

# Prione, Prions

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**

<http://www.the-scientist.com/?articles.view/articleNo/38721/title/The-Bright-Side-of-Prions/>

**Prions** [http://www.rr-africa.oie.int/docspdf/en/2019/REMESA18/session%20202/9%20Remesa\\_26\\_27\\_June\\_2019.pdf](http://www.rr-africa.oie.int/docspdf/en/2019/REMESA18/session%20202/9%20Remesa_26_27_June_2019.pdf)

Monod J (1970) **Le hazard et la Nécessité**. Edition du Seuil, Paris. Zufall und Notwendigkeit. Piper & Co München 1971

Eigen M (1971) **Selforganization of matter and the evolution of biological macromolecules**. Naturwissenschaften 58, 465-523.

Dickinson AG (1976) Scrapie in sheep and goats. In: Kimberlin RH, editor. **Slow virus** diseases of animals and man. Amsterdam: Elsevier/North Holland. pp. 209-241.

Kimberlin RH, Walker C (1977) Characteristics of a short incubation model of scrapie in the golden hamster. J Gen Virol 34, 295-304.

Prusiner SB (1982) **Novel proteinaceous infectious particles cause scrapie**, [Science](#), 216, 136-44

Kimberlin RH, Walker CA, Fraser H (1989) The genomic identity of different strains of mouse scrapie is expressed in hamsters and preserved on reisolation in mice. J Gen Virol 70, 2017-2025.

Prusiner SB (1991) **Molecular biology of prion diseases**. Science 252(5012), 1515-1522 DOI: 10.1126/science.1675487 <http://www.sciencemag.org/content/252/5012/1515.abstract>  
[http://www.ncbi.nlm.nih.gov/sites/entrez?cmd=search&db=pubmed&term=Prusiner%20S\[au\]&dispmax=50](http://www.ncbi.nlm.nih.gov/sites/entrez?cmd=search&db=pubmed&term=Prusiner%20S[au]&dispmax=50)

Bäßeler H, Aguzzi A, Sailer A, Greiner RA, Autenried P, et al. (1993) **Mice devoid of PrP** are resistant to scrapie. Cell 73, 1339-1347.

[Prusiner](#) SB (1994) **Neurodegeneration in humans caused by prions**. [West J Med](#). 161(3), 264–272. PMID: PMC1011408 <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1011408/>

Wickner RB (1994) [URE3] as an altered URE2 protein: evidence for a **prion analog** in *Saccharomyces cerevisiae*. [Science](#), 264, 566-69

Lansbury PT, Caughey B (1995) The chemistry of scrapie infection: implications of the ‘ice 9’ metaphor. Chem Biol, 2, 1-5.

Kauffman S. (1996) **Self-replication. Even peptides do it**. Nature. 382(6591), 496-497

Lee DH et al. (1996) **A SELF-REPLICATING PEPTIDE**. Nature, 382(6591), 1996, pp. 525-528  
<http://www.ncbi.nlm.nih.gov/pubmed/8700218>

Taylor DM, McConnell I, Fernie K (1996) The effect of dry heat on the ME7 strain of mouse-passaged scrapie agent. J Gen Virol 77, 3161–3164

Appel TR et al.(2001) Heat stability of prion rods and recombinant prion protein in water, lipid and lipid-water mixtures. *J Gen Virol* 82, 465–473 14.

Collinge J (2001) Prion diseases of humans and animals: their causes and molecular basis. *Annu Rev Neurosci* 24, 519–20. [CrossRef](#) [MEDLINE](#)

Kyle RA (2001) **Amyloidosis**: a convoluted story. *Br J Haematol*, 114, 529-38

Abschlussbericht der Task Force vCJK (2002) **Die Variante der Creutzfeldt-Jakob-Krankheit (vCJK)**. Bundesgesundheitsbl Gesundheitsforsch Gesundheitsschutz 45, 376–394

(2002) **Update of the opinion of the SCIENTIFIC STEERING COMMITTEE on the Geographical Risk of Bovine Spongiform Encephalopathy (GBR)**  
[http://ec.europa.eu/food/fs/sc/ssc/out243\\_en.pdf](http://ec.europa.eu/food/fs/sc/ssc/out243_en.pdf)

Klohn PC, Stoltze L, Flechsig E, Enari M, Weissmann C (2003) A quantitative, highly sensitive cell-based infectivity assay for mouse scrapie prions. *Proc Natl Acad Sci U S A* 100, 11666-11671.

