Agitans-like 3 Jackson; a neurological remutation to agitans-like on Chromosome 14.

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Mutation (allele) symbol: *agil*^{3J}

Mutation (allele) name: agitans-like 3 Jackson

Gene symbol: *agil*

Strain of origin: CBA/J

Current strain name: CBA/J-*agil*^{3J}/GrsrJ

Stock #014084 (jaxmice.jax.org)

Phenotype categories: neurological

Abstract
We have identified a recessive remutation to agitans-like (*agil*) that causes affected mice to exhibit the same abnormal gait as the original agitans-like mutation. The phenotype can be identified when the affected mice are about 2 weeks of age. A direct test for allelism produced affected animals proving allelism with *agil*.

Origin and Description
The new mutation was found by Helen Tracey in a production colony of CBA/J mice at The Jackson Laboratory in 2007 and was initially identified by its wobbling gait and the retraction of its back legs toward the body when picked up by the tail. This mutant subline has been maintained by progeny test. Female and male homozygotes have not bred. Homozygotes are smaller in size than their littermates and usually do not live to adulthood.

Genetic Analysis
A direct test for allelism was performed by mating a female mouse heterozygous for the *agil* mutation to a male mouse heterozygous for the new mutation. This mating produced three affected homozygotes out of fifteen total progeny born, proving allelism to the original *agil* mutation. This new remutation has been named agitans-like 3 Jackson (*agil*^{3J}).

Pathology
A routine pathological screen[1] showed dystrophic axons, mild degeneration of white matter in the spinal cord, and atrophic enterocytes in the small intestine in one male homozygote at 3 weeks of age. There was no sperm in the testis of either the 3-week-old homozygote or the wild type sibling control. One female at 2 weeks of age showed focal

necrosis in the pancreas but the axonal dystrophy was not found. Retinal degeneration was detected in both homozygotes and heterozygotes as this is a strain characteristic of CBA/J.

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[1]**Standard Histology Protocol used in The Mouse Mutant Resource**
For fixation of tissues, mice were deeply anesthetized with tribromoethanol (avertin) until they no longer displayed a withdrawal reflex in the hind limbs and then perfused intracardially with Bouin's fixative following a flush of the vasculature with saline solution. After soaking in Bouin's for one week to demineralize bones, tissues were dissected. Six segments of spine with axial muscles and spinal cord in situ, representing cervical, thoracic and lumbar spinal segments, were dissected. The brain was removed and sliced into 6 cross sectional pieces at the levels of olfactory lobes, frontal cortex, striatum, thalamus, midbrain, rostral and caudal medulla with cerebellum. Midsagittal slices of hind leg through the knees were prepared. Slices of basal skull through the pituitary and inner ears were taken. Both eyes, salivary glands and submandibular lymph node, trachea plus thyroid and sometimes parathyroid were removed and cassetted. A longitudinal slice of skin from the back was removed. The thymus, slices of lung, and a longitudinal slice of heart were cassetted. Similarly slices of liver through gall bladder, kidney with adrenal attached, pancreas and spleen were prepared. The stomach was sliced longitudinally to include both squamous and glandular portions. Loops of small intestine from 3 levels and slices of large intestine and cecum were removed, as were slices of urinary bladder. The whole uterus, with ovaries attached, was taken. In males testes were sliced longitudinally. The accessory male organs including seminal vesicles, coagulating gland and prostate were removed en block. Altogether in most cases all tissue fit into a total of 10 cassettes. The cassettes were processed in an automatic tissue processor to dehydrate tissues which were then embedded in paraffin. Six micron sections were cut and stained with hematoxylin and eosin (H&E). Sections of brain and spinal cord in vertebral bones also were stained with luxol fast blue (LFB) for myelin and cresylecht violet (CV) for cellular detail.