

**BTBR-*Pah*<sup>enu2</sup>/J**  
Stock No: **002232** | PAH<sup>enu2</sup>

 [Chemically Induced Mutation](#)

Sized to accommodate orders of up to 10 or more with age range. Ask Customer Service for details.

PLACE ORDER

[Email](#) [Download PDF](#) [Help](#)

gene and are a useful model of severe hyperphenylalanemia.

### Donating Investigator

Dr. Alexandra Shedlovsky, University of Wisconsin , Madison

READ MORE +

## GENETIC OVERVIEW

### Genetic Background

002282 BTBR *T*<sup>+</sup> *Itpr3*<sup>fl</sup>/J

### Generation

N2p+F9  
(2020-11-03 00:00:00)

### *Pah*<sup>enu2</sup>

#### Alele Type

Chemically induced (ENU)

#### Gene Symbol

*Pah*

#### Gene Name

phenylalanine hydroxylase

VIEW GENETICS

## RESEARCH APPLICATIONS

Metabolism Research

Neurobiology Research

Mouse/Human Gene Homologs

VIEW ALL RESEARCH APPLICATIONS

## BASE PRICE

Starting at:

---

\$255.00 Domestic price for female 4-week

---

510.00 Domestic price for breeder pair

---

V I E W   P R I C E   L I S T

### Details

#### Detailed Description

Phenylketonuria (PKU) in humans results from a deficiency in phenylalanine hydroxylase (PAH). If untreated, this condition results in severe and irreversible mental retardation.

Homozygous *Pah*<sup>enu2</sup> mutant mice show severe hyperphenylalanemia. They typically have small heads and display behavioral abnormalities starting at about 2 weeks and lasting into adulthood. Levels of serum phenylalanine are elevated 10-20 fold and urinary ketone concentrations are significantly increased in adults. As in human patients, the mutant mice are hypopigmented unless maintained on a low phenylalanine diet. Under conditions of elevated phenylalanine (e.g. at 25 g/dl in drinking water), *Pah*<sup>enu2</sup> animals sicken within days.

The coat color of the background strain, BTBR *T*<sup>+</sup> *Itpr3*<sup>tf</sup> /J (Stock No. [002282](#)) is black and tan (*a*<sup>t</sup> / *a*<sup>t</sup>). This strain is also homozygous for the gene tufted (*tf/tf*) resulting in various molting patterns in the mouse coat. These effects, limited to the mouse coat, may make the mice appear malformed.

In The Jackson Laboratory colony, homozygous prewean pups are black and tan. They molt at 3-4 weeks of age, but when the fur grows back, the mice are brown and tan. When breeders lose their fur at an older age, it doesn't generally grow back. Our colony shows a very low percentage of mice that appear black and tan after 6-7 weeks old.

Female homozygous mice are fertile but do not rear their young when maintained on a standard mouse diet; none of their pups survive beyond several hours. Like maternal PKU in humans, this effect depends entirely upon the genotype of the mother.

Of note, the *Pah*<sup>enu2</sup> mutation is also available on a congenic C57BL/6J background as Stock No. [029218](#).

---

#### Development

---

#### Control Suggestions

---

#### Selected References

---

### Genetics

---

+ [Pah<sup>enu2</sup>](#)

---

## – Disease/Phenotype

+ [Disease Terms](#)

---

+ [Research Areas By Phenotype](#)

---

+ [Mammalian Phenotype Terms by Genotype](#)

---

+ [References](#)

---

## – Technical Support

C O N T A C T   T E C H N I C A L   S U P P O R T

### Genotyping Protocols

Sanger sequencing:[Pah](#)

End Point Analysis:[Pah Alt 1](#)

[Genotyping resources and troubleshooting](#)

### Dietary Information

LabDiet® 5K52 formulation (6% fat)

### Breeding Considerations

Crosses of heterozygous females with homozygous males are recommended. Homozygous females are fertile but do not rear their young when maintained on a standard mouse diet; none of their pups survive beyond several hours.

*Technician's Note:* This strain breeds well and does not require any special husbandry. The Jackson Laboratory maintains their colony on a 6% fat diet. The average litter size is 8-12 pups. The expected coat color from breeding is Black with Brown.

### [Additional Breeding and Husbandry Support](#)

#### Mating System

Heterozygote x Homozygote

#### Appearance

black and tan, tufted

Related Genotype:  $a^t/a^t +^T/+^T$   $tf/tf$

Homozygous prewean pups are black and tan. They molt at 3-4 weeks of age, but when the fur grows back, the mice are brown and tan. When breeders lose their fur at an older age, it doesn't generally grow back.