Si K et al. (2003) A neuronal isoform of the *Aplysia* CPEB has **prion-like** properties. *Cell*, 115, 879-91.

**Entfernung von Prionen bei der Aufbereitung von Medizinprodukten Ein Beitrag zur Prüfung und Deklaration geeigneter Verfahren.** Bundesgesundheitsbl - Gesundheitsforsch Gesundheitsschutz 2004 · 47:36–40 DOI 10.1007/s00103-003-0761-8  
[https://www.rki.de/DE/Content/Infekt/Krankenhaushygiene/Erreger\\_ausgewaehlt/CJK/CJK\\_pdf\\_04.pdf?\\_blob=publicationFile](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]“**  
**To the temperature sensitivity of prions it should be noted that in steam sterilization even under application of 18 minutes sterilization time at 134-137 ° C a certain proportion of prions remain infectious [1, 13] and dry heat has not eliminated residual infectivity even at extreme temperatures [14]“**

Castilla J, Saa P, Hetz C, Soto C (2005) In vitro generation of infectious scrapie prions. *Cell* 121, 195-206.

[Jackson](#) GS, [McKintosh](#) E, [Flechsig](#) E et al. (2005) **An enzyme–detergent method for effective prion decontamination of surgical steel.** *J Gen Virol* 86 no. 3 869-878 doi: 10.1099/vir.0.80484-0  
<http://jgv.sgmjournals.org/content/86/3/869.short>

Johnson RT. (2005) Prion diseases. *Lancet Neurol.* 4(10), 635-642

Tanaka M, Collins SR, Toyama BH, Weissman JS (2006) The physical basis of how prion conformations determine strain phenotypes. *Nature* 442, 585-589.

Collinge J, Clarke AR (2007) A general model of prion strains and their pathogenicity. *Science* 318, 930-936.

Deleault NR, Harris BT, Rees JR, Supattapone S (2007) Formation of native prions from minimal components in vitro. *Proc Natl Acad Sci U S A* 104, 9741-9746.

Mahal SP, Baker CA, Demczyk CA, Smith EW, Julius C, et al. (2007) Prion strain discrimination in cell culture: the cell panel assay. *Proc Natl Acad Sci U S A* 104, 20908-20913.

Richt JA, Kasinathan P, Hamir AN, Castilla J, Sathiyaseelan T, et al. (2007) Production of cattle lacking prion protein. *Nat Biotechnol* 25, 132-138.

Yang L, Zhou X, Yang J, Yin X, Han L, Zhao D. (2008) [Aspirin inhibits cytotoxicity of prion peptide PrP106-126 to neuronal cells associated with microglia activation in vitro.](#) *J Neuroimmunol.* 199 (1-2), 10-7. doi: 10.1016/j.jneuroim.2008.04.028.

Weissmann C (2009) Thoughts on mammalian prion strains. *Folia Neuropathol* 47: 104113.

