

Addition/Correction

The Emerging Principles of Mammalian Prion Propagation and Transmissibility Barriers: Insight from Studies in Vitro

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ADDITIONS AND CORRECTIONS

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Witold K. Surewicz, Eric M. Jones, and Adrian C. Apetri : The Emerging Principles of Mammalian Prion Propagation and Transmissibility Barriers: Insight from Studies in Vitro

Pages 656 and 661. The references were misnumbered in the version of this paper published on the Web on May 19, 2006, and in the September 19, 2006 print issue. As a result of this misnumbering, two citations on page 656 had to be replaced in addition to the references being renumbered. In the second full paragraph on page 656, the original text read as follows: "...suggesting a role of partially folded intermediates in the conformational transition.²⁷ Importantly, the α -helix \rightarrow β -sheet transition occurs concomitantly with oligomerization of the protein,^{26,27}". The corrected text reads as follows: "...suggesting a role of partially folded intermediates in the conformational transition.²⁶ Importantly, the α -helix \rightarrow β -sheet transition occurs concomitantly with oligomerization of the protein,^{25,26}".

The electronic version of this paper, with the references correctly numbered and the citations updated, was replaced on the Web on September 27, 2006.

The complete list of references, correctly numbered, is presented below.

References

- (1) Prusiner, S. B. Prions. *Proc. Natl. Acad. Sci. U.S.A.* **1998**, *95*, 13363–13383.
- (2) Collinge, J. Prion diseases of humans and animals: their causes and molecular basis. *Annu. Rev. Neurosci.* **2001**, *24*, 519–550.
- (3) Aguzzi, A.; Polymenidou, M. Mammalian prion biology: one century of evolving concepts. *Cell* **2004**, *116*, 313–327.
- (4) Weissmann, C. The state of the prion. *Nat. Rev. Microbiol.* **2004**, *2*, 861–871.
- (5) Prusiner, S. B. Molecular biology of prion diseases. *Science* **1991**, *252*, 1515–1522.
- (6) Chien, P.; Weissman, J. S.; DePace, A. H. Emerging principles of conformation-based prion inheritance. *Annu. Rev. Biochem.* **2004**, *73*, 617–656.
- (7) Tanaka, M.; Chien, P.; Naber, N.; Cooke, R.; Weissman, J. S. Conformational variations in an infectious protein determine prion strain differences. *Nature* **2004**, *428*, 323–328.
- (8) Brachmann, A.; Baxa, U.; Wickner, R. B. Prion generation in vitro: amyloid of Ure2p is infectious. *EMBO J.* **2005**, *24*, 3082–3092.
- (9) Millhauser, G. L. Copper binding in the prion protein. *Acc. Chem. Res.* **2004**, *37*, 79–85.
- (10) Riek, R.; Hornemann, S.; Wider, G.; Billeter, M.; Glockshuber, R.; Wuthrich, K. NMR structure of the mouse prion protein domain PrP(121–231). *Nature* **1996**, *382*, 180–182.
- (11) Donne, D. G.; Viles, J. H.; Groth, D.; Mehlhorn, I.; James, T. L.; Cohen, F. E.; Prusiner, S. B.; Wright, P. E.; Dyson, H. J. Structure of the recombinant full-length hamster prion protein PrP(29–231): the N-terminus is highly flexible. *Proc. Natl. Acad. Sci. U.S.A.* **1997**, *94*, 13452–13457.
- (12) Zahn, R.; Liu, A.; Luhrs, T.; Riek, R.; von Schroetter, C.; Lopez Garcia, F.; Billeter, M.; Calzolai, L.; Wider, G.; Wuthrich, K. NMR solution structure of the human prion protein. *Proc. Natl. Acad. Sci. U.S.A.* **2000**, *97*, 145–150.
- (13) Knaus, K. J.; Morillas, M.; Swietnicki, W.; Malone, M.; Surewicz, W. K.; Yee, V. C. Crystal structure of the human prion protein reveals a mechanism for oligomerization. *Nat. Struct. Biol.* **2001**, *8*, 770–774.
- (14) Kelly, J. W. The alternative conformations of amyloidogenic proteins and their multi-step assembly pathways. *Curr. Opin. Struct. Biol.* **1998**, *8*, 101–106.
