

Prione

Is it necessary that we must focus our attention again? [BDH](#)

„Prione machen nicht immer krank. Prione können auch z.B. spezielle Fähigkeiten und Eigenschaften unter gesunden Wirten weiter vererben.

Prions may not be universally pathogenic and may even act as protein-based elements of inheritance in healthy organisms”. Halfmann R

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https://www.rki.de/DE/Content/Infekt/Krankenhaushygiene/Erreger_ausgewaehlt/CJK/CJK_pdf_04.pdf?__blob=publicationFile

„Zur Temperaturempfindlichkeit von Prionen ist anzumerken, dass bei Dampfsterilisation selbst unter Anwendung von 18 Minuten Sterilisierzeit bei 134–137°C ein gewisser Anteil der Prionen infektiös bleiben kann [1, 13] und trockene Hitze sogar bei extremen Temperaturen eine Restinfektiosität nicht beseitigt hat [14]“

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"First, we successfully propagated various Creutzfeldt-Jakob disease isolates (sporadic, variant and iatrogenic CJD) in neuronal cultures expressing the human prion protein. Then, we found that doxycycline was the most effective compound, with important variations between isolates. Isolates from sporadic CJD, the most common form of prion diseases showed the highest sensitivity."

Van Ba IA-T, Imberdis,T, Perrier V (2013) Review Article. From Prion Diseases to Prion-Like Propagation Mechanisms of Neurodegenerative Diseases. *International Journal of Cell Biology* Volume 2013 Article ID 975832, 8 pages <http://dx.doi.org/10.1155/2013/975832>
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"Amyloid" is a generic term and all amyloids, irrespective of amino acid sequence, are formed in a seeded nucleation mechanism in which a small aggregate (oligomers) of a few amyloid moieties (a seed or a nucleus) seed (nucleate) normal amyloid precursor moieties to change conformation to a β -sheet. ...

There are several protein misfolding disorders - the most widely known include Alzheimer's disease, Parkinson's disease and other α -synucleinopathies, amyotrophic lateral sclerosis (ALS), frontotemporal dementias in which abnormally phosphorylated MAP- τ protein accumulates and finally, polyglutamine expansion diseases such as Huntington's disease and certain spinocerebellar ataxias. The proteins involved differ in each of these disorders but the molecular mechanism is almost exactly the same, a seeding-nucleation mechanism."

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« Research efforts should focus on this bacterium for development of therapeutic regimens for Creutzfeldt-Jakob disease. »

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Science Translational Medicine, 8(370), 370ra182, DOI: 10.1126/scitranslmed.aag1257

<http://stm.sciencemag.org/content/8/370/370ra182.full>

Varges D et al. (2016) **Doxycycline in early CJD: a double-blinded randomised phase II and observational study.** J Neurol Neurosurg Psychiatry 0, 1–7. doi:10.1136/jnnp-2016-31354 <https://pdfs.semanticscholar.org/61e1/ec1edfbf0b18fd85c0da417591f9b8eb83e4.pdf>

« On the basis of our results, we recommend doxycycline treatment, especially in MM patients with sCJD in earlier disease stages, as a useful therapeutic option, unless other treatment options become available ».

Tzehow M et al. (2017) **Variant Creutzfeldt-Jakob disease in a patient with heterozygosity at PRNP codon 129. Correspondence.** N Engl J Med 376 (3) 292–4. doi:

10.1056/NEJMc1610003 [CrossRef](#). <https://www.ncbi.nlm.nih.gov/labs/articles/28099827/>
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<https://www.aerzteblatt.de/archiv/186263/Creutzfeldt-Jakob-Krankheit-Der-juengste-Fall-passt-nicht-in-das-bisherige-Muster#literatur>

„Ungewöhnlich bei diesem Patienten ist ein Detail im Codon 129 seines Prion-Protein-Gens: Normalerweise kann dieses Kodon für die Aminosäuren Methionin (M) oder Valin (V) kodieren. Menschen können somit hinsichtlich dieses Merkmals homozygot (MM, VV) oder heterozygot (MV) sein. Eine vCJK war aber bislang nur bei Patienten diagnostiziert worden, die den homozygoten MM-Typ aufweisen. In Großbritannien ist nun 1 Patient verstorben, der erstmals die heterozygote Form des Prion-Protein-Gens (MV-Typ) aufweist (2). Man geht davon aus, dass in der Normalbevölkerung 37 % homozygot für die Methionin sind (MM), 12 % homozygot für Valin (VV) und 51 % heterozygot für Methionin und Valin (MV)“.

➔ **Video:** Scientists Observe Infectious Prion Proteins Invade and Move Within Brain Cells. http://www.niaid.nih.gov/topics/prion/Pages/prion_video.aspx
<https://www.youtube.com/watch?v=dkmemumGEMs>

[Bernt - Dieter Huismans](#), last revision april 2017 www.Huismans.click



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