

1 **Detection of prion seeding activity in the olfactory mucosa of patients with Fatal**  
2 **Familial Insomnia**

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34 **SUPPLEMENTARY MATERIALS**

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50 **Supplementary Figure 1. PMCA analysis of OM samples from patients with other**

51 **neurodegenerative disorders.** PMCA analysis of OM samples collected from 2 FFI subjects, 3

52 patients with other neurodegenerative diseases, including AD (n=1), PD (n=1) and FTD (n=1) and 2

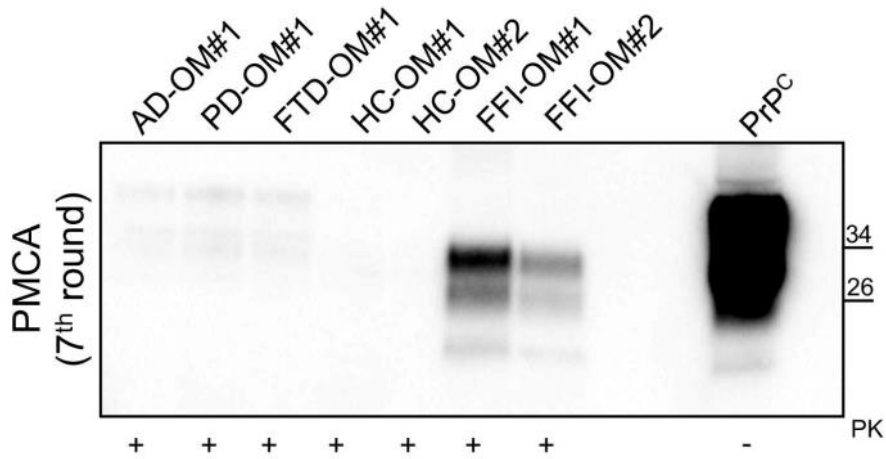
53 healthy controls. Seven rounds of amplification were performed. The signal of PrP<sup>Sc</sup> was assessed by

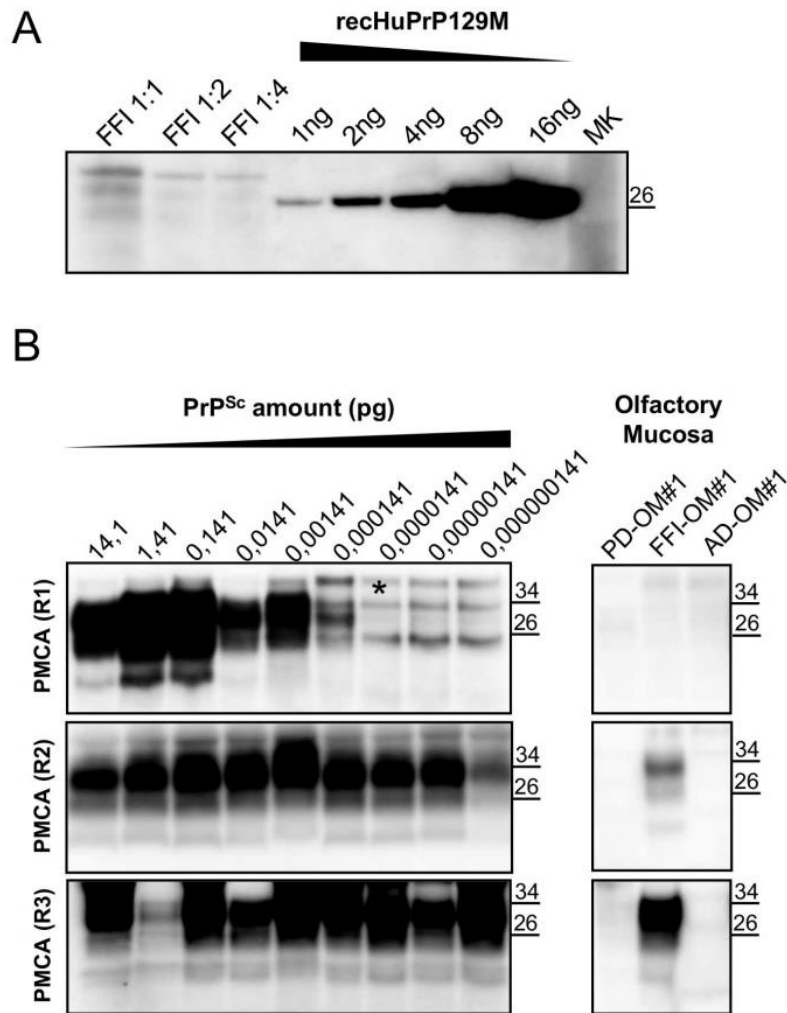
54 means of Western blotting, after PK digestion, with the 6D11 antibody. Numbers in the right of the

55 blots indicate the position of molecular weight marker.

