

APPENDIX

Beers/Appel/Simpson/Henkel (BASH) Mouse ALS Score*

0 = Healthy

1 = Tremulousness

2 = Gait Abnormalities

- waddle
- lower hindquarters
- limp tail

3 = Hindlimb Weakness/Paresis

- foot drag
- trouble with hindlimb extension

4 = Weight Loss

- $\geq 10\%$ loss of maximum weight

5 = Spasticity

- one or both hindlimbs spastic
- incontinence

6 = Paralysis

- one or both hindlimb paralyzed

AND

- able to ambulate around cage with forelimbs

7 = End Stage

- inability to ambulate around cage

8 = Euthanasia

*Stages of disease progression. Shown are the criteria used to score mSOD1^{G93A}/ PU.1^{+/+}, mSOD1^{G93A}/PU.1^{+/-}, or mSOD1^{G93A}/PU.1^{-/-} mice as they develop an ALS-like disease.