

Fragile X Syndrome

Fmr1-KO Mouse Model

Fmr1-KO mice

The mouse model contains a neomycin resistance cassette substituting exon 5 of the fragile X mental retardation syndrome 1 (*Fmr1*) gene. The knockdown causes an increase in the number of CGG repeats that lead to hypermethylation of the *Fmr1* gene and therefore inhibiting FMR protein production.

At the age of 7 weeks mice start to present core and secondary phenotypic traits of Fragile X syndrome such as:

- Hyperactivity
- Altered anxiety levels
- Repetitive behavior
- Social behavior deficits
- Vocalization deficits

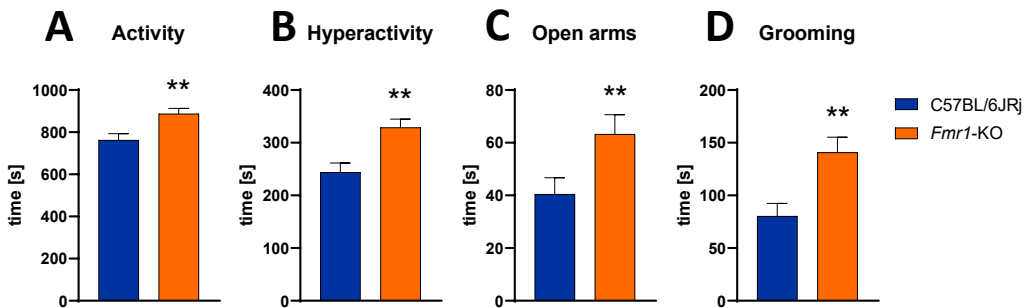


Figure: Activity, hyperactivity, anxiety, and repetitive behavior of male *Fmr1*-KO mice at the age of 7 weeks. Activity (A) and hyperactivity (B) measured in the open field test, time spent in open arms of the open field test (C), and time spent grooming in the auto-grooming test (D) of *Fmr1*-KO compared to C57BL/6Jrj mice. n = 15 per group. Unpaired t-test; Mean + SEM; **p<0.01.

References:

Bakker et al. 1994. *Fmr1* knockout mice: a model to study fragile x mental retardation. The Dutch-Belgium Fragile X Consortium. *Cell* 78(1):23-33.