- Ghaemmaghami S, Ahn M, Lessard P, Giles K, Legname G, et al. (2009) Continuous quinacrine treatment results in the formation of drug-resistant prions. *PLoS Pathog* 5: e1000673. doi:10.1371/journal.ppat.1000673.
- Manuelidis L (2010) Transmissible encephalopathy agents: virulence, geography and clockwork. *Virulence* 1, 101-104.
- Shorter J (2010) Emergence and natural selection of drug-resistant prions. *Mol Biosyst* 6, 1115-1130.
- Eisele YS, Obermüller U, Heilbronner G (2010) peripherally applied A $\beta$ -containing inoculates induce cerebral  $\beta$ -amyloidosis. *Science* 330, 980–2. [CrossRef](#) [MEDLINE](#) [PubMed Central](#)
- Morales R, Estrada LD, Diaz-Espinosa R et al. (2010) **Molecular cross talk between misfolded proteins in animal models of Alzheimer's and prion diseases.** *J Neurosci* 30, 4528–35. [CrossRef](#) [MEDLINE](#) [PubMed Central](#)
- Soscia SJ, Kirby JE, Washicosky KJ, et al. (2010) **The Alzheimer's disease-associated amyloid betaprotein is an antimicrobial peptide.** In: *PLoS ONE*. 5(3), e9505. doi:10.1371/journal.pone.0009505. PMID 20209079. Volltext bei PMC: 2831066. <http://www.ncbi.nlm.nih.gov/pubmed/20209079>
- Li J, Browning S, Mahal SP, Oelschlegel AM, Weissmann C (2010) Darwinian evolution of prions in cell culture. *Science* 327, 869-872.
- Wang F et al. (2010) Generating a prion with bacterially expressed recombinant prion protein. [Science](#), 327, 1132-35.
- Adapınar DÖ, Saylısoy S, Yenilmez C et al. (2011) The First Report of a Patient with Probable Variant Creutzfeldt-Jakob Disease in Turkey. *Dement Geriatr Cogn Dis Extra*. 1(1), 429–432. Published online 2011 December 24. doi: [10.1159/000332024](https://doi.org/10.1159/000332024) <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3265806/>
- Li J, Mahal SP, Demczyk CA, Weissmann C (2011) Mutability of prions. *EMBO Rep* 14, 191.
- Browning S, Baker CA, Smith E et al. (2011) Abrogation of complex glycosylation by Swainsonine results in strain- and cell-specific inhibition of prion replication. *J Biol Chem* 19, 19.
- Santo KP, Berjanskii M, Wishart DS et al. (2011) **Comparative analysis of essential collective dynamics and NMR-derived flexibility profiles in evolutionarily diverse prion proteins.** *Prion* 5(3), 188-200 <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3226046/>
- Hou F. et al. (2011) "MAVS forms functional prion-like aggregates to activate and propagate antiviral innate immune response," [Cell](#), 146, 448-61. <http://www.cell.com/retrieve/pii/S0092867411007161?cc=y>
- Weissmann C, Li J, Mahal SP et al. (2011) **Prions on the move.** *EMBO Rep* 12, 1109-1117.
- Carter C. (2011) Alzheimer's Disease: APP, Gamma Secretase, APOE, CLU, CR1, PICALM, ABCA7, BIN1, CD2AP, CD33, EPHA1, and MS4A2, and Their Relationships with Herpes Simplex, C. Pneumoniae, Other Suspect Pathogens, and the Immune System. doi: [10.4061/2011/501862](https://doi.org/10.4061/2011/501862) PMID: PMC3255168 <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3255168/?tool=pubmed>
- Shin DM, Eun-Kyeong Jo (2011) Antimicrobial Peptides in Innate Immunity against Mycobacteria. *Immune Netw*. 11(5), 245–252. <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3242998/?tool=pubmed>
- Jeong JK, Moon MH, Seol JW, Seo JS, Lee YJ, Park SY. (2011) [Prion peptide-mediated cellular prion protein overexpression and neuronal cell death can be blocked by aspirin treatment.](#) *Int J Mol Med*. 27(5), 689-93. doi: 10.3892/ijmm.2011.626
- Zhou M, Ottenberg G, Sferrazza GF. (2012) Highly neurotoxic monomeric  $\alpha$ -helical prion protein. <http://www.pnas.org/content/early/2012/02/07/1118090109>

Harris F, Dennison SR, Phoenix DA. (2012) **Aberrant action of amyloidogenic host defense peptides: a new paradigm to investigate neurodegenerative disorders?** doi: 10.1096/fj.11-199208 <http://www.fasebj.org/content/early/2012/02/03/fj.11-199208.abstract>

Grübler B (2012) Alzheimer-Demenz: Die Forschung steht unter Druck. Dtsch Arztebl 109(1–2), 26–8. [VOLLTEXT](#)