- (15) Jarrett, J. T.; Lansbury, P. T., Jr. Seeding "one-dimensional crystallization" of amyloid: a pathogenic mechanism in Alzheimer's disease and scrapie? *Cell* **1993**, *73*, 1055–1058.
- (16) Silveira, J. R.; Raymond, G. J.; Hughson, A. G.; Race, R. E.; Sim, V. L.; Hayes, S. F.; Caughey, B. The most infectious prion protein particles. *Nature* **2005**, *437*, 257–261.
- (17) Kocisko, D. A.; Come, J. H.; Priola, S. A.; Chesebro, B.; Raymond, G. J.; Lansbury, P. T.; Caughey, B. Cell-free formation of protease-resistant prion protein. *Nature* **1994**, *370*, 471–474.
- (18) Caughey, B. Interactions between prion protein isoforms: the kiss of death? *Trends Biochem. Sci.* **2001**, *26*, 235–242.
- (19) Supattapone, S. Prion protein conversion in vitro. *J. Mol. Med.* **2004**, *82*, 348–356.
- (20) Saborio, G. P.; Permanne, B.; Soto, C. Sensitive detection of pathological prion protein by cyclic amplification of protein misfolding. *Nature* **2001**, *411*, 810–813.
- (21) Castilla, J.; Saa, P.; Hetz, C.; Soto, C. In vitro generation of infectious scrapie prions. *Cell* **2005**, *121*, 195–206.
- (22) Swietnicki, W.; Petersen, R.; Gambetti, P.; Surewicz, W. K. pH-dependent stability and conformation of the recombinant human prion protein PrP(90–231). *J. Biol. Chem.* **1997**, *272*, 27517–27520.
- (23) Hornemann, S.; Glockshuber, R. A scrapie-like unfolding intermediate of the prion protein domain PrP(121–231) induced by acidic pH. *Proc. Natl. Acad. Sci. U.S.A.* **1998**, *95*, 6010–6014.
- (24) Jackson, G. S.; Hosszu, L. L.; Power, A.; Hill, A. F.; Kenney, J.; Saibil, H.; Craven, C. J.; Walther, J. P.; Clarke, A. R.; Collinge, J. Reversible conversion of monomeric human prion protein between native and fibrillogenic conformations. *Science* **1999**, *283*, 1935–1937.
- (25) Swietnicki, W.; Morillas, M.; Chen, S. G.; Gambetti, P.; Surewicz, W. K. Aggregation and fibrillization of the recombinant human prion protein huPrP90–231. *Biochemistry* **2000**, *39*, 424–431.
- (26) Morillas, M.; Vanik, D. L.; Surewicz, W. K. On the mechanism of α -helix \rightarrow β -sheet transition in the recombinant prion protein. *Biochemistry* **2001**, *40*, 6982–6987.
- (27) Maiti, N. R.; Surewicz, W. K. The role of disulfide bridge in the folding and stability of the recombinant human prion protein. *J. Biol. Chem.* **2001**, *276*, 2427–2431.
- (28) Lee, S.; Eisenberg, D. Seeded conversion of recombinant prion protein to a disulfide-bonded oligomer by a reduction–oxidation process. *Nat. Struct. Biol.* **2003**, *10*, 725–730.
- (29) Baskakov, I. V.; Legname, G.; Baldwin, M. A.; Prusiner, S. B.; Cohen, F. E. Pathway complexity of prion protein assembly into amyloid. *J. Biol. Chem.* **2002**, *277*, 21140–21148.
- (30) Baskakov, I. V. Autocatalytic conversion of recombinant prion proteins displays a species barrier. *J. Biol. Chem.* **2004**, *279*, 7671–7677.
- (31) Apetri, A. C.; Vanik, D. L.; Surewicz, W. K. Polymorphism at residue 129 modulates the conformational conversion of the D178N variant of human prion protein 90–231. *Biochemistry* **2005**, *44*, 15880–15888.
- (32) Legname, G.; Baskakov, I. V.; Nguyen, H. O.; Riesner, D.; Cohen, F. E.; DeArmond, S. J.; Prusiner, S. B. Synthetic mammalian prions. *Science* **2004**, *305*, 673–676.
- (33) Weissmann, C. Birth of a prion: spontaneous generation revisited. *Cell* **2005**, *122*, 165–168.
- (34) Wildegg, G.; Liemann, S.; Glockshuber, R. Extremely rapid folding of the C-terminal domain of the prion protein without kinetic intermediates. *Nat. Struct. Biol.* **1999**, *6*, 550–553.
- (35) Hosszu, L. L.; Baxter, N. J.; Jackson, G. S.; Power, A.; Clarke, A. R.; Walther, J. P.; Craven, C. J.; Collinge, J. Structural mobility of the human prion protein probed by backbone hydrogen exchange. *Nat. Struct. Biol.* **1999**, *6*, 740–743.