## Citation

When using the PAH<sup>enu2</sup> mouse strain in a publication, please [cite the originating article\(s\)](#) and include JAX stock #002232 in your Materials and Methods section.

## Animal Health Reports

[Facility Barrier Level Descriptions](#)

 [AX11 \(Maximum\)](#)

## ➔ Pricing & Availability



Available Now

Sized to accommodate orders of up to 10 or more with age range. Ask Customer Service for details.

## Domestic **International**

Pricing effective for USA, Canada and Mexico shipping destinations

LIVE MOUSE			
AGE	SEX	GENOTYPE	PRICE
4 weeks	Female	Heterozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Heterozygous for Pah <sup>enu2</sup>	\$255.00
4 weeks	Female	Homozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Homozygous for Pah <sup>enu2</sup>	\$255.00
5 weeks	Female	Heterozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Heterozygous for Pah <sup>enu2</sup>	\$255.00
5 weeks	Female	Homozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Homozygous for Pah <sup>enu2</sup>	\$255.00
6 weeks	Female	Heterozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Heterozygous for Pah <sup>enu2</sup>	\$255.00
6 weeks	Female	Homozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Homozygous for Pah <sup>enu2</sup>	\$255.00
7 weeks	Female	Heterozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Heterozygous for Pah <sup>enu2</sup>	\$255.00
7 weeks	Female	Homozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Homozygous for Pah <sup>enu2</sup>	\$255.00
8 weeks	Female	Heterozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Heterozygous for Pah <sup>enu2</sup>	\$255.00
8 weeks	Female	Homozygous for Pah <sup>enu2</sup>	\$255.00

	SEX	Homozygous for Pah <sup>enu2</sup>	\$255.00
9 weeks	Female	Homozygous for Pah <sup>enu2</sup>	\$255.00
	Male	Homozygous for Pah <sup>enu2</sup>	\$255.00

BREEDER PAIR			
SEX	GENOTYPE		PRICE
Female	Heterozygous for Pah <sup>enu2</sup>		\$510.00
Male	Homozygous for Pah <sup>enu2</sup>		

RELATED PRODUCTS AND SERVICES		
<a href="#">Frozen Mouse Embryo</a>	BTBR-Pah <sup>enu2</sup> /J Frozen Embryos	\$2595.00

## PAYMENT TERMS AND CONDITIONS

Terms are granted by individual review and stated on the customer invoice(s) and account statement. These transactions are payable in U.S. currency within the granted terms. Payment for services, products, shipping containers, and shipping costs that are rendered are expected within the payment terms indicated on the invoice or stated by contract. Invoices and account balances in arrears of stated terms may result in The Jackson Laboratory pursuing collection activities including but not limited to outside agencies and court filings.

## THE JACKSON LABORATORY'S GENOTYPE PROMISE

The Jackson Laboratory has rigorous genetic quality control and mutant gene genotyping programs to ensure the genetic background of JAX® Mice strains as well as the genotypes of strains with identified molecular mutations. JAX® Mice strains are only made available to researchers after meeting our standards. However, the phenotype of each strain may not be fully characterized and/or captured in the strain data sheets. **Therefore, we cannot guarantee a strain's phenotype will meet all expectations.** To ensure that JAX® Mice will meet the needs of individual research projects or when requesting a strain that is new to your research, we suggest ordering and performing tests on a small number of mice to determine suitability for your particular project. We do not guarantee [breeding performance](#) and therefore suggest that investigators order more than one breeding pair to avoid delays in their research.

## Terms Of Use

### TERMS OF USE

[General Terms and Conditions](#)

QUESTIONS ABOUT TERMS OF USE

## LICENSING INFORMATION

Phone: 207-288-6470

## ☰ Related Strains

All

By Allele

By Gene

By Collection



### DO YOU NEED BALB/c MICE?

Rely on JAX to provide the models you need, when you need them.

[LEARN MORE](#)



CONTACT



DONATE



SUBSCRIBE

[JAX HOME](#) [CAREERS](#) [LEGAL INFORMATION](#)


[RESEARCH CENTERS](#) [MOUSE GENOME INFORMATICS](#)

*Leading the search for*

# TOMORROW'S CURES



©2021 THE JACKSON LABORATORY

Choose other country or region 

 [E](#) [E](#) [E](#) [D](#) [B](#)

Did you find what you were looking for?

Yes  No