

6 References

- Alper, T., D. A. Haig and M. Clarke (1966). "The exceptionally small size of the scrapie agent." Biochem. Biophys. Res. Commun. 22: 278-84.
- Appel, T. R., C. Dumpitak, U. Matthiesen and D. Riesner (1999). "Prion rods contain an inert polysaccharide scaffold." Biol. Chem. 380: 1295-1306.
- Appel, T. R., M. Wolff, F. v. Rheinbaden, M. Heinzl and D. Reisner (2001). "Heat stability of prion rods and recombinant prion protein in water, lipid and lipid-water mixtures." J. Gen. Virol. 82: 465-473.
- Arjona, A., L. Simarro, F. Islinger, N. Nishida and L. Manuelidis (2004). "Two Creutzfeldt-Jakob disease agents reproduce prion protein-independent identities in cell cultures." Proc. Natl Acad. Sci. U.S.A. 101(23): 8768-8773.
- Arnold, J., C. Tipler, L. Laszlo, J. Hope, M. Landon and R. Mayer (1995). "The abnormal isoform of the prion protein accumulates in late-endosome-like organelles in scrapie-infected mouse brain." J. Pathol. 176: 403-411.
- Arosio, P., T. G. Adelman and J. W. Drysdale (1978). "On ferritin heterogeneity. Further evidence for heteropolymers." J. Biol. Chem. 253(12): 4451-4458.
- Ashwath, M., S. Dearmond and T. Culclasure (2005). "Prion-associated dilated cardiomyopathy." Arch Intern Med. 14: 338-340.
- Aucouturier, P., R. J. Kascsak, B. Frangione and T. Wisniewski (1999). "Biochemical and conformational variability of human prion strains in sporadic Creutzfeldt-Jakob disease." Neurosci. Lett. 274(1): 33-36.
- Backmann, J., C. Schultz, H. Fabian, U. Hahn, W. Saenger and D. Naumann (1996). "Thermally induced hydrogen exchange processes in small proteins as seen by FTIR spectroscopy." Proteins 24: 379-387.
- Bamborough, P., H. Wille, G. C. Telling, F. Yehiely, S. B. Prusiner and F. C. Cohen (1996). "Prion Protein Structure and Scrapie Replication: Theoretical, Spectroscopic, and Genetic Investigation." Cold Spring Harb. Symp. Quant. Biol. 61: 495-509.
- Bandekar, J. (1992). "Amide modes and protein conformation." Biochim. Biophys. Acta 1120: 123-143.
- Baron, G. S., K. Wehrly, D. W. Dorward, C. Bruce and B. Caughey (2002). "Conversion of raft associated prion protein to the protease-resistant state requires insertion of PrP-res (PrP^{Sc}) into contiguous membranes." EMBO J. 21: 1031-1040.
- Barrachina, M., E. Dalfo, B. Puig, N. Vidal, M. Freixes, E. Castano and I. Ferrer (2005). "Amyloid- β deposition in the cerebral cortex in Dementia with Lewy bodies is accompanied by a relative increase in A β PP mRNA isoforms containing the Kunitz protease inhibitor." Neurochem. Int. 46(3): 253-260.
- Barth, A. (2000). "The infrared absorption of amino acid side chains." Prog. Biophys. Mol. Biol. 74: 141-173.
- Barth, A. and C. Zscherp (2002). "What vibrations tell us about proteins." Q. Rev. Biophys. 35: 369-430.
- Baskakov, I. V. and O. V. Bocharova (2005). "In Vitro Conversion of Mammalian Prion Protein into Amyloid Fibrils Displays Unusual Features." Biochemistry 44: 2339 - 2348.
- Basler, K., B. Oesch, Scott M, Westaway D, Walchli M, Groth DF, McKinley MP, Prusiner SB and W. C (1986). "Scrapie and cellular PrP isoforms are encoded by the same chromosomal gene." Cell 46: 417-428.

References

- Bastian, F. O., S. Dash and R. F. Garry (2004). "Linking chronic wasting disease to scrapie by comparison of *Spiroplasma mirum* ribosomal DNA sequences." Exp. Mol. Pathol. 77(1): 49-56.
- Beekes, M., E. Baldauf, S. Caßens, H. Diringer, P. Keyes, A. C. Scott, G. A. Wells, P. Brown, C. J. Gibbs and D. C. Gajdushek (1995). "Western blot mapping of disease-specific amyloid in various animal species and humans with transmissible spongiform encephalopathies using a high-yield purification method." J. Gen. Virol. 76: 2567-2579.
- Beekes, M., E. Baldauf and H. Diringer (1996). "Sequential appearance and accumulation of pathognomonic markers in the central nervous system of hamsters orally infected with scrapie." J. Gen. Virol. 77: 1952-1934.
- Beekes, M., P. McBride and E. Baldauf (1998). "Cerebral targeting indicates vagal spread of infection in hamsters fed with scrapie." J. Gen. Virol. 79: 601-607.
- Begara-McGorum, I., L. Gonzalez, M. Simmons, N. Hunter, F. Houston and M. Jeffrey (2002). "Vacuolar Lesion Profile in Sheep Scrapie: Factors Influencing its Variation and Relationship to Disease-specific PrP Accumulation." J. Comp. Pathol. 127(1): 59-68.
- Bendheim, P., B. HR., R. RD., S. LJ., G. NL., W. GY., K. RJ., C. NR. and B. DC. (1992). "Nearly ubiquitous tissue distribution of the scrapie agent precursor protein." Neurology 42: 149-156.
- Beringue, V., G. Mallinson, M. Kaiser, M. Tayebi, Z. Sattar, G. Jackson, D. Anstee, J. Collinge and S. Hawke (2003). "Regional heterogeneity of cellular prion protein isoforms in the mouse brain." Brain 126(9): 2065-2073.
- Bessen, R. A. and R. F. March (1992). "Biochemical and physical properties of the prion protein from two strains of the transmissible mink encephalopathy agent." J. Virol. 66: 2096-2101.
- Bessen, R. A. and R. F. Marsh (1994). "Distinct PrP Proteins Suggest the Molecular basis of Strain Variation in Transmissible Mink Encephalopathy." J. Virol. 68: 7859-7868.
- Bieschke, J., P. Weber, N. Sarafoff, M. Beekes, A. Giese and H. Kretzschmar (2004). "Autocatalytic self-propagation of misfolded prion protein." Proc. Natl Acad. Sci. U.S.A. 101(33): 12207-12211.
- Bolton, D. C. (1998). "Prion distribution in hamster lung and brain following intraperitoneal inoculation." J. Gen. Virol. 79: 2557-2562.
- Bolton, D. C. and P. E. Bendheim (1991). "Purification of Scrapie Agents: How Far Have We Come?" CTMI 172: 39-55.
- Bolton, D. C., P. E. Bendheim, A. D. Marmorstein and A. Potempska (1987). "Isolation and Structural Studies of the Intact Scrapie Agent Protein." Arch. Biochem. Biophys. 258: 579-590.
- Bolton, D. C., M. P. McKimley and S. B. Prusiner (1982). "Identification of a protein that purifies with the scrapie prion." Science 218: 1309-1310.
- Bolton, D. C., R. K. Meyer and S. B. Prusiner (1985). "Scrapie PrP 27-30 Is a Sialoglycoprotein." J. Virol. 53: 596-606.
- Borchelt, D., M. Scott, A. Taraboulos, N. Stahl and S. Prusiner (1990). "Scrapie and cellular prion proteins differ in their kinetics of synthesis and topology in cultured cells." J. Cell Biol. 110(3): 743-752.
- Brown, D. R., I. K. Jordanova, B.-S. Wong, C. Venien-Bryan, F. Hafiz, L. L. Glasssmith, M.-S. Sy, P. Gambetti, I. M. Jones, C. Clive and S. J. Haswell (2000). "Functional and structural differences between the prion protein from two alleles prnp(a) and prnp(b) of mouse." Eur. J. Biochem. 267(8): 2452-2459.