R. Halfmann et al. (2012) Prions are a common mechanism for phenotypic inheritance in wild yeasts, Nature, 482, 363-8. <http://web.wi.mit.edu/lindquist/pub/Publications.html>

Marchant Jo. (2012) Prion diseases hide out in the spleen. UK population could harbour thousands of silent infections. <http://www.nature.com/news/prion-diseases-hide-out-in-the-spleen-1.9904>

Wang F et al (2012) Genetic informational RNA is not required for recombinant prion infectivity. J Virol. 86(3), 1874-6 <http://jvi.asm.org/content/86/3/1874.abstract>

Blood test for mad cow disease (2012) <http://www.channel4.com/news/blood-test-breakthrough-for-mad-cow-disease>

Saunders SE, Bartelt-Hunt SL, Bartz JC (2012) Occurrence, Transmission, and Zoonotic Potential of Chronic Wasting Disease. Emerging Infectious Diseases • www.cdc.gov/eid • Vol. 18, No. 3 <http://wwwnc.cdc.gov/eid/article/18/3/pdfs/11-0685.pdf>

Liu L, Drouet V, Wu JW et al. (2012) Trans-synaptic spread of Tau pathology in vivo. PLoS One 7, e3102. [MEDLINE](#)

Siegmund Schultze N. (2012) Neurodegenerative Erkrankungen. Gemeinsamkeiten mit Prionen. Deutsches Ärzteblatt 109, 12 <http://www.aerzteblatt.de/lit1212>

Creutzfeldt-Jakob-Krankheit (CJK) <http://de.wikipedia.org/wiki/Creutzfeldt-Jakob-Krankheit>

CJD Figures (Statistik1990 bis 2012) <http://www.cjd.ed.ac.uk/figures.htm>  
<http://www.cjd.ed.ac.uk/index.htm>

[Miyazawa K](#), [Kipkorir T](#), [Tittman S](#), [Manuelidis L](#). (2012) **Continuous production of prions after infectious particles are eliminated: implications for Alzheimer's disease.** *PLoS One*. 7(4), e35471. Epub 2012 Apr 11. <https://journals.plos.org/plosone/article?id=10.1371/journal.pone.0035471>

Soto C (2012) Transmissible proteins: expanding the prion heresy. *Cell*, 149, 968-77

Halfmann R et al. (2012) Prions are a common mechanism for phenotypic inheritance in wild yeasts. *Nature*, 482, 363-68

**University of Edinburgh (2012) The Nationan CJDRSU** <http://www.cjd.ed.ac.uk/data.html>

[Bastian FO](#), [Elzer PH](#), [Wu X](#). (2012) **Spiroplasma spp. biofilm formation is instrumental for their role in the pathogenesis of plant, insect and animal diseases.** *Exp Mol Pathol*. 93(1), 116-28. doi: 10.1016/j.yexmp.2012.04.017. <http://www.ncbi.nlm.nih.gov/pubmed/22552100>

Kim HJ et al. (2013) Mutations in prion-like domains in hnRNPA2B1 and hnRNPA1 cause multisystem proteinopathy and **ALS**. *Nature*, doi:10.1038/nature11922. <http://www.the-scientist.com/?articles.view/articleNo/34571/title/Prion-like-Proteins-Cause-Disease/>

Gill et al. (2013) **Prevalent abnormal prion protein in human appendixes after bovine spongiform encephalopathy epizootic: large scale survey.** *BMJ* 347, f5675 <http://dx.doi.org/10.1136/bmj.f5675>

Salmon R (2013) **How widespread is variant Creutzfeldt-Jakob disease?** *BMJ* 347, f5994 <http://dx.doi.org/10.1136/bmi.f5994> <http://www.bmj.com/content/347/bmj.f5994>

Hannaoui S, Gougerot A, Privat N, Levavasseur E, Bizat N, Hauw JJ, Brandel JP, Haïk S. (2013) **Cycline** efficacy on the propagation of human prions in primary cultured neurons is strain-specific. *J Infect Dis*. <http://www.ncbi.nlm.nih.gov/pubmed/24265435>

**“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>  
<http://www.hindawi.com/journals/ijcb/2013/975832/>

Holmes DL et al. (2013) Heritable remodeling of yeast multicellularity by an environmentally responsive prion. *Cell*, 153, 153-65.

