Efficient transmission of human prion diseases to a glycan-free prion protein-expressing host

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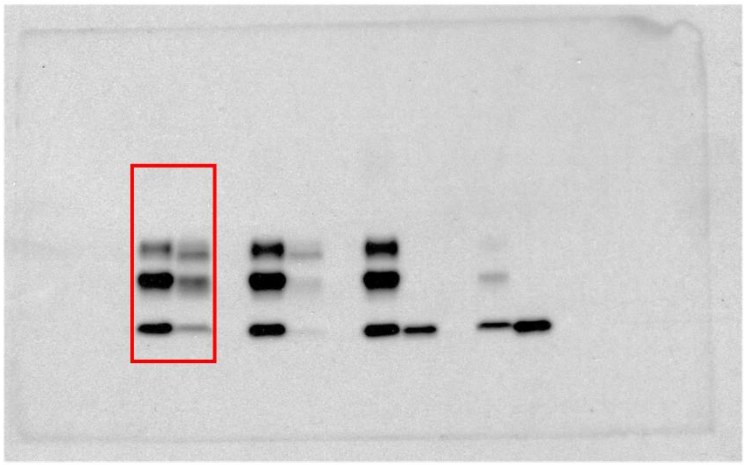
Supplementary Table 1: Summary of the main findings

Parameters	Sporadic CJD MM2	Sporadic Fatal Insomnia	Familial Fatal Insomnia
Histopathology in human diseases	SD: Large, confluent vacuoles w. PrP deposits, preferentially in the neocortex	SD: Similar to sCJDMM2 but less severe, its presence depends on disease duration (>24 months). Focal thalamic atrophy	SD: Similar to sCJDMM2 but much less severe and strictly dependent on disease duration. Focal thalamic atrophy
Inoculated mice			
Glyc- mice average incubation periods (in dpi)	1st p.: 266 (~2x shorter than Glyc+ controls). 2nd & 3rd p.: 78	1st p.: 346 (~1.8x shorter than Glyc+ controls. 2nd & 3rd p.: 114	1st p.: 289 (no transmission in controls). 2nd p.: 193
Histopathology in Gyc- mice and in Glyc+ controls	SD & PrP deposition similar to hu. disease; ~4x more severe than controls, w. large PrP deposit & high astro-microglia reactions	Same as after sCJDMM2 transmission and more severe than in controls	Same as in sFI-Glyc- mice but slightly less severe
Histopathology severity & incubation period	Severity, accurate SD replication and presence of the large PrP deposits directly correlated w. the length of the incubation period		
resPrP^D WB in Glyc- mice	Same electrophoretic mobility as unglycosylated in Glyc+ and 3 hu. diseases		
resPrP^D quantitative assessment in Glyc- mice	~40x > Glyc+ control	Definitely > controls	High; no controls avail.
Tot & resPrP^D SE profiles	Glyc- mice: prevalence of high-density aggregates Glyc+ mice: prevalence of low-density aggregates		Not tested
Glyc- mice: CSSA	Significant increase in resPrP^D stability	Not tested	Not tested
Glyc- mice: Anchorless resPrP^D as % of total	21.1: Similar to that of the sCJDMM2 (17.4)	Not tested	Not tested

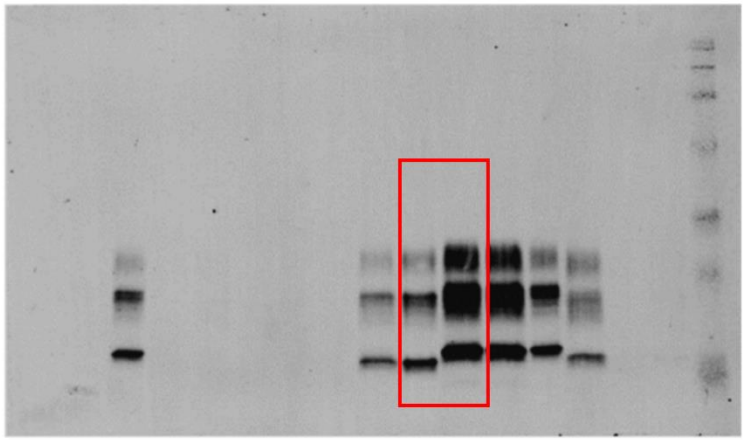
Abbreviations: avail.: available; CSSA: conformational solubility and stability assay; dpi: days post-inoculations; Glyc-: TgGlyc mice; hu.: human; mo.: months; p.: passage; SD: spongiform degeneration; w.: with; WB: western blot; SE: sedimentation equilibrium.

Supplementary Figure 1 Original uncropped immunoblots shown in Figures 5 A, and 5 B. Red boxes indicate the cropped region shown in the corresponding panel.