- Brown, P. and R. Bradley (1998). "1755 and all that: a historical primer of transmissible spongiform encephalopathy." BMJ 317(7174): 1688-1692.
- Broxmeyer, L. (2004). "Is mad cow disease caused by a bacteria?" Med. Hypotheses 63(4): 731-739.
- Bruce, M., A. Chree, I. McConnell, J. Foster, G. Pearson and H. Fraser (1994). "Transmission of bovine spongiform encephalopathy and scrapie to mice: strain variation and the species barrier." Philos. Trans. R. Soc. Lond., B, Biol. Sci. 343(1306): 405-411.
- Bruce, M. and A. Dickinson (1987). "Biological evidence that scrapie agent has an independent genome." J. Gen. Virol. 68: 79-89.
- Bruce, M. and H. Fraser (1982). "Focal and asymmetrical vacuolar lesions in the brains of mice infected with certain strains of scrapie." Acta Neuropathol. 58: 133-140.
- Bruce, M. E. (2003). "TSE strain variation." Br. Med. Bull. 66(1): 99-108.
- Bruce, M. E. and H. Fraser (1991). "Scrapie strain Variation and its Implications." CTMI 172: 125-138.
- Bruce, M. E., R. G. Will, J. W. Ironside, I. McConnell, D. Drummond, A. Suttie, L. McCardle, A. Chree, J. Hope, C. Birket, S. Cousens, H. Fraser and C. J. Bostock (1997). "Transmissions to mice indicate that 'new variant' CJD is caused by the BSE agent." Nature 389: 498-501.
- Burnette, W. N. (1981). "'Western blotting': electrophoretic transfer of proteins from sodium dodecyl sulfate--polyacrylamide gels to unmodified nitrocellulose and radiographic detection with antibody and radioiodinated protein A." Anal. Biochem. 112(2): 195-203.
- Byler, D. M. and H. Susi (1986). "Examination of the Secondary Structure of Proteins by Deconvolved FTIR Spectra." Biopolymers 25: 469-487.
- Carp, R., S. Callahan, E. Sersen and R. Moretz (1984). "Preclinical changes in weight of scrapie-infected mice as a function of scrapie agent-mouse strain combination." Intervirology 21: 61-69.
- Carp, R. I., H. Meeker, E. Sersen and P. Kozlovski (1998). "Analysis of the incubation periods, induction of obesity and histopathological changes in senescence-prone and senescence-resistant mice infected with various scrapie strains." J. Gen. Virol. 79: 2863-2869.
- Carp, R. I., H. Meeker and E. Sersen (1997). "Scrapie strains retain their distinctive characteristic following passages of homogenates from different brain regions and spleen." J. Gen. Virol. 78: 283-290.
- Castilla, J., P. Saa, C. Hetz and C. Soto (2005). "In Vitro Generation of Infectious Scrapie Prions." Cell 121(2): 195.
- Caughey, B. (2003). "Prion protein conversions: insight into mechanisms, TSE transmission barriers and strains." Br. Med. Bull. 66(1): 109-120.
- Caughey, B., K. Neary, R. Buller, D. Ernst, L. Perry, B. Chesebro and R. RE. (1990). "Normal and scrapie-associated forms of prion protein differ in their sensitivities to phospholipase and proteases in intact neuroblastoma cells." J. Virol. 64: 1093-1101.
- Caughey, B., R. E. Race, D. Ernst, M. J. Buchmeier and B. Chesebro (1989). "Prion protein biosynthesis in scrapie-infected and uninfected neuroblastoma cells." J. Virol. 63: 175-181.
- Caughey, B. and G. Raymond (1991a). "The scrapie-associated form of PrP is made from a cell surface precursor that is both protease- and phospholipase-sensitive." J. Biol. Chem. 266(27): 18217-18223.
- Caughey, B., G. J. Raymond and R. Bessen (1998). "Strain-dependent Differences in beta Sheet Conformations of Abnormal Prion Protein." J. Biol. Chem.: 32230-32235.

References

- Caughey, B. W., A. Dong, K. S. Bath, D. Ernst, S. F. Hayest and W. S. Caughey (1991b). "Secondary Structure Analysis of the Scrapie-Associated Protein PrP 27-30 in Water by Infrared Spectroscopy." Biochemistry 30: 7672-7678.
- Chen, S. G., D. B. Teplow, P. Parchi, J. K. Teller, P. Gambetti and L. Autilio-Gambetti (1995). "Truncated Forms of the Human Prion Protein in Normal Brain and in Prion Diseases." J. Biol. Chem. 270(32): 19173-19180.
- Chesebro, B. (2003). "Introduction to the transmissible spongiform encephalopathies or prion diseases." Br. Med. Bull. 66(1): 1-20.
- Chesebro, B., M. Trifilo, R. Race, K. Meade-White, C. Teng, R. LaCasse, L. Raymond, C. Favara, G. Baron, S. Priola, B. Caughey, E. Masliah and M. Oldstone (2005). "Anchorless Prion Protein Results in Infectious Amyloid Disease Without Clinical Scrapie." Science 308(5727): 1435-1439.
- Choi, J., H. D. Rees, S. T. Weintraub, A. I. Levey, L.-S. Chin and L. Li (2005). "Oxidative modifications and aggregation of Cu/Zn superoxide dismutase associated with Alzheimer's and Parkinson's diseases." J. Biol. Chem.: M414327200.
- Colchester, A. C. F. and N. T. H. Colchester (2005). "The origin of bovine spongiform encephalopathy: the human prion disease hypothesis." The Lancet 366(9488): 856.
- Collinge, J., KCLSidle, M. J, I. J and H. AF (1996). "Molecular analysis of prion strain variation and the aetiology of 'new variant' CJD." Nature 383: 685-690.
- Collins, S., M. G. Law, A. Fletcher, A. Boyd, J. Kaldor and C. L. Masters (1999). "Surgical treatment and risk of sporadic Creutzfeldt-Jakob disease: a case-control study." Lancet 353(9154): 693-697.
- Collins, S., C. A. McLean and C. L. Masters (2001). "Gerstmann-Straussler-Scheinker syndrome, fatal familial insomnia, and kuru: a review of these less common human transmissible spongiform encephalopathies." J Clin Neurosci 8(5): 387-397.
- Collins, S. R., A. Douglass, R. D. Vale and J. S. Weissman (2004). "Mechanism of Prion Propagation: Amyloid Growth Occurs by Monomer Addition." PLoS Biology 2(10): e321.
- Cordeiro, Y., F. Machado, L. Juliano, M. A. Juliano, R. R. Brentani, D. Foguel and J. L. Silva (2001). "DNA converts cellular prion protein into the beta-sheet conformation and inhibits prion peptide aggregation." J Biol Chem 276(52): 49400-9.
- Couzin, J. (2004). "BIOMEDICINE: An End to the Prion Debate? Don't Count on It." Science 305(5684): 589a-.
- Czub, M., H. R. Braig and H. Diringer (1986). "Pathogenesis of scrapie: study of the temporal development of clinical symptoms, of infectivity titres and scrapie-associated fibrils in brains of hamsters infected intraperitoneally." J. Gen. Virol. 67: 2005-2009.
- DeArmond, S., M. McKinley, R. Barry, M. Braunfeld, J. McColloch and S. Prusiner (1985). "Identification of prion amyloid filaments in scrapie-infected brain." Cell 41: 221-235.
- DeArmond, S. J., S.-L. Yang, A. Lee, R. Bowler, A. Taraboulos, D. Groth and S. B. prusiner (1993). "Three scrapie prion isolats exhibit different accumulation patterns of the prion protein scrapie isoform." proc. Natl. Acad. Sci. U.S.A 90: 6449-6453.
- DeBurman, S. K., G. J. Raymond, B. Gaughey and S. Lindquist (1997). "Chaperone-supervised conversion of prion protein to its protease-resistant form." Proc. Natl. Acad. Sci. U.S.A 94: 13938-13943.
- Deleault, N. R., R. W. Lucassen and S. Supattapone (2003). "RNA molecules stimulate prion protein conversion." Nature 425(6959): 717-20.
- Dell'Omo, G., E. Vannoni, A. L. Vyssotski, M. A. Di Bari, R. Nonno, U. Agrimi and H.-P. Lipp (2002). "Early behavioural changes in mice infected with BSE and scrapie: automated home cage monitoring reveals prion strain differences." Eur J Neurosci 16(4): 735-742.

- Dickinson, A. and V. Meikle (1971). "Host-genotype and agent effects in scrapie incubation: change in allelic interaction with different strains of agent." Mol. Gen. Genet. 112: 73-79.
- Dickinson, A., V. Meikle and H. Fraser (1968). "Identification of a gene which controls the incubation period of some strains of scrapie agent in mice." J. Comp. Pathol. 78: 293-299.
- Dickinson, A. G. (1976). Slow virus diseases of animals and man Scrapie in sheep and goats. Amsterdam, North-Holland Publishing Company.
- Dickinson, A. G. and G. W. Outram (1988). "Genetic aspects of unconventional virus infections: the basis of the virino hypothesis." Ciba Found. Symp. 135: 63-83.
- Diringer, H. (1991). "Transmissible spongiform encephalopathies (TSE) virus-induced amyloidoses of the central nervous system (CNS)." Eur. J. Epidemiol. 7: 562-566.
- Diringer, H., M. Beekes, M. Özel, D. Simon, I. Queck, F. Cardone, M. Pocchiari and J. W. Ironside (1997). "Highly infectious purified preparations of disease-specific amyloid of transmissible spongiform encephalopathies are not devoid of nucleic acids of viral size." Intervirology 40: 238-246.
- Diringer, H., H. Hilmert, D. Simon, E. Werner and B. Ehlers (1983). "Towards Purification of Scrapie Agent." Eur. J. Biochem 134: 555-560.
- Dobson, C. (2001). "The structural basis of protein folding and its links with human disease." Philos. Trans. R. Soc. Lond., B, Biol. Sci. 356: 133-45.
- Dong, A., P. Huang and W. S. Caughey (1990). "Protein Secondary Structures in Water from Second-Derivative Amide I Infrared Spectra." Biochemistry 29: 3303-3308.
- Donne, D. G., J. H. Viles, D. Groth, I. Mehlhorn, T. L. James, F. E. Cohen, S. B. Prusiner, P. E. Wright and H. J. Dyson (1997). "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. 94(25): 13452-13457.
- Ebringer, A., T. Rashid and C. Wilson (2005). "Bovine Spongiform Encephalopathy, Multiple Sclerosis, and Creutzfeldt-Jakob Disease Are Probably Autoimmune Diseases Evoked by Acinetobacter Bacteria." Ann NY Acad Sci 1050(1): 417-428.
- Ermonval, M., S. Mouillet-Richard, P. Codogno, O. Kellermann and J. Botti (2003). "Evolving views in prion glycosylation: functional and pathological implications." Biochimie 85(1-2): 33-45.
- Fabian, H., L.-P. i. Choo, G. I. Szenderei, M. Jackson, W. C. Halliday, L. Otvos and H. H. Mantsch (1993). "Infrared Spectroscopic Characterization of Alzheimer Plaques." Appl. Spectrosc. 47: 1513-1518.
- Fabian, H., P. Lasch, M. Boese and W. Haensch (2003). "Infrared microspectroscopic imaging of benign breast tumor tissue sections." Journal of Molecular Structure 661-662: 411.
- Fabian, H. and W. Mäntele (2002). Infrared Spectroscopy of Proteins. Handbook of Vibrational Spectroscopy. J. M. C. a. P. R. Griffiths. Chichester, John Wiley & Sons Ltd. 5: 3399-3425.
- Fabian, H. and H. H. Mantsch (1995). "Ibionuclease A Revisited: Infrared Spectroscopic Evidence for Lack of Native-like Secondary Structures in the Thermally Denatured State." Biochemistry 34: 13651-13655.
- Fabian, H. and C. P. Schultz (2000). Fourier Transform Infrared Spectroscopy in Peptide and Protein Analysis. Encyclopedia of Analytical Chemistry: 5779-5803.
- Fernaes, S. and T. Land (2005). "Increased iron-induced oxidative stress and toxicity in scrapie-infected neuroblastoma cells." Neuroscience Letters 382(3): 217.

References

- Fraser, H. and A. Dickinson (1985). "Targeting of scrapie lesions and spread of agent via the retino-tectal projection." Brain Res. 346: 32-41.
- Fraser, H. and A. G. Dickinson (1967). "Distribution of experimentally induced scrapie lesions in the brain." Nature 216: 1310-1311.
- Fraser, H. and A. G. Dickinson (1968). "The Sequential development of the Brain Lesions of Scrapie in Three Strains of Mice." J. Comp. Path. 78: 301-310.
- Gajdushek, D. C. (1977). "Unconventional Viruses and the Origin and Disappearance of Kuru." Science 197: 943-960.
- Gambetti, P. (1996). Fatal familial insomnia and familial Creutzfeldt-Jakob disease: A tale of two diseases with the same genetic mutation, Springer-Verlag Berlin Heidelberg New York.
- Gambetti, P., Q. Kong, W. Zou, P. Parchi and S. G. Chen (2003a). "Sporadic and familial CJD: classification and characterisation." Br. Med. Bull. 66(1): 213-239.
- Gambetti, P., P. Parchi and S. Chen (2003b). "Hereditary Creutzfeldt-Jakob disease and fatal familial insomnia." Clin Lab Med 23: 43-64.
- Gasset, M., M. A. Baldwin, R. J. Fletterick and S. B. Prusiner (1993). "Perturbation of the secondary structure of the scrapie prion protein under conditions that alter infectivity." Proc. Natl Acad. Sci. U.S.A. 90: 1-5.
- Glover, J. R., A. S. Kowal, E. C. Schirmer, M. M. Patino, J. J. Liu and S. Lindquist (1997). "Self-seeded fibers formed by Sup35, the protein determinant of [PSI+], a heritable prion-like factor of *S. cerevisiae*." Cell 89(5): 811-9.
- Goormaghtigh, E. (1990). "Secondary structure and dosage of soluble and membrane proteins by attenuated total reflection Fourier-transform infrared spectroscopy on hydrated films." Eur. J. Biochem. 193: 409-420.
- Govaerts, C., H. Wille, S. B. Prusiner and F. E. Cohen (2004). "Evidence for assembly of prions with left-handed {beta}-helices into trimers." Proc. Natl Acad. Sci. U.S.A. 101(22): 8342-8347.
- Gretzschel, A., A. Buschmann, M. Eiden, U. Ziegler, G. Luhken, G. Erhardt and M. H. Groschup (2005). "Strain typing of German transmissible spongiform encephalopathies field cases in small ruminants by biochemical methods." J. Vet. Med. B Infect. Dis. Vet. Public Health 52(2): 55-63.
- Griffith, J. S. (1967). "Self-replication and scrapie." Nature 215: 1043-4.
- Groschup, M. H., M. Beekes, P. A. McBride, M. Hardt, J. A. Hainfellner and H. Budka (1999). "Deposition of disease-associated prion protein involves the peripheral nervous system in experimental scrapie." Acta Neuropathol. 98: 453-457.
- Hadlow, W. J. (1999). "Reflections on the transmissible spongiform encephalopathies." Vet. Pathol. 36(6): 523-529.
- Haraguchi, T., S. Fisher, S. Olofsson, T. Endo, D. Groth, A. Tarentino, D. Borchelt, D. Teplow, L. Hood and A. Burlingame (1989). "Asparagine-linked glycosylation of the scrapie and cellular prion proteins." Arch. Biochem. Biophys. 274: 1-13.
- Harris, D. A. (1999). "Cellular Biology of Prion Diseases." Clin. Microbiol. Rev. 12: 429-444.
- Hegde, R., P. Tremblay, D. Groth, S. DeArmond, S. Prusiner and V. Lingappa (1999). "Transmissible and genetic prion diseases share a common pathway of neurodegeneration." Nature 402: 822-6.
- Helm, D., H. Labischinski, G. Schallehn and D. Naumann (1991). "Classification and identification of bacteria by Fourier-transform infrared spectroscopy." J Gen Microbiol 137: 69-79.
- Hill, A., Desbruslais M, Joiner S, Sidle KC, Gowland I, Collinge J, Doey LJ and L. P (1997). "The same prion strain causes vCJD and BSE." Nature 389: 448-450.

- Hill, A. F., S. Joiner, J. D. F. Wadsworth, K. C. L. Sidle, J. E. Bell, H. Budka, J. W. Ironside and J. Collinge (2003). "Molecular classification of sporadic Creutzfeldt-Jakob disease." Brain 126(6): 1333-1346.
- Hilmert, H. and H. Diring (1984). "A rapid and efficient method to enrich SAF-protein from scrapie brains of hamsters." Biosci. Rep. 4: 165-70.
- Horiuchi, M., T. Nemoto, N. Ishiguro, H. Furuoka, S. Mohri and M. Shinagawa (2002). "Biological and Biochemical Characterization of Sheep Scrapie in Japan." J. Clin. Microbiol. 40(9): 3421-3426.
- Hsiao, K., H. F. Baker, T. Crow, M. Poulter, F. Owen, J. D. Terwilliger, D. Westaway, J. Ott and S. B. Prusiner (1989). "Linkage of a prion protein missense variant to Gerstmann-Sträussler syndrome." Nature 338: 342-345.
- Hunter, G. D. (1979). The enigma of the scrapie agent: Biochemical approach and the involvement of membranes and nucleic acids. Slow transmissible diseases of the nervous system. S. B. Prusiner and W. J. Hadlow. New York London Sydney Toronto San Francisco, Academic Press. 2: 365-385.
- Jackson, G. S., A. F. Hill, C. Joseph, L. Hosszu, A. Power, J. P. Waltho, A. R. Clarke and J. Collinge (1999). "Multiple folding pathways for heterologously expressed human prion protein." Biochim Biophys Acta 1431(1): 1-13.
- Jackson, M. and H. H. Mantsch (1995). "The use and the Misuse of FTIR Spectroscopy in the Determination of the Protein Structure." Crit. Rev. Biochem. Mol. Biol. 30: 95-120.
- Jarrett, J., T., and P. T. J. Lansbury (1983). "Seeding "one-dimensional crystallization" of amyloid: A pathogenic mechanism in Alzheimer's disease and scrapie?" Cell 73: 1055-1058.
- Jeffrey, M., I. A. Goodbrand and C. M. Goodsir (1995). "Pathology of the transmissible spongiform encephalopathies with special emphasis on ultrastructure." Micron 26: 277-298.
- Jeffrey, M., C. Goodsir, M. Bruce, P. McBride, J. Scott and W. Halliday (1992a). "Infection specific prion protein (PrP) accumulates on neuronal plasmalemma in scrapie infected mice." Neurosci. Lett. 23: 106-109.
- Jeffrey, M., C. M. Goodsir, M. E. Bruce, N. Fowler and J. R. Scott (1994). "Murine scrapie-infected neurons in vivo release excess prion protein into the extracellular space." Neurosci. Lett. 174: 39-42.
- Jeffrey, M., J. R. Scott, A. Williams and H. Fraser (1992b). "Ultrastructural features of spongiform encephalopathy transmitted to mice from three species of bovidae." Acta neuropathologica 84: 559-569.
- Jimenez, J. L., E. J. Nettleton, M. Bouchard, C. V. Robinson, C. M. Dobson and H. R. Saibil (2002). "The protofilament structure of insulin amyloid fibrils." Proc. Natl Acad. Sci. U.S.A. 99(14): 9196-9201.
- Jones, E. M. and W. K. Surewicz (2005). "Fibril Conformation as the Basis of Species- and Strain-Dependent Seeding Specificity of Mammalian Prion Amyloids." Cell 121(1): 63.
- Kasczak, R. J., R. Rubenstein, P. A. Merz, M. Tonna-DeMasi, R. Fersko, R. I. Carp, H. M. Wisniewski and H. Diring (1987). "Mouse polyclonal and monoclonal antibody to scrapie-associated fibril proteins." J. Virol. 61: 3688-3693.
- Kellings, K., N. Meyer, C. miranda, S. B. Prusiner and D. Reisner (1992). "Further analysis of nucleic acids in purified scrapie prion preparations by improved return refocusing gel electrophoresis." J. Gen. Virol. 73: 1025-1029.
- Khurana, R. and A. L. Fink (2000). "Do Parallel beta -Helix Proteins Have a Unique Fourier Transform Infrared Spectrum?" Biophys. J. 78(2): 994-1000.

References

- Khurana, R., C. Ionescu-Zanetti, M. Pope, J. Li, L. Nielson, M. Ramirez-Alvarado, L. Regan, A. L. Fink and S. A. Carter (2003). "A General Model for Amyloid Fibril Assembly Based on Morphological Studies Using Atomic Force Microscopy." Biophys. J. 85(2): 1135-1144.
- Kim, J. and E. Manuelidis (1989). "Neuronal alterations in experimental Creutzfeldt-Jakob disease: a Golgi study." J. Neurol. Sci. 89: 93-101.
- Kimberlin, R. and R. Marsh (1975). "Comparison of scrapie and transmissible mink encephalopathy in hamsters. I. Biochemical studies of brain during development of disease." J Infect Dis. 131: 97-103.
- Kimberlin, R. and C. Walker (1978). "Evidence that the transmission of one source of scrapie agent to hamsters involves separation of agent strains from a mixture." J. Gen. Virol. 39: 487-496.
- Kimberlin, R., C. Walker and H. Fraser (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.
- Kimberlin, R. H. and C. A. Walker (1977). "Characteristic of Short Incubation Model of Scrapie in Golden Hamster." J. Gen. Vrol. 34: 295-304.
- Kingsbury, D., K. Kasper, D. Stites, J. Watson, R. Hogan and S. Prusiner (1983). "Genetic control of scrapie and Creutzfeldt-Jakob disease in mice." J Immunol 131(1): 491-496.
- Kitamoto, T., J. Tateishi, T. Tashima, I. Takeshita, R. Barry, S. DeArmond and P. SB (1986). "Amyloid plaques in Creutzfeldt-Jakob disease stain with prion protein antibodies." Ann. Neurol. 20: 204-208.
- Klein, T. R., D. Kirsch, R. Kaufmann and D. Reisner (1998). "Prion rods contain small amounts of two host sphingolipids as revealed by thin-layer chromatography and mass spectrometry." Biol. Chem. 379: 655-666.
- Kneipp, J., M. Beekes, P. Lasch and D. Naumann (2002). "Molecular changes of preclinical scrapie can be detected by infrared spectroscopy." J. Neurosci. 22(8): 2989-2997.
- Kneipp, J., P. Lasch, E. Baldauf, M. Beekes and D. Naumann (2000). "Detection of pathological molecular alterations in scrapie-infected hamster brain by Fourier transform infrared (FT-IR) spectroscopy." Biochim. Biophys. Acta 1501: 189-199.
- Kneipp, J., L. Miller, M., S. Spassov, F. Sokolowski, P. Lasch, M. Beekes and D. Naumann (2004a). "Prion structure investigated in situ, ex vivo, and in vitro by FTIR spectroscopy." Proc. SPIE 5321: 17-25.
- Kneipp, J., L. Miller, S. Spassov, F. Sokolowski, P. Lasch, M. Beekes and D. Naumann (2004b). "Scrapie-infected cells, isolated prions, and recombinant prion protein: a comparative study." Biopolymers 74: 163-167.
- Korth, C., P. Streit and B. Oesch (1999). "Monoclonal antibodies specific for the native, disease-associated isoform of the prion protein." Methods Enzymol 309: 106-22.
- Krimm, S. and Y. Abe (1972). "Intermolecular interaction effects in the amide I vibrations of polypeptides." Proc. Natl. Acad. Sci. U.S.A 69: 2788-92.
- Kubelka, J. and T. A. Keiderling (2001). "Differentiation of beta-Sheet structures: Ab Initio-Based Simulations of IR Absorptions and Vibrational CD for Model Peptide and Protein beta-Sheets." J. Amer. Chem. Soc. 123: 12048-12058.
- Kuczus, T. and M. Groschup (1999). "Differences in proteinase K resistance and neuronal deposition of abnormal prion proteins characterize bovine spongiform encephalopathy (BSE) and scrapie strains." Mol. Med. 5: 406-418.
- Kühbacher, M., G. Weseloh, A. Thomzig, H. Bertelsmann, G. Falkenberg, M. Radtke, H. Riesemeier, A. Kyriakopoulos, M. Beekes and D. Behne (2005). "Analysis and localization of metal- and metalloid-containing proteins by synchrotron radiation x-ray fluorescence spectrometry." X-Ray Spectrometry 34(2): 112-117.

- Laemmli, U. K. (1970). "Cleavage of Structural Proteins during the Assamby of the Head of Bacteriophage T4." Nature 227: 680-684.
- Landis, D., R. Williams and C. Masters (1981). "Golgi and electronmicroscopic studies of spongiform encephalopathy." Neurology 31: 538-49.
- Lasmezas, C. I. (2003). "Putative functions of PrPC." Br. Med. Bull. 66(1): 61-70.
- Legname, G., I. V. Baskakov, H.-O. B. Nguyen, D. Riesner, F. E. Cohen, S. J. DeArmond and S. B. Prusiner (2004). "Synthetic Mammalian Prions." Science 305(5684): 673-676.
- Legname, G., H.-O. B. Nguyen, I. V. Baskakov, F. E. Cohen, S. J. DeArmond and S. B. Prusiner (2005). "Strain-specified characteristics of mouse synthetic prions." Proc. Natl Acad. Sci. U.S.A.: 0409079102.
- Li, L., T. A. Darden, L. Bartolotti, D. Kominos and L. G. Pedersen (1999). "An Atomic Model for the Pleated beta -Sheet Structure of Abeta Amyloid Protofilaments." Biophys. J. 76(6): 2871-2878.
- Liberski, P. P., D. M. Asher, R. Yanagihara, J. C. J. Gibbs and D. C. Gajdusek (1989). "Serial Ultrastructural Studies of Scrapie In Hamsters." J. Comp. Path. 101: 429-356.
- Liberski, P. P., B. Sikorska, J. Bratosiewicz-Wasik, D. Carleton Gajdusek and P. Brown (2004). "Neuronal cell death in transmissible spongiform encephalopathies (prion diseases) revisited: from apoptosis to autophagy." Int. J. Biochem. Cell Biol. 36(12): 2473-2490.
- Liu, H., S. Farr-Jones, N. B. Ulyanov, M. Llinas, S. Marqusee, D. Groth, F. E. Cohen, S. B. Prusiner and T. L. James (1999). "Solution Structure Of Syrian Hamster Prion Protein rPrP(90-231)." Biochemistry 38: 5362-5377.
- Liu, T., T. Zwingman, R. Li, T. Pan, B.-S. Wong, R. B. Petersen, P. Gambetti, K. Herrup and M.-S. Sy (2001). "Differential expression of cellular prion protein in mouse brain as detected with multiple anti-PrP monoclonal antibodies." Brain Res. 896(1-2): 118-129.
- Lowenstein, D., D. Butler, D. Westaway, M. McKinley, S. DeArmond and P. SB. (1990). "Three hamster species with different scrapie incubation times and neuropathological features encode distinct prion proteins." Mol. Cell. Biol. 10: 1153-1163.
- Lugaresi, E., R. Medori, P. Montagna, A. Baruzzi, P. Cortelli and A. Lugaresi (1986). "Fatal familial insomnia and dysautonomia with selective degeneration of thalamic nuclei." New English Journal of Medicine 315: 997-1004.
- Madec, J. Y., A. Vanier, A. Dorier, J. Bernillon, P. Belli and Th. Baron (1997). "Biochemical properties of protease resistant prion protein PrPsc in natural sheep scrapie." Arch Virol 142: 1603-1612.
- Malone, T. G., R. P. Marsh, R. P. Hanson and J. S. Semancik (1979). Hamstert scrapie agent: properties, partial purification, and a DNA component. Slow transmissible diseases of the nervous system. S. B. Prusiner and W. J. Hadlow. New York London Sydney Toronto San Francisco, Academic Press. 2: 479-.
- Manuelidis, L. (2003). "Transmissible encephalopathies: speculations and realities." Viral Immunol. 16: 123-39.
- Manuelidis, L., T. Sklaviadis, A. Akowitz and W. Fritch (1995). "Viral Particles are Required for Infection in Neurodegenerative Creutzfeldt-Jakob Disease." Proc. Natl Acad. Sci. U.S.A. 92(11): 5124-5128.
- Marsh, R. (1992). Transmissible mink encephalopathy. London, Ellis Horwood.
- Marsh, R. and R. Kimberlin (1975). "Comparison of scrapie and transmissible mink encephalopathy in hamsters. II. Clinical signs, pathology, and pathogenesis." J Infect Dis 131: 104-110.
- Marsh, R. F. (1993). "Bovine spongiform encephalopathy: a new disease of cattle?" Arch Virol Suppl 7: 255-9.

References

- Masel, J. and V. A. A. Jansen (1999). "The kinetics of proteinase K digestion of linear prion polymers." Proc. R. Soc. Lond. 266: 1927-1931.
- Masel, J. and V. A. A. Jansen (2001). "The measured level of prion infectivity varies in a predictable way according to the aggregation state of the infectious agent." Biochim. Biophys. Acta 1535(2): 164.
- Masters, C. L., D. C. Gajdusek and C. J. Gibbs, Jr (1981). "Creutzfeldt-Jakob disease virus isolations from the Gerstmann-Straussler syndrome with an analysis of the various forms of amyloid plaque deposition in the virus-induced spongiform encephalopathies." Brain 104(3): 559-588.
- McBride, P. A. and M. Beekes (1999). "Pathological PrP is abundant in sympathetic and sensory ganglia of hamsters fed with scrapie." Neurosci. Lett. 265: 135-138.
- McKinley, M., A. Taraboulos, L. Kenaga, D. Serban, A. Stieber, S. DeArmond, S. Prusiner and N. Gonatas (1991a). "Ultrastructural localization of scrapie prion proteins in cytoplasmic vesicles of infected cultured cells." Lab. Invest. 65: 622-630.
- McKinley, M. P., M. B. Braunfeld, C. G. Bellinger and S. B. Prusiner (1986). "Molecular Characteristics of Prion Rods Purified from Scrapie-Infected Hamster Brains." J Infect Dis 154: 110-120.
- McKinley, M. P., R. K. Meyer, L. Kenaga, F. Rahbar, R. Cotter, A. Serban and S. B. Prusiner (1991b). "Scrapie Prion Rod Formation In Vitro Requires both Detergent Extraction and Limited Proteolysis." J. Virol. 65: 1340-1351.
- Merz, P. A., R. A. Somerville, H. M. Wisniewski and K. Iqbal (1981). "Abnormal fibrils from scrapie-infected brain." Acta Neuropathologica 54(1): 63-74.
- Milhavet, O., H. E. M. McMahon, W. Rachidi, N. Nishida, S. Katamine, A. Mange, M. Arlotto, D. Casanova, J. Riandel, A. Favier and S. Lehmann (2000). "Prion infection impairs the cellular response to oxidative stress." Proc. Natl Acad. Sci. U.S.A. 97(25): 13937-13942.
- Millson, G. C. and J. Manning (1979). The effect of selected detergents on scrapie infectivity. Slow transmissible diseases of the nervous system. S. B. Prusiner and W. J. Hadlow. New York London Sydney Toronto San Francisco, Academic Press. 2: 409-463.
- Millson, G. D., Hunter and R. H. Kimberlin (1976). "Slow Virus Disease of Animals and Man." Front Biol. 44: 241-266.
- Minor, P., J. Newham, N. Jones, C. Bergeron, L. Gregori, D. Asher, F. van Engelenburg, T. Stroebel, M. Vey, G. Barnard, M. Head and the WHO Working Group on International Reference Materials for the Diagnosis and Study of Transmissible Spongiform Encephalopathies (2004). "Standards for the assay of Creutzfeldt-Jakob disease specimens." J Gen Virol 85(6): 1777-1784.
- Mishra, R. S., S. Basu, Y. Gu, X. Luo, W.-Q. Zou, R. Mishra, R. Li, S. G. Chen, P. Gambetti, H. Fujioka and N. Singh (2004). "Protease-Resistant Human Prion Protein and Ferritin Are Cotransported across Caco-2 Epithelial Cells: Implications for Species Barrier in Prion Uptake from the Intestine." J. Neurosci. 24(50): 11280-11290.
- Nakamura, K., T. Tanaka, A. Kuwahara and K. Takeo (1985). "Microassay for Proteins on Nitrocellulose Filter Using Protein Day-Staining Procedure." Anal. Biochem. 148: 311-319.
- Narang, H. (2002). "A critical review of the nature of the spongiform encephalopathy agent: protein theory versus virus theory." Exp. Biol. Med. 227(1): 4-19.
- Nelson, R., M. R. Sawaya, M. Balbirnie, A. O. Madsen, C. Riek, R. Grothe and D. Eisenberg (2005). "Structure of the cross-[beta] spine of amyloid-like fibrils." Nature 435(7043): 773.
- Neuhoff, V., K. Philipp, H. G. Zimmer and S. Mesecke (1979). "A Simple, Versative, Sensitive and Volume-Independent Method for Quantitative Protein Determination

- wich is Independent of Other External Influences." Hoppe-Seyler's Z. Physiol. Chem. 360: 1657-1670.
- Nguyen, J. T., H. Inouye, M. A. Baldwin, R. J. Fletterick, F. E. Cohen, S. B. Prusiner and D. A. Kirschner (1995). "X-ray Diffraction of Scrapie Prion Rods and PrP Peptides." J. Mol. Biol. 252(4): 412-422.
- Notari, S., S. Capellari, A. Giese, I. Westner, A. Baruzzi, B. Ghetti, P. Gambetti, H. A. Kretzschmar and P. Parchi (2004). "Effects of Different Experimental Conditions on the PrP^{Sc} Core Generated by Protease Digestion: IMPLICATIONS FOR STRAIN TYPING AND MOLECULAR CLASSIFICATION OF CJD." J. Biol. Chem. 279(16): 16797-16804.
- Oakley, B. R., D. R. Kirsch and N. R. Moriss (1980). "A Simplified Ultrasensitive Silver Stain for Detecting Proteins in Polyacrylamide Gels." Anal. Biochem. 105: 361-363.
- Oesch, B., M. Jensen, P. Nilsson and J. Fogh (1994). "Properties of the scrapie prion protein: quantitative analysis of protease resistance." Biochemistry 33: 5926-5931.
- Outram, G. (1976). "The pathogenesis of scrapie in mice." Front Biol. 44: 325-57.
- Pan, K.-M., M. Baldwin, J. Nguyen, M. Gasset, A. Serban, D. Groth, I. Mehlhorn, Z. Huang, R. J. Fletterick, F. E. Cohen and S. B. Prusiner (1993). "Conversion of alpha-helices into beta-sheets features in the formation of the scrapie prion proteins." Proc. Natl. Acad. Sci. U.S.A. 90: 10962-10966.
- Pan, T., M. Colucci, B.-S. Wong, R. Li, T. Liu, R. B. Petersen, S. Chen, P. Gambetti and M.-S. Sy (2001). "Novel Differences between Two Human Prion Strains Revealed by Two-dimensional Gel Electrophoresis." J. Biol. Chem. 276(40): 37284-37288.
- Pan, T., R. Li, B.-S. Wong, T. Liu, P. Gambetti and M.-C. Sy (2002). "Heterogeneity of normal prion protein in two-dimensional immunoblot: presence of various glycosylated and truncated forms." J. Neurochem. 81: 1092-1101.
- Parchi, P., S. Capellari, S. G. Chen, R. B. Petersen and P. Gambetti (1997). "Typing prion isoforms." Nature 386: 232-233.
- Parchi, P., R. Castellani, S. Capellari, B. Ghetti, K. Young, S. G. Chen, M. Farlow, D. W. Dickson, A. A. Sima and A. Trojanowski et (1996). "Molecular basis of phenotypic variability in sporadic Creutzfeldt-Jakob disease." Ann. Neurol. 39(6): 767-778.
- Parchi, P., A. Giese, S. Capellari, Brown P, Schulz-Schaeffer W, Windl O, Zerr I, Budka H, Kopp N, Piccardo P, Poser S, Rojiani A, Streichemberger N, Julien J, Vital C, Ghetti B, Gambetti P and K. H. (1999). "Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects." Ann. Neurol. 46: 224-33.
- Parchi, P., W. Zou, W. Wang, P. Brown, S. Capellari, B. Ghetti, N. Kopp, W. J. Schulz-Schaeffer, H. A. Kretzschmar, M. W. Head, J. W. Ironside, P. Gambetti and S. G. Chen (2000). "Genetic influence on the structural variations of the abnormal prion protein." Proc. Natl Acad. Sci. U.S.A. 97(18): 10168-10172.
- Pattison, I. and G. Millson (1961). "Scrapie produced experimentally in goats with special reference to the clinical syndrome." J. Comp. Pathol. 71: 107-109.
- Pattison, I. H. (1966). "The relative susceptibility of sheep, goats and mice to two types of the goat scrapie agent." Res. Vet. Sci. 7: 207-12.
- Pattison, J. (1998). "The Emergence of Bovine Spongiform Encephalopathy and Related Diseases." Emerging Infect. Dis. 4: 390-394.
- Peretz, D., M. R. Scott, D. Groth, R. A. Williamson, D. R. Burton, F. E. Cohen and S. B. Prusiner (2001). "Strain-specified relative conformational stability of the scrapie prion protein." Protein Sci 10(4): 854-863.

References

- Peretz, D., R. A. Williamson, Y. Matsunaga, H. Serban, C. Pinilla, R. B. Bastidas, R. Rozenshteyn, T. L. James, R. A. Houghten and F. E. Cohen (1997). "A conformational transition at the N terminus of the prion protein features in formation of the scrapie isoform." J. Mol. Biol. 273(3): 614-622.
- Peri, S. e. a. (2003). "Development of human protein reference database as an initial platform for approaching systems biology in humans." Genome Res. 13: 2363-2371.
- Polymenidou, M., K. Stoeck, M. Glatzel, M. Vey, A. Bellon and A. Aguzzi (2005). "Coexistence of multiple PrP(Sc) types in individuals with Creutzfeldt-Jakob disease." Lancet Neurol 4(12): 805-14.
- Poser, C. M. (2002a). "Notes on the history of the prion diseases. Part I." Clin Neurol Neurosurg 104(1): 1-9.
- Poser, C. M. (2002b). "Notes on the history of the prion diseases. Part II." Clin Neurol Neurosurg 104(2): 77-86.
- Priola, S. A. (2001). "Prion Protein Diversity and Disease in the Transmissible Spongiform Encephalopathies." Adv. Protein. Chem. 57: 1-27.
- Priola, S. A., B. Chesebro and B. Caughey (2003). "BIOMEDICINE: A View from the Top--Prion Diseases from 10,000 Feet." Science 300(5621): 917-919.
- Prusiner, S., M. McKinley, Groth DF, Bowman KA, Mock NI, Cochran SP and M. FR (1981). "Scrapie agent contains a hydrophobic protein." Proc. Natl. Acad. Sci. U.S.A. 78: 6675-6679.
- Prusiner, S. B. (1982). "Novel proteinaceous infectious particles cause scrapie." Science 246: 136-144.
- Prusiner, S. B. (1991). "Molecular Biology of Prion Diseases." Science 252: 1515-1522.
- Prusiner, S. B. (1996a). "Molecular Biology and Genetics of Prion Diseases." Cold Spring Harb. Symp. Quant. Biol. 61: 473-493.
- Prusiner, S. B. (1996b). Prions Prions Prions, Springer-Verlag Berlin Heidelberg New York.
- Prusiner, S. B. (1997). "Prion Diseases and the BSE Crisis." Science 278(5336): 245-251.
- Prusiner, S. B. (1999). Development of the prion concept. Cold Spring Harbor, N.Y., Cold Spring Harbor Press.
- Prusiner, S. B., W. J. Hadlow, C. M. Eklund and R. E. Race (1977). "Sedimentation properties of the scrapie agent." Proc. Natl. Acad. Sci. U.S.A 74: 4656-4660.
- Prusiner, S. B., W. J. Hadlow, D. E. Garfin, S. P. Cochran, J. R. Baringer, R. E. Race and C. M. Eklund (1987). "Partial purification and evidence for multiple molecular forms of the scrapie agent." Biochemistry 17: 4993-9.
- Prusiner, S. B., M. P. McKinley, K. A. Bowman, D. C. Bolton, P. E. Bendheim, D. F. Groth and G. G. Glenner (1983). "Scrapie Prions Aggregate to Form Amyloid-like Birefringent Rods." Cell 35: 349-358.
- Prusiner, S. B., M. R. Scott, S. J. DeArmond and F. E. Cohen (1998). "Prion Protein Biology." Cell 93: 337-348.
- Puoti, G., G. Giaccone, G. Rossi, B. Canciani, O. Bugiani and F. Tagliavini (1999). "Sporadic Creutzfeldt-Jakob disease: Co-occurrence of different types of PrPSc in the same brain." Neurology 53(9): 2173-.
- Qin, K., Y. Yang, P. Mastrangelo and D. Westaway (2002). "Mapping Cu(II) Binding Sites in Prion Proteins by Diethyl Pyrocarbonate Modification and Matrix-assisted Laser Desorption Ionization-Time of Flight (MALDI-TOF) Mass Spectrometric Footprinting." J. Biol. Chem. 277: 1981-1990.
- Quaglio, E., R. Chiesa and D. A. Harris (2001). "Copper Converts the Cellular Prion Protein into a Protease-resistant Species That Is Distinct from the Scrapie Isoform." J. Biol. Chem. 276(14): 11432-11438.

- Rabilloud, T. (1990). "Mechanisms of protein silver staining in polyacrylamide gels: a 10-year synthesis." Electrophoresis 11(10): 785-94.
- Race, R., K. Graham, D. Ernst, B. Caughey and B. Chesebro (1990). "Analysis of linkage between scrapie incubation period and the prion protein gene in mice." J Gen Virol 71(2): 493-497.
- Reinstädler, D., H. Fabian, J. Beckmann and D. Naumann (1996). "Refolding of thermally and urea-denatured ribonuclease A monitored by time-resolved FTIR spectroscopy." Biochemistry 35: 15822-15830.
- Rezaie, P. and P. L. Lantos (2001). "Microglia and the pathogenesis of spongiform encephalopathies." Brain Res. Rev. 35(1): 55-72.
- Riek, R., S. Hornemann, G. Wider, R. Glockshuber and K. Wuthrich (1997). "NMR characterization of the full-length recombinant murine prion protein, mPrP(23-231)." FEBS Lett. 413(2): 282-288.
- Riesner, D. (2003). "Biochemistry and structure of PrPC and PrPSc." Br. Med. Bull. 66(1): 21-33.
- Roberts, G., R. Lofthouse, D. Allsop, M. Landon, M. Kidd, S. Prusiner and C. TJ. (1988). "CNS amyloid proteins in neurodegenerative diseases." Neurology 38: 1534-1540.
- Rogers, M., D. Serban, T. Gyuris, M. Scott, T. Torchia and S. B. Prusiner (1991). "Epitope mapping of the Syrian hamster prion protein utilizing chimeric and mutant genes in a vaccinia virus expression system." J. Immunol. 147(10): 3568-3574.
- Rohwer, R. G., P. W. Brown and D. C. Gajdusek (1979). The use of sedimentation to equilibrium as a step in the purification of the scrapie agent. Slow transmissible diseases of the nervous system. S. B. Prusiner and W. J. Hadlow. New York London Sydney Toronto San Francisco, Academic Press. 2: 465-478.
- Rubenstein, R., P. C. Gray, C. M. Wehlburg, J. S. Wagner and G. C. Tisone (1998). "Detection and Discrimination of PrP-Sc by Multi-spectral Ultraviolet Fluorescence." Biochemical and Biophysical Research Communication 246: 100-106.
- Rudd, P. M., T. Endo, C. Colominas, D. Groth, S. F. Wheeler, D. J. Harvey, M. R. Wormald, H. Serban, S. B. Prusiner, A. Kobata and R. A. Dwek (1999). "Glycosylation differences between the normal and pathogenic prion protein isoforms." Proc. Natl Acad. Sci. U.S.A. 96(23): 13044-13049.
- Saborio, G. P., B. Permanne and C. Soto (2001). "Sensitive detection of pathological amplification of protein misfolding." Nature 411: 810-813.
- Safar, J., P. P. Roller, D. C. Gajdusek and J. Clarence J. Gibbs (1993a). "Thermal stability and conformational transitions of scrapie amyloid (prion) protein correlate with infectivity." Protein Sci. 2: 2206-2216.
- Safar, J., P. P. Roller, D. G. Gajdusek and J. Clarence J. Gibbs (1993b). "Conformational Transitions, Dissociation, and Unfolding of Scrapie Amyloid (Prion) Protein." J. Biol. Chem. 268: 20276-20284.
- Safar, J., H. Wille, V. Itri, D. Groth, N. Serban, M. Torchia, F. E. Cohen and S. B. Prusiner (1998). "Eight prion strains have PrP-Sc molecules with different conformations." Nat. Med. 4: 1157-1165.
- Safar, J. G., M. D. Geschwind, C. Deering, S. Didorenko, M. Sattavat, H. Sanchez, A. Serban, M. Vey, H. Baron, K. Giles, B. L. Miller, S. J. DeArmond and S. B. Prusiner (2005a). "Diagnosis of human prion disease." Proc. Natl Acad. Sci. U.S.A. 102(9): 3501-3506.
- Safar, J. G., K. Kellings, A. Serban, D. Groth, J. E. Cleaver, S. B. Prusiner and D. Riesner (2005b). "Search for a prion-specific nucleic Acid." J. Virol. 79(16): 10796-806.
- Schreuder, B. E. (1993). "General aspects of transmissible spongiform encephalopathies and hypotheses about the agents." Vet. Q. 15: 167-174.

References

- Schulz-Schaeffer, W. J., S. Tschöke, N. Kranefuss, W. Dