# Ndufs4 Knockout Mice for Modeling Leigh Syndrome

### Biomaterial – Mouse

### Biomaterial Description

The Ndufs4 Knockout (KO) Mice are an innovative model for studying mitochondrial complex I deficiency, specifically designed to model the severe mitochondrial disease known as Leigh syndrome. These mice are genetically engineered to lack the Ndufs4 gene, which encodes a critical subunit of the NADH:ubiquinone oxidoreductase (Complex I) in the mitochondrial electron transport chain. Defects in mitochondrial complex I are the most prevalent among mitochondrial disorders. This deficiency leads to a range of neurometabolic symptoms, providing a robust model for studying mitochondrial disorders and their systemic effects.

### Applications

-Drug Development: Use these mice to screen and evaluate the efficacy of potential drugs targeting mitochondrial dysfunction and related neurodegenerative conditions.

-Gene Therapy: Investigate gene therapy approaches to correct mitochondrial deficiencies and improve neurological outcomes.

-Metabolic Studies: Explore the metabolic pathways affected by mitochondrial dysfunction and identify potential metabolic interventions.

-Aging Studies: Study the impact of mitochondrial dysfunction on aging and test interventions that could extend healthy lifespan.

### Advantages

-Versatile Research Tool: These mice can be used to study mitochondrial dysfunction, neurodegenerative diseases, and metabolic disorders.

-Therapeutic Development: Ideal for preclinical testing of novel therapeutic interventions aimed at mitigating mitochondrial diseases.

-Conditional KO Model Available.

### Distributor Information

Non-Exclusive License available.

### Technology ID

INV 45257

### Category

Research Tools/Biological Materials/Mouse

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### Learn more

Shipped from JAX:

JAX Stock #026963 – Conditional Cre Mice

JAX Stock #027058 – KO mice

## References


1. Kruse, S. E., Watt, W. C., Marcinek, D. J., Kapur, R. P., Schenkman, K. A., Palmiter, R. D.(2008) , https://pmc.ncbi.nlm.nih.gov/articles/PMC2593686/, https://www.sciencedirect.com/journal/cell-metabolism, 7, 312-320