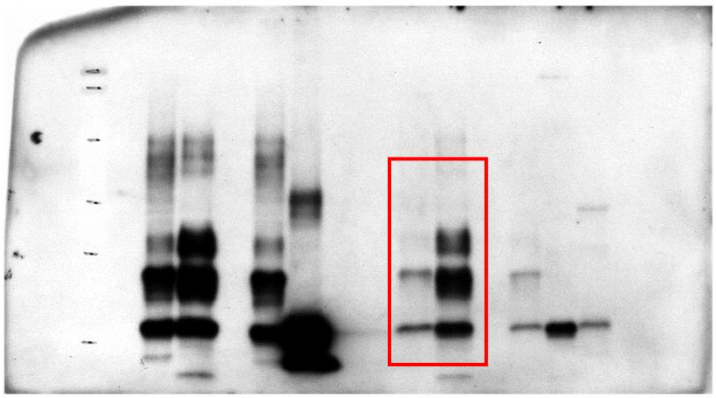
Fig. 5 A



Panel 1

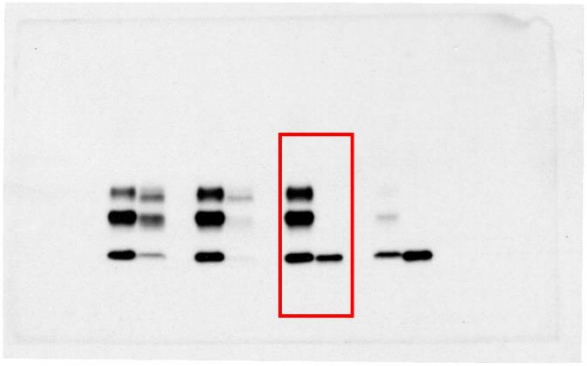


Panel 2

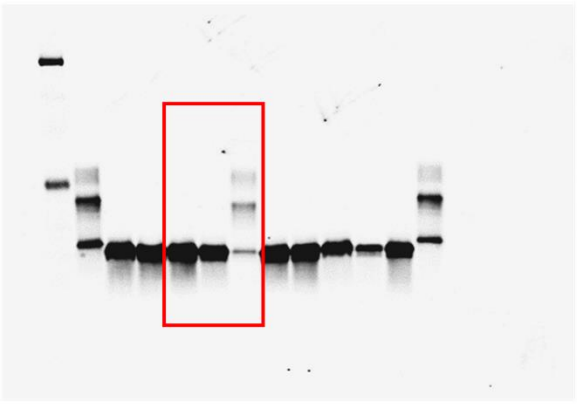


Panel 3

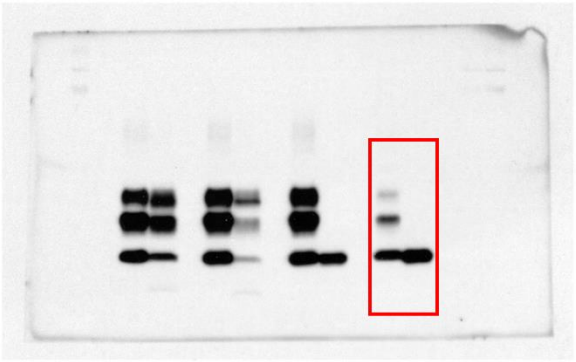
Fig. 5 B



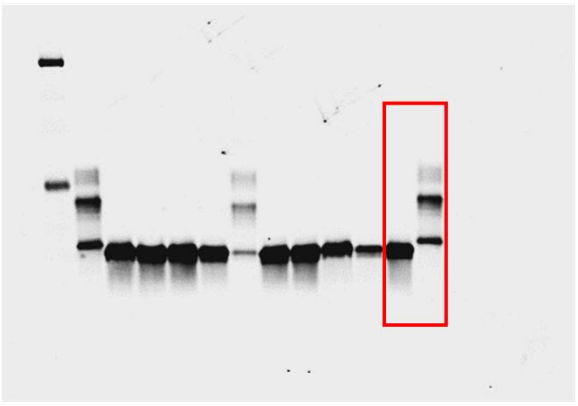
Panel 1



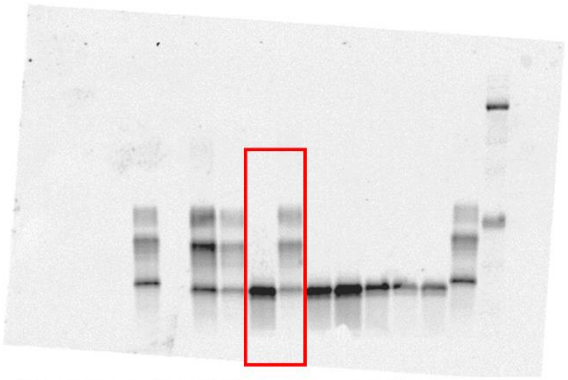
Panel 2



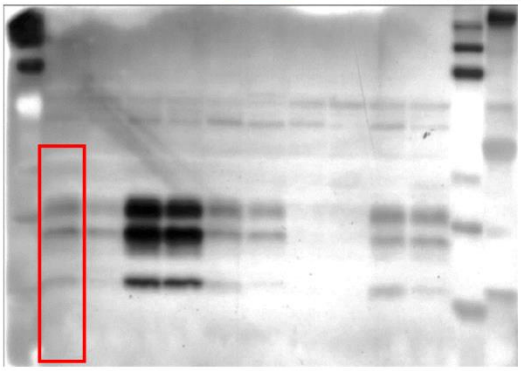
Panel 3



Panel 4



Panel 5



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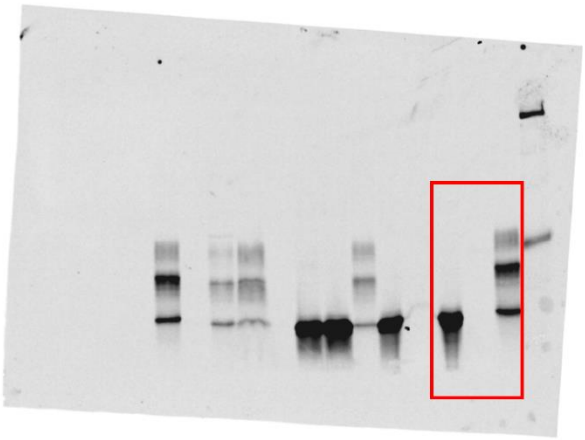
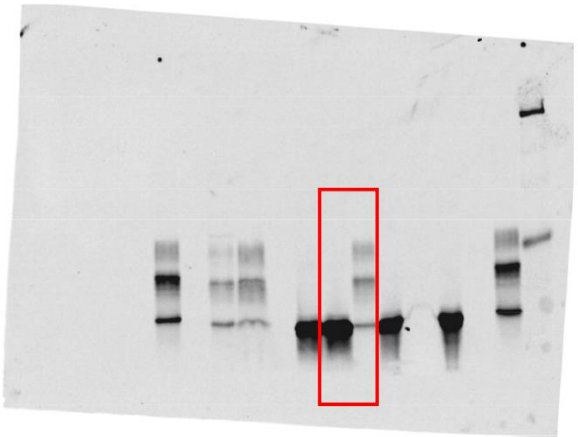


