1. Introduction

Transgenic mice overexpressing the mutant human form of SOD1 is widely recognized to rapidly develop progressive motor neuron disease that strongly resembles to ALS. At about 2.5 - 3 months of age, indeed, SOD1 mice demonstrate progressively muscle weakness along with nerve dysfunction indicated by a decrease and an increase in the amplitude and latency of compound muscle action potential (CMAP), respectively. By 4 months of age, death by paralysis and respiratory failure appears. Riluzole, the only therapy available for the treatment of ALS patients shows a modest extension of survival and no effect on motor function in the clinic. Similar profile of results is observed following treatment of transgenic SOD1 mice with riluzole.

2. Compound testing

Compound testing addresses the effect of chronic treatment (typically starting at day 60 of age) on:
- survival or
- survival and nerve function (electrophysiology measures) or
- survival, nerve function and motor performance (behavioural measures).

Cumulative survival curve of transgenic SOD1 mice. Motor performance of transgenic mice as assessed by the rotarod test.
Motor performance of transgenic mice as assessed by the grid test.

Motor performance of transgenic mice as assessed by the string test.

Change in the amplitude of CMAP in transgenic SOD1 mice.

Change in the latency of CMAP in transgenic SOD1 mice.

3. References


