These mice carry a spontaneous deletion that results in polycystic kidney disease and other dysmorphic features with death by 3 weeks after birth. This strain offers a model for human Meckel Syndrome Type 3.
<table>
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<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tbody>
<tr>
<td>Spontaneous</td>
<td>a</td>
<td>nonagouti</td>
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**bpck**

<table>
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<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tbody>
<tr>
<td>Spontaneous</td>
<td>bpck</td>
<td>bilateral polycystic kidney deletion region</td>
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</table>

Mice homozygous for the *bpck* deletion develop bilateral polycystic kidneys and die by 3 weeks of age. There is an increased incidence of hydrocephalus. Homozygotes can be identified by their smaller size and swollen abdomens. At 2 weeks of age elevated blood urea nitrogen is found. Ovaries and testes are smaller than normal and progression of maturing cells from spermatocytes to spermatids is disorganized at 3 weeks of age. The primary cilia on the kidney proximal tubule epithelial cells are dysmorphic and vary in length at birth and by 14 days of age cilia are significantly longer than normal. Through overlapping BAC rescues the polycystic kidney disease and hydrocephalus has been traced to the absence of the *Tmem67* gene. Although lacking some of the ancillary phenotypes associated with Meckel Syndrome Type 3 in humans, this deletion offers a model for that disease.
Genotyping Protocols
QPCR: bpck QPCR
Genotyping resources and troubleshooting

Citation
When using the B6C3Fe \textit{a}/\textit{a}-bpck/J mouse strain in a publication, please cite the originating article(s) and include JAX stock #008044 in your Materials and Methods section.

Production of mice from cryopreserved embryos or sperm occurs in a maximum barrier room, \textit{G200}

Pricing & Availability
Typically mice are recovered in 10-14 weeks. Contact Customer Service to place an order or for more information.

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<tr>
<th>SERVICE/PRODUCT</th>
<th>DESCRIPTION</th>
<th>PRICE</th>
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<tr>
<td>Cryo Recovery</td>
<td>Heterozygous or Wild-type for bpck</td>
<td>$2,854.50</td>
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</table>
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**All Related Strains**