Goñi F, Mathiason CK, Yim L et al. (2014) **Mucosal immunization with an attenuated Salmonella vaccine partially protects white-tailed deer from chronic wasting disease**. *Vaccine*, DOI: [10.1016/j.vaccine.2014.11.035](https://doi.org/10.1016/j.vaccine.2014.11.035)

Liberski PP (2014) **Prion, prionoids and infectious amyloid**. *Parkinsonism Relat Disord*. 20S1, 80-84. doi: 10.1016/S1353-8020(13)70021-X. <http://www.ncbi.nlm.nih.gov/pubmed/24262195>

**““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  $\beta$ -sheet. ... There are several protein misfolding disorders - the most widely known include Alzheimer's disease, Parkinson's disease and other  $\alpha$ -synucleinopathies, amyotrophic lateral sclerosis (ALS), frontotemporal dementias in which abnormally phosphorylated MAP- $\tau$  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.”**

ProMED Mail post (2014) **BSE, bovine - Germany (02): (BB) new case, atypical H-type, OIE**. <http://www.promedmail.org/post/20140225.2300162>

Jackson GS, Burk-Rafel J, Edgeworth JA et al. (2014) **Population Screening for Variant Creutzfeldt-Jakob Disease Using a Novel Blood Test Diagnostic Accuracy and Feasibility Study**. *JAMA Neurol*. Published online doi:10.1001/jamaneurol.2013.6001  
<http://archneur.jamanetwork.com/article.aspx?articleid=1834620>

Orrú CD, Bongianni M, Tonoli G, Ferrari S, Hughson AG et al. (2014) **A Test for Creutzfeldt–Jakob Disease Using Nasal Brushings**. *N Engl J Med* 371, 519-529 [August 7, 2014](https://doi.org/10.1056/NEJMoa1315200) DOI: 10.1056/NEJMoa1315200

Minikel EV (2014) **Why doxycycline's failure in CJD clinical trials is no surprise**. <http://www.cureffi.org/2014/02/04/why-doxycyclines-failure-in-cjd-clinical-trials-is-no-surprise/>  
**„The result in two words: no efficacy“.**

Bastian FO (2014) **The case for involvement of spiroplasma in the pathogenesis of transmissible spongiform encephalopathies**. *J Neuropathol Exp Neurol*. 73(2), 104-14. doi: 10.1097/NEN.0000000000000033. <https://www.ncbi.nlm.nih.gov/pubmed/24423635>  
**« Research efforts should focus on this bacterium for development of therapeutic regimens for Creutzfeldt-Jakob disease. »**

Pritzkow S, Morales R (2015) **Grass Plants Bind, Retain, Uptake, and Transport Infectious Prions**. *Cell Reports* 11, 1168–1175 <http://dx.doi.org/10.1016/j.celrep.2015.04.036>  
<http://www.cell.com/cell-reports/pdfExtended/S2211-1247%2815%2900437-4>

Burchell JT, Panegyres P (2016) **Prion diseases: immunotargets and therapy**. [ImmunoTargets and Therapy](https://doi.org/10.1080/17513758.2016.1185888) » [Volume 5](https://doi.org/10.1080/17513758.2016.1185888)  
<https://www.dovepress.com/prion-diseases-immunotargets-and-therapy-peer-reviewed-fulltext-article-ITT>



Boelaert F, Hugas M, Pelaez AO et al. (2016) **The European Union summary report on data of the surveillance of ruminants for the presence of transmissible spongiform encephalopathies (TSEs) in 2015.** European Food Safety Authority (EFSA) [Full publication history](#) DOI: 10.2903/j.efsa.2016.4643 [View](#) <http://onlinelibrary.wiley.com/doi/10.2903/j.efsa.2016.4643/full>