Panel 7



Panel 8

Supplementary Figure 2 Original uncropped immunoblots shown in Figures 7 B(i), 7 B(ii), 7 C and 7 D. Red boxes indicate the cropped region shown in the corresponding panel.

Fig. 7 B(i)

Fig. 7 B(ii)

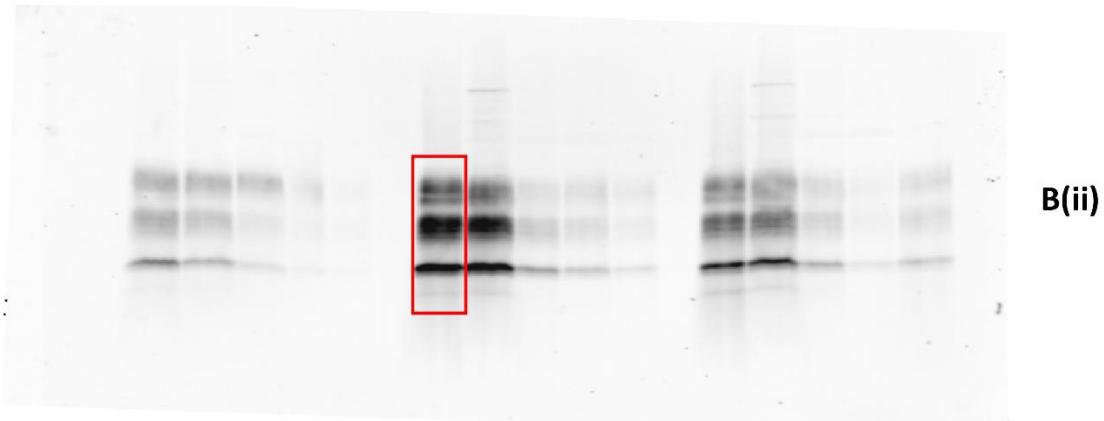


Fig. 7 C

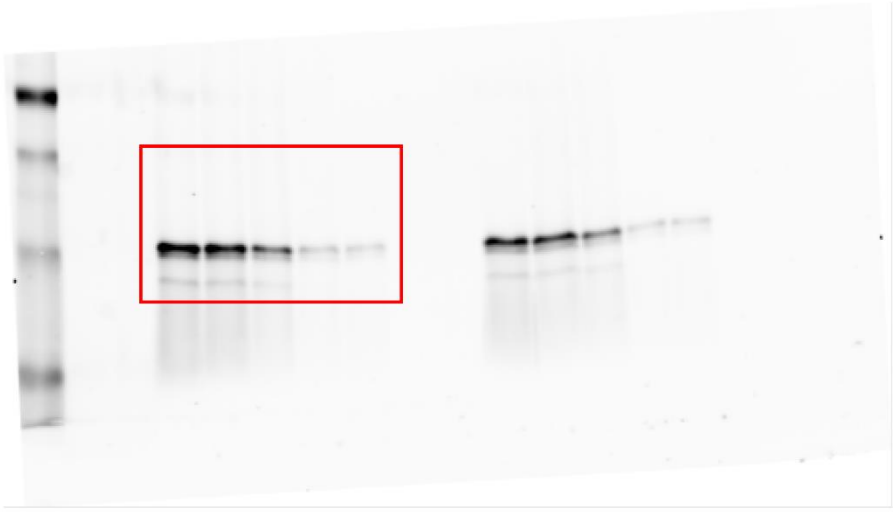


Fig. 7 D