

### Semi-quantitative scoring of PrP<sup>Sc</sup> distribution in the brain

| Genotype                       | Goat# | Olfactory lobe | Frontal cortex | N. caud/putamen | Parietal cortex | Thalamus | Hippocampus | Midbrain | Cerebellum | Pons | Medulla oblongata | Trigeminal ganglion |
|--------------------------------|-------|----------------|----------------|-----------------|-----------------|----------|-------------|----------|------------|------|-------------------|---------------------|
| <i>PRNP</i> <sup>+/+</sup>     | 416   | 0.5            | 0.5            | 0.5             | 0.5             | 1.5      | 0.5         | 2.0      | 0.0        | 0.5  | 1.5               | 0.5                 |
| <i>PRNP</i> <sup>+/+</sup>     | 417   | 2.5            | 1.0            | 1.0             | 1.5             | 3.0      | 3.0         | 3.0      | 2.5        | 3.0  | 2.5               | 1.0                 |
| <i>PRNP</i> <sup>+/+</sup>     | 529   | 2.5            | 2.0            | 1.0             | 1.5             | 3.0      | 1.5         | 3.0      | 1.0        | 2.0  | 2.0               | 1.5                 |
| <i>PRNP</i> <sup>+/+</sup>     | 536   | 1.0            | 0.5            | 0.5             | 0.5             | 0.5      | 0.5         | 2.0      | 0.5        | 1.5  | 1.5               | 0.5                 |
|                                |       | 1.6            | 1.0            | 0.8             | 1.0             | 2.0      | 1.4         | 2.5      | 1.0        | 1.8  | 1.8               | 0.9                 |
| <i>PRNP</i> <sup>+/Ter</sup>   | 451   | 3.0            | 2.5            | 2.0             | 2.5             | 2.5      | 2.5         | 2.0      | 0.5        | 1.0  | 0.5               | NE                  |
| <i>PRNP</i> <sup>+/Ter</sup>   | 469   | NE             | 1.5            | 1.5             | 1.0             | 2.5      | 1.5         | 2.0      | 1.0        | 1.5  | 1.0               | 0.5                 |
| <i>PRNP</i> <sup>+/Ter</sup>   | 527   | 3.0            | 2.5            | 1.5             | 2.0             | 2.5      | 2.0         | 2.0      | 1.5        | 2.5  | 2.5               | 1.0                 |
| <i>PRNP</i> <sup>+/Ter</sup>   | 533   | NE             | 2.0            | 1.5             | 2.0             | 2.5      | 1.5         | 2.0      | 0.5        | 2.0  | 1.5               | 1.0                 |
|                                |       | 3.0            | 2.1            | 1.6             | 1.9             | 2.5      | 1.9         | 2.0      | 0.9        | 1.8  | 1.8               | 0.8                 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 413   | 0.0            | 0.0            | 0.0             | 0.0             | 0.0      | 0.0         | 0.0      | 0.0        | 0.0  | 0.0               | 0.0                 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 457   | 0.0            | 0.0            | 0.0             | 0.0             | 0.0      | 0.0         | 0.0      | 0.0        | 0.0  | 0.0               | 0.0                 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 476   | 0.0            | 0.0            | 0.0             | 0.0             | 0.0      | 0.0         | 0.0      | 0.0        | 0.0  | 0.0               | 0.0                 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 490   | 0.0            | 0.0            | 0.0             | 0.0             | 0.0      | 0.0         | 0.0      | 0.0        | 0.0  | 0.0               | 0.0                 |
|                                |       | 0.0            | 0.0            | 0.0             | 0.0             | 0.0      | 0.0         | 0.0      | 0.0        | 0.0  | 0.0               | 0.0                 |

