoreductase core subunit S4; mutant strain|congenic strain: Ndufs4

Affected Gene: NADH:ubiquinone oxidoreductase core subunit S4

Genomic Alteration: targeted mutation 1.1; Richard D Palmiter

Catalog Number: JAX:027058

Database: JAX Mice and Services

Database Abbreviation: JAX

Availability: live

Organism Name: B6.129S4-Ndufs4^{tm1.1Rpa/J}

Record Creation Time: 20250513T053759+0000

Record Last Update: 20260725T044505+0000

Ratings and Alerts

No rating or validation information has been found for B6.129S4-Ndufs4^{tm1.1Rpa/J}.

No alerts have been found for B6.129S4-Ndufs4^{tm1.1Rpa/J}.

Data and Source Information

Source: [Integrated Animals](#)

Source Database: JAX Mice and Services

Usage and Citation Metrics

We found 15 mentions in open access literature.

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

Blume SY, et al. (2025) HypoxyStat, a small-molecule form of hypoxia therapy that increases oxygen-hemoglobin affinity. *Cell*, 188(6), 1580.

Meisel JD, et al. (2024) Hypoxia and intra-complex genetic suppressors rescue complex I mutants by a shared mechanism. *Cell*, 187(3), 659.

Hernansanz-Agustín P, et al. (2024) A transmitochondrial sodium gradient controls membrane potential in mammalian mitochondria. *Cell*.

Willemse L, et al. (2023) A ketogenic diet alters mTOR activity, systemic metabolism and potentially prevents collagen degradation associated with chronic alcohol consumption in mice. *Metabolomics : Official journal of the Metabolomic Society*, 19(5), 43.

Baik AH, et al. (2023) Oxygen toxicity causes cyclic damage by destabilizing specific Fe-S cluster-containing protein complexes. *Molecular cell*, 83(6), 942.

Avrutsky MI, et al. (2022) Noninvasive Ophthalmic Imaging Measures Retinal Degeneration and Vision Deficits in *Ndufs4*^{-/-} Mouse Model of Mitochondrial Complex I Deficiency. *Translational vision science & technology*, 11(8), 5.

van der Walt G, et al. (2021) Sub-Cellular Metabolomics Contributes Mitochondria-Specific Metabolic Insights to a Mouse Model of Leigh Syndrome. *Metabolites*, 11(10).

Bertan F, et al. (2021) Comparative analysis of CI- and CIV-containing respiratory supercomplexes at single-cell resolution. *Cell reports methods*, 1(1), 100002.

Shil SK, et al. (2021) *Ndufs4* ablation decreases synaptophysin expression in hippocampus. *Scientific reports*, 11(1), 10969.

Perry EA, et al. (2021) Tetracyclines promote survival and fitness in mitochondrial disease models. *Nature metabolism*, 3(1), 33.

Liu S, et al. (2021) Glycerol-3-phosphate biosynthesis regenerates cytosolic NAD⁺ to alleviate mitochondrial disease. *Cell metabolism*, 33(10), 1974.

Kagawa Y, et al. (2020) Mitochondrial dysfunction in GnRH neurons impaired GnRH production. *Biochemical and biophysical research communications*, 530(1), 329.

Bisbach CM, et al. (2020) Succinate Can Shuttle Reducing Power from the Hypoxic Retina to the O₂-Rich Pigment Epithelium. *Cell reports*, 31(5), 107606.

Yang L, et al. (2020) Serine Catabolism Feeds NADH when Respiration Is Impaired. *Cell metabolism*, 31(4), 809.

Bolea I, et al. (2019) Defined neuronal populations drive fatal phenotype in a mouse model of Leigh syndrome. *eLife*, 8.