- (36) Apetri, A. C.; Surewicz, W. K. Kinetic intermediate in the folding of human prion protein. *J. Biol. Chem.* **2002**, *277*, 44589–44592.
- (37) Nicholson, E. M.; Mo, H.; Prusiner, S. B.; Cohen, F. E.; Marqusee, S. Differences between the prion protein and its homolog Doppel: a partially structured state with implications for scrapie formation. *J. Mol. Biol.* **2002**, *316*, 807–815.
- (38) Kuwata, K.; Li, H.; Yamada, H.; Legname, G.; Prusiner, S. B.; Akasaka, K.; James, T. L. Locally disordered conformer of the hamster prion protein: a crucial intermediate to PrP^{Sc}? *Biochemistry* **2002**, *41*, 12277–12283.
- (39) Apetri, A. C.; Surewicz, K.; Surewicz, W. K. The effect of disease-associated mutations on the folding pathway of human prion protein. *J. Biol. Chem.* **2004**, *279*, 18008–18014.
- (40) Kundu, B.; Maiti, N. R.; Jones, E. M.; Surewicz, K. A.; Vanik, D. L.; Surewicz, W. K. Nucleation-dependent conformational conversion of the Y145Stop variant of human prion protein: structural clues for prion propagation. *Proc. Natl. Acad. Sci. U.S.A.* **2003**, *100*, 12069–12074.
- (41) Scott, M.; Peretz, D.; Ridley, R. M.; Baker, H. F.; DeArmond, S. J.; Prusiner, S. B. Transgenic investigations of the species barrier and prion strains. In *Prion Biology and Diseases*, 2nd ed.; Prusiner, S. B., Ed.; Cold Spring Harbor Monograph Series 41; Cold Spring Harbor Press: Cold Spring Harbor, NY, 2004; Chapter 9.
- (42) Priola, S. A.; Vorberg, I. Molecular aspects of disease pathogenesis in the transmissible spongiform encephalopathies. *Methods Mol. Biol.* **2004**, *268*, 517–540.
- (43) Vanik, D. L.; Surewicz, K. A.; Surewicz, W. K. Molecular basis of barriers for interspecies transmissibility of mammalian prions. *Mol. Cell* **2004**, *14*, 139–145.
- (44) Jones, E. M.; Surewicz, W. K. Fibril conformation as the basis of species- and strain-dependent seeding specificity of mammalian prion amyloids. *Cell* **2005**, *121*, 63–72.
- (45) Bessen, R. A.; Marsh, R. F. Distinct prion properties suggest the molecular basis of strain variation in transmissible mink encephalopathy. *J. Virol.* **1994**, *68*, 7859–7868.
- (46) Jones, E. M.; Surewicz, K. A.; Surewicz, W. K. Role of N-terminal familial mutations in prion protein fibrillization and prion amyloid propagation in vitro. *J. Biol. Chem.* **2006**, *281*, 8190–8196.
- (47) Tanaka, M.; Chien, P.; Yonekura, K.; Weissman, J. S. Mechanism of cross-species prion transmission: an infectious conformation compatible with two highly divergent yeast prion proteins. *Cell* **2005**, *121*, 49–62.
- (48) Petkova, A. T.; Leapman, R. D.; Guo, Z.; Yau, W. M.; Mattson, M. P.; Tycko, R. Self-propagating, molecular-level polymorphism in Alzheimer's beta-amyloid fibrils. *Science* **2005**, *307*, 262–265.

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