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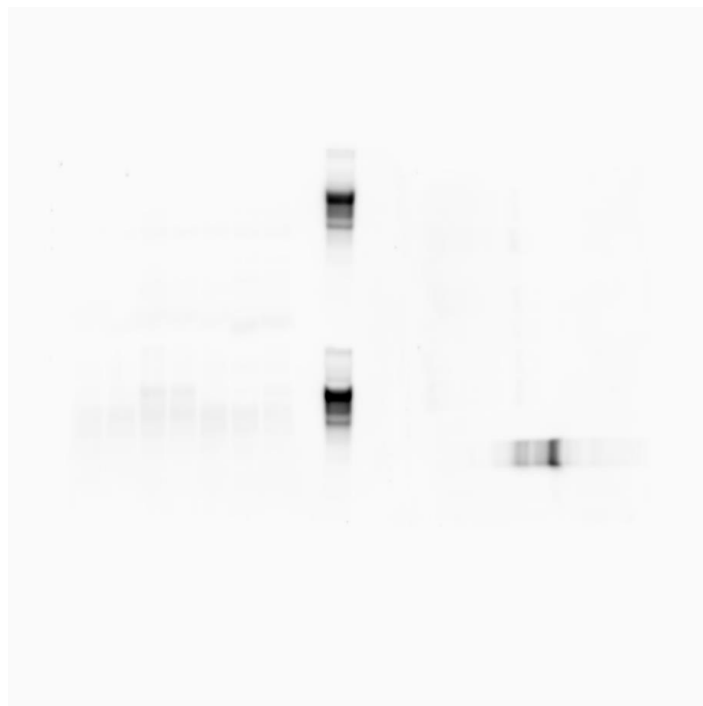
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59 **Supplementary Figure 2. Quantitative PMCA (qPMCA) for estimating PrP<sup>Sc</sup> concentration in**  
 60 **OM samples of FFI patients. (A)** The intensity of the PrP<sup>Sc</sup> signal obtained after PK digestion from  
 61 different dilutions of FFI brain homogenate (20-fold concentrated) was compared to that of known  
 62 concentrations of recombinant human prion protein with methionine at codon 129 (recHuPrP129M).  
 63 Mw refers to molecular weight marker. **(B)** Quantified FFI-PrP<sup>Sc</sup> was spiked in bank vole brain  
 64 homogenate and PMCA experiments were performed along with FFI-OM#1 sample. OM collected  
 65 from patients with PD and AD were used as negative controls. The signal of PrP<sup>Sc</sup> was assessed by  
 66 means of Western blotting, after PK digestion, with the 6D11 antibody. Asterisk indicates unspecific  
 67 signal. Numbers in the right of the blots indicate the position of molecular weight marker.

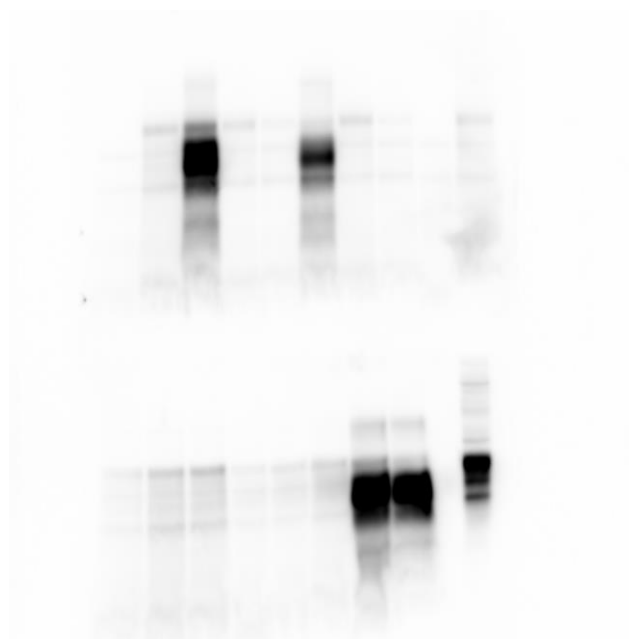
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**A (Figure 2A. Before PMCA)**



**B (Figure 2A. After PMCA 3<sup>rd</sup> round)**



94 **Raw pictures.** (A) Raw gels used to prepare Figure 2A “Before PMCA”. The picture on top/left and  
95 the picture on the right were used. (B) Raw gels used to prepare Figure 2A “After PMCA 3<sup>rd</sup> round”.  
96 Both pictures were used.