Authors: Patricia Aguilar-Calvo, Christine Fast, Kerstin Tauscher, Juan-Carlos Espinosa, Martin H. Groschup, Muhammad Nadeem, Wilfred Goldmann, Jan Langeveld, Alex Bossers, Olivier Andreoletti, and Juan-María Torres.

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Background. The prion protein-encoding gene (PRNP) is one of the major determinants for scrapie occurrence in sheep and goats. However, its effect on bovine spongiform encephalopathy (BSE) transmission to goats is not clear. Methods. Goats harboring wild type (WT), R/Q211 or Q/K222 PRNP genotypes were orally inoculated with a goat-BSE isolate to assess their relative susceptibility to BSE infection. Goats were culled at different time points during the incubation period and after the onset of clinical signs, and their brains as well as several peripheral tissues were analyzed for the accumulation of pathological prion protein (PrPSc) and prion infectivity by mouse bioassay.

Results. R/Q211 goats displayed delayed clinical signs compared to WT goats. PrPSc was detected only in brain whereas infectivity was present in peripheral tissues too. In contrast, none of the Q/K222 goats showed any evidence of clinical prion disease. No PrPSc accumulation was observed in their brains or peripheral tissues but very low infectivity was detected in some tissues after very long post-inoculation times (44-45 months).

Conclusions. These results demonstrate that transmission of goat-BSE is genotype dependent and highlight the pivotal protective effect of the K222 PRNP variant in the oral susceptibility of goats to BSE.