Bougard D, Brandel J-P, Bélondrade M et al. (2016) **Detection of prions in the plasma of presymptomatic and symptomatic patients with variant Creutzfeldt-Jakob disease.** *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/). <https://www.ncbi.nlm.nih.gov/labs/articles/28099827/>  
<http://www.nejm.org/doi/full/10.1056/NEJMc1610003>

Zylka-Menhorn, V (2017) **Creutzfeldt-Jakob-Krankheit: Der jüngste Fall passt nicht in das bisherige Muster.** *Dtsch Arztebl* 114(6) A-266 / B-238 / C-235  
<https://www.aerzteblatt.de/archiv/186263/Creutzfeldt-Jakob-Krankheit-Der-juengste-Fall-passt-nicht-in-das-bisherige-Muster>  
<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)**“.

Waddell L, Greig J, Mascarenhas M et al. (2017) **Current evidence on the transmissibility of chronic wasting disease prions to humans -- A systematic review.** *Transbound Emerg Dis*. 00: 1-13. doi:10.1111/tbed.12612; <http://onlinelibrary.wiley.com/doi/10.1111/tbed.12612/full>.

Babelhadj B, Di Bari MA, Pirisinu L, Chiappini B, Gaouar SBS, Riccardi G et al. (2018) **Prion disease in dromedary camels, Algeria.** *Emerg Infect Dis*. <https://doi.org/10.3201/eid2406.172007>  
DOI: 10.3201/eid2406.172007


**Toward Therapy of Human Prion Diseases.** *2018 Annual Review of Pharmacology and Toxicology* 58(1), 331-351 DOI: 10.1146/annurev-pharmtox-010617-052745 Adriano Aguzzi, [Asvin K.K. Lakkaraju](#), [Karl Frontzek](#)  
<https://academic.microsoft.com/paper/2588040733/reference/search?q=Toward%20Therapy%20of%20Human%20Prion%20Diseases&qe=%2540%2540%2540REFERENCES%253D2588040733&f=&orderBy=0>

Moreno JA, Halliday M, Molloy C et al. (2013) **Oral Treatment Targeting the Unfolded Protein Response Prevents Neurodegeneration and Clinical Disease in Prion-Infected Mice.** *Science Translational Medicine* 5(206), 206ra138 DOI: 10.1126/scitranslmed.3006767 <https://www.ncbi.nlm.nih.gov/pubmed/24107777>

**We show that oral treatment with a specific inhibitor of the kinase PERK (protein kinase RNA-like endoplasmic reticulum kinase), a key mediator of this UPR pathway, prevented UPR-mediated translational repression and abrogated development of clinical prion disease in mice, with neuroprotection observed throughout the mouse brain.**

**Wir zeigen, dass die orale Behandlung mit einem spezifischen Inhibitor der Kinase PERK (Proteinkinase-RNA-ähnliche endoplasmatische Retikulumkinase), einem Schlüsselmediator dieses UPR-Signalwegs, die UPR-vermittelte translationale Repression verhinderte und die Entwicklung einer klinischen Prionkrankheit bei Mäusen unter Neuroprotektion verhinderte und zwar im gesamten Maushirn.**

- ➔ Zabel M, Warner R, Brasch S (2017) Can Scientists Stop The 'Zombie Disease' Killing Elk And Deer?  
<http://www.cpr.org/news/story/can-scientists-stop-the-zombie-disease-killing-elk-and-deer>  
<http://www.cpr.org/news/story/could-colorado-s-zombie-deer-disease-ever-spread-to-people>
- ➔ (2005) Scientists Observe Infectious Prion Proteins Invade and Move Within Brain Cells.  
[https://www.eurekalert.org/pub\\_releases/2005-05/nioa-soi052305.php](https://www.eurekalert.org/pub_releases/2005-05/nioa-soi052305.php)

[Bernt - Dieter Huismans](#), last revision January 2020 [www.Huismans.click](http://www.Huismans.click)   
Back to top: <http://www.erlebnishaft.de/prione.pdf>