Protective Effect of Val₁₂₉-PrP against Bovine Spongiform Encephalopathy but not Variant Creutzfeldt-Jakob Disease

Technical Appendix

Technical Appendix Table 1. Transmission of Ca-BSE derived isolates adapted in different human PrP polymorphic variants to BoPrP-Tg110 mice

	Mean survival time in days±SD (n/n₀)*†	
Isolates	BoPrP-Tg110	
Ca-BSE ₀	303 ± 10 (13/13)(1)	
Ca-BSE ₀ →TgVaI ₁₂₉	>700‡ (0/7)	
Ca-BSE ₂	$308 \pm 5 (5/5) (1)$	
$Ca-BSE_2 \rightarrow TgMet_{129} \rightarrow TgMet_{129}$	$308 \pm 13 (5/5)(2)$	
Ca-BSE/Sh(ARQ)	234 ± 5 (16/16)(1)	
Ca-BSE/Sh(ARQ) →TgVal ₁₂₉	>700‡ (0/6)	
Ca-BSE/Sh(ARQ) →TgVal ₁₂₉ →TgVal ₁₂₉	>700‡ (0/5)	
Ca-BSE/Sh(ARQ) →TgMet ₁₂₉	278 ± 7 (7/7)	
Ca-BSE/Sh(ARQ) →TgMet ₁₂₉ →TgVal ₁₂₉	311 ± 9 (6/6)	
Ca-BSE/Go	239 ± 66 (6/6)(3)	
Ca-BSE/Go→TgVal ₁₂₉	427 ± 38 (3/6)	
Hu-vCJD₁	$370 \pm 33 (9/9)(1)$	
Hu-vCJD₁→TgVal ₁₂₉	>700 ^b (0/5)	
Hu-vCJD ₂	276 ± 9 (6/6)	
Hu-vCJD₂→TgVal ₁₂₉	371 ± 5 (3/3)§	
Hu-vCJD ₂ →TgMet/Val ₁₂₉ →TgVal ₁₂₉	260 ± 41 (6/6)	
*-/- discount D-D(PSiti/indetdeinl-	·	

^{*}n/n₀: diseased PrP^{res} positive/inoculated animals.

Technical Appendix Table 2. Transmission of vCJD and cattle, sheep and goat BSE isolates to TgVal₁₂₉ (8x) mice

	Mean survival time in days ±SD (n/n ₀)* TgVal ₁₂₉ (8x)	
Isolates	1 st passage	2 nd passage
Hu-sCJD MM1	295 ± 21 (6/6)	215 ± 4 (7/7)
Hu-sCJD VV2	$228 \pm 7 (5/5)$	228 ± 14 (5/5)
Hu-TSE negative	>700† (0/6)	>700† (0/6)
Ca-BSE₀	>700† (0/6)	>700† (0/5)
Ca-BSE ₂	>700† (0/3)‡	>700† (0/3)‡
Ca-BSE/Sh(ARQ)	>700† (0/5)	>700† (0/6)
Ca-BSE/Go	>700† (0/5)	
Hu-vCJD ₂	>700† (0/5)	>700† (0/6)

^{*}Survival time is indicated as mean±SD dpi of all the mice scored positive for PrPres. n/n₀: diseased PrPres positive/inoculated animals.

References

1. Padilla D, Béringue V, Espinosa JC, Andreoletti O, Jaumain E, Reine F, et al. Sheep and goat BSE propagate more efficiently than cattle BSE in human PrP transgenic mice. PLoS Pathog. 2011;7:e1001319. PubMed http://dx.doi.org/10.1371/journal.ppat.1001319

[†]Survival time is indicated as mean±SD dpi of all the mice scored positive for PrPres.

[‡]Animals were euthanized without clinical signs at 700 dpi.

^{\$}Three additional animals had to be culled before the end of the experiment because of intercurrent illnesses; all were negative for brain PrP^{res} on WB.

[†]Animals were euthanized without clinical signs at 700 dpi.

[‡]Three additional animals had to be culled before the end of the experiment because of intercurrent diseases; all were negative for brain PrPres on WB.

- Torres JM, Espinosa JC, Aguilar-Calvo P, Herva ME, Relaño-Ginés A, Villa-Diaz A, et al. Elements modulating the prion species barrier and its passage consequences. PLoS One. 2014;9:e89722. PubMed http://dx.doi.org/10.1371/journal.pone.0089722
- Aguilar-Calvo P, Fast C, Tauscher K, Espinosa JC, Groschup MH, Nadeem M, et al. Effect of Q211 and K222 PRNP polymorphic variants in the susceptibility of goats to oral infection with goat bovine spongiform encephalopathy. J Infect Dis. 2015;212:664–72. PubMed http://dx.doi.org/10.1093/infdis/jiv112