Suppl. Fig 1 Western blot analysis of VPSPr and sCJD VV2 using anti-PrP primary antibodies recognising internal and C-terminal epitopes.

Western blot analysis of cerebral cortex homogenate from an sCJD VV2 case or VPSPr case 1, with (+) or without (-) PK treatment, using the anti-PrP monoclonal antibodies 3F4 (amino acids 106-112) or 94B4 (C-terminal, amino acids 187-193). The volumes of homogenates analysed are indicated. Numbers in brackets [] indicate the volumes that were centrifugally concentrated.
Suppl. Fig. 2 D-N and N values for whole brain homogenate versus centrifugal pellet for VPSPr versus non-CJD

D-N (red bar) values for brain frontal cortex homogenate and centrifugal pellet from a case of VPSPr and a non-CJD case (ALS/FTLD). Samples (100μl) of a 10% (w/v) homogenate of frontal cortex (H) from VPSPr case 1 or a non-CJD case were analysed directly by CDI. Alternatively, 100μl samples were centrifuged at 20,000g for 1h at 4°C and the pellets were analysed by CDI. The results are the means + S.D. for triplicate analyses of single samples.
Suppl. Fig. 3

**FC**  
GdnHCl½ = 2.21, CI=0.11

**TC**  
GdnHCl½ = 2.01, CI=0.42

**PC**  
GdnHCl½ = 2.07, CI=0.39

**OC**  
GdnHCl½ = 1.63, CI=0.21

**Me**  
GdnHCl½ = 2.87, CI=0.35

**PM**  
GdnHCl½ = 2.92, CI=0.23

**Midbrain**  
GdnHCl½ = 1.80, CI=0.61

**CB**  
GdnHCl½ = 3.52 CI=0.11

Continued………
BG

GdnHCl½ = 2.08, CI=0.21

Hip

GdnHCl½ = 2.2, CI=0.26

Thal

GdnHCl½ = 2.32, CI=0.31

GSS FC

GdnHCl½ = 2.57, CI=0.06

GSS CB

GdnHCl½ = 2.64, CI=0.06

GSS small fragment FC

GdnHCl½ = 1.68, CI=0.36

sCJD VV2 FC

GdnHCl½ = 2.44, CI=0.22

Continued......
Suppl. Fig. 3 Equilibrium unfolding analysis
The stability of PrP^Sc in various regions of a VPSPr brain, two cases of GSS and a case of sCJD VV2 was investigated by CDI following denaturation with various concentrations of GdnHCl. The y-axis on this curve is the fraction of PrP^Sc that is unfolded, calculated as described in the methods. The abbreviations used for the various brain regions are the same as in Fig. 5. For the analysis of FC, CB and PC, the data were merged from the analysis of 4, 3 and 2 samples, respectively. Single samples were analysed for the other regions. FC was analysed from one GSS case (P102L, small fragment), and CB and FC was analysed from another GSS case (P102L, type 1). Underneath each figure is given the [GdnHCl]1/2 value and the 95% CI for the curve fit.