Supplemental Figure 1.

A

B

mRNA levels (% of Control)

Log Conc. (µM)

Inactive ASO

ASO 1

ASO 2

ASO 333611

SOD1 mRNA (% Vehicle)

Vehicle ASO 1

B

SOD1 mRNA (% Vehicle)

Vehicle ASO 1

***

Log Conc. (µM)

Vehicle ASO 1
Supplemental Figure 2.

A. Male Rats

- Inactive ASO
- aCSF
- ASO 333611
- ASO 1
- ASO 2

B. Male Rats

- Inactive ASO
- aCSF
- ASO 333611
- ASO 1
- ASO 2

C. Female Rats

- Inactive ASO
- aCSF
- ASO 333611
- ASO 1
- ASO 2

D. Female Rats

- Inactive ASO
- aCSF
- ASO 333611
- ASO 1
- ASO 2
Supplemental Figure 3.

A

B

Inactive ASO

ASO 1

--- 80 days ICV

--- 110 days ICV

--- 110 days ICV

--- 80 days ICV

% Survival

% Weight peak

Age (days)

Age (days)
Supplemental Figure 4.

![Graph showing SOD1 mRNA (% Vehicle) against dose (µg). The ED<sub>50</sub> is 43 µg.]
Supplemental Figure 5.

A

<table>
<thead>
<tr>
<th>Concentration (ng/ml)</th>
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<tr>
<td>pNFH serum (aCSF)</td>
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<tr>
<td>pNFH CSF (A)</td>
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<tr>
<td>miR-218 CSF (ASO 2)</td>
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B

<table>
<thead>
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<th>pNFH serum vs miR-218</th>
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<td>p = 0.053</td>
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<tr>
<td>R² = 0.65</td>
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C

<table>
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<th>pNFH CSF vs miR-218</th>
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</thead>
<tbody>
<tr>
<td>p = 0.119</td>
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<tr>
<td>R² = 0.49</td>
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</tbody>
</table>
Supplemental Figure 6.

A

ASO 1: 3' -TCGACATCTTTACATAGGAC- 5'

NHP SOD1: UGCUAGCUGUAGAAUGUAUCUGAUU

B

![Graph showing SOD1 mRNA levels vs. log concentration (µM)]

- ASO 1
- Inactive ASO

Log Concentration (µM)

SOD1 mRNA (% control)
Supplemental Figure 7.

Lumbar cord

Thoracic cord

Cervical cord

Pons

Frontal cortex

Hippocampus

EC$_{50}$ = 22.4

EC$_{50}$ = 26.4

EC$_{50}$ = 24.1

EC$_{50}$ = 28.9

EC$_{50}$ = 22.5

EC$_{50}$ = 17.4