Abbreviation: N. caud, Nucleus caudatus

### Semi-quantitative scoring of PrP<sup>Sc</sup> distribution in the spinal cord

| Genotype                       | Goat# | Cervical |     |     | Thoracal |     |     | Lumbal |     |     | Cauda equina |     |     |
|--------------------------------|-------|----------|-----|-----|----------|-----|-----|--------|-----|-----|--------------|-----|-----|
|                                |       | DH       | VH  | DRG | DH       | VH  | DRG | DH     | VH  | DRG | DH           | VH  | DRG |
| <i>PRNP</i> <sup>+/+</sup>     | 416   | 1.5      | 1.5 | 0.5 | 1.0      | 1.0 | 0.5 | 0.5    | 0.5 | NE  | 1.0          | 0.5 | 0.0 |
| <i>PRNP</i> <sup>+/+</sup>     | 417   | 3.0      | 2.5 | 2.0 | 2.0      | 1.0 | NE  | 3.0    | 2.0 | 2.0 | 2.5          | 0.5 | 1.0 |
| <i>PRNP</i> <sup>+/+</sup>     | 529   | 2.0      | 0.5 | 0.5 | 2.5      | 1.0 | 1.0 | 2.5    | 0.5 | 1.0 | 3.0          | 1.0 | 1.0 |
| <i>PRNP</i> <sup>+/+</sup>     | 536   | 1.5      | 1.0 | NE  | 1.0      | 0.5 | 0.5 | 0.5    | 0.5 | NE  | 1.5          | 0.5 | NE  |
|                                |       | 2.0      | 1.4 | 1.0 | 1.6      | 0.9 | 0.7 | 1.6    | 0.9 | 1.5 | 2.0          | 0.6 | 0.7 |
| <i>PRNP</i> <sup>+/Ter</sup>   | 451   | 3.0      | 1.5 | 1.0 | 2.5      | 1.0 | 0.0 | 2.0    | 1.5 | NE  | 2.5          | 2.0 | 0.5 |
| <i>PRNP</i> <sup>+/Ter</sup>   | 469   | 2.0      | 1.0 | NE  | 1.5      | 0.5 | 0.5 | 2.0    | 1.5 | 0.5 | NE           | 1.5 | NE  |
| <i>PRNP</i> <sup>+/Ter</sup>   | 527   | 3.0      | 2.0 | 0.5 | 2.5      | 2.0 | 0.5 | 3.0    | 2.0 | 0.5 | 3.0          | 2.5 | 0.5 |
| <i>PRNP</i> <sup>+/Ter</sup>   | 533   | 1.5      | 1.0 | 0.0 | 1.0      | 1.0 | 1.0 | 1.5    | 1.5 | 0.0 | 3.0          | 2.5 | 1.0 |
|                                |       | 2.4      | 1.4 | 0.5 | 1.9      | 1.1 | 0.5 | 2.1    | 1.6 | 0.3 | 2.8          | 2.1 | 0.7 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 413   | 0.0      | 0.0 | 0.0 | 0.0      | 0.0 | 0.0 | 0.0    | 0.0 | 0.0 | 0.0          | 0.0 | 0.0 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 457   | 0.0      | 0.0 | 0.0 | 0.0      | 0.0 | 0.0 | 0.0    | 0.0 | 0.0 | 0.0          | 0.0 | 0.0 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 476   | 0.0      | 0.0 | 0.0 | 0.0      | 0.0 | 0.0 | 0.0    | 0.0 | 0.0 | 0.0          | 0.0 | 0.0 |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 490   | 0.0      | 0.0 | 0.0 | 0.0      | 0.0 | 0.0 | 0.0    | 0.0 | 0.0 | 0.0          | 0.0 | 0.0 |
|                                |       | 0.0      | 0.0 | 0.0 | 0.0      | 0.0 | 0.0 | 0.0    | 0.0 | 0.0 | 0.0          | 0.0 | 0.0 |

Abbreviations: DH, dorsal horn; VH, ventral horn; DRG, dorsal root ganglion

### Semi-quantitative scoring of PrP<sup>Sc</sup> distribution in peripheral tissues

| Genotype                       | Goat# | Parotid lymph node | RPLN | Superficial cervical lymph node | Spleen | RAMALT | Adrenal gland | Parotid gland |
|--------------------------------|-------|--------------------|------|---------------------------------|--------|--------|---------------|---------------|
| <i>PRNP</i> <sup>+/+</sup>     | 416   | NE                 | 0.5  | 0.0                             | 0.0    | 0.0    | 0.0           | NE            |
| <i>PRNP</i> <sup>+/+</sup>     | 417   | NE                 | 0.5  | 0.0                             | 0.0    | 0.0    | 0.0           | NE            |
| <i>PRNP</i> <sup>+/+</sup>     | 529   | NE                 | 0.5  | 0.0                             | 0.0    | 0.0    | 0.0           | NE            |
| <i>PRNP</i> <sup>+/+</sup>     | 536   | NE                 | 2.0  | 0.0                             | 0.0    | 0.0    | 0.0           | NE            |
|                                |       |                    | 0.9  | 0.0                             | 0.0    | 0.0    | 0.0           |               |
| <i>PRNP</i> <sup>+/Ter</sup>   | 451   | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>+/Ter</sup>   | 469   | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>+/Ter</sup>   | 527   | 1.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>+/Ter</sup>   | 533   | NE                 | 0.0  | NE                              | 0.0    | 0.0    | 0.0           | 0.0           |
|                                |       | 0.3                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 413   | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 457   | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 476   | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
| <i>PRNP</i> <sup>Ter/Ter</sup> | 490   | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |
|                                |       | 0.0                | 0.0  | 0.0                             | 0.0    | 0.0    | 0.0           | 0.0           |

Abbreviations: RPLN, retropharyngeal lymph node; RAMALT, rectoanal mucosa-associated lymphoid tissue