ALS: Lou Gehrig's Disease

Why has progress on this disease been so slow?
140 years and counting since diagnosis

Normal

Normal health condition

DISEASED


Disease Mechanisms

Transgenic Mouse

What does Mck do?

How is Mck used?

TABLE 1. CAT activity in tissues of adult
−3300MCK-CAT transgenic mice

<table>
<thead>
<tr>
<th>Tissue</th>
<th>CAT activity (nanomoles of protein ± SD) in given mouse line**</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1</td>
</tr>
<tr>
<td>Skeletal muscle</td>
<td>3 x 10^(-3)</td>
</tr>
<tr>
<td>Cardiac muscle</td>
<td>1 x 10^(-3)</td>
</tr>
<tr>
<td>Lung</td>
<td>6 x 10^(-3)</td>
</tr>
<tr>
<td>Brain</td>
<td>1 x 10^(-3)</td>
</tr>
<tr>
<td>Testis</td>
<td>2 x 10^(-3)</td>
</tr>
<tr>
<td>Uterus</td>
<td>1 x 10^(-3)</td>
</tr>
<tr>
<td>Blood</td>
<td>6 x 10^(-3)</td>
</tr>
<tr>
<td>Kidney</td>
<td>1 x 10^(-3)</td>
</tr>
<tr>
<td>Liver</td>
<td>8 x 10^(-3)</td>
</tr>
<tr>
<td>Spleen</td>
<td>8 x 10^(-3)</td>
</tr>
</tbody>
</table>

* A unit of CAT activity is defined as the amount sufficient to acetylate 1 
μmol of chloramphenicol per min (17). Data are from two to eight adult mice.
Standard deviations are given only for data averaged from tissues of eight
mice. See legend to Fig. 1 for description of the transgene −3300MCK-CAT.
Transgenic mouse line 1 contains 3 to 5 copies of the transgene −3300MCK,
transgenic mouse line 2 contains 1 to 3 copies of the transgene −3300MCK.

** Superficial peritoneum muscle.
--- Not determined.

Is Muscle Function Improved?


Muscle Atrophy?

Neuronal Degeneration?


Motor Neurons?

Number of motor neurons at endstage
(Spinal cord ventral horn)

D

SOD1G93A Non Tg
m=3
SOD1G93A MCK-PGC-1α
m=3
PDC-1α
m=3
Survival?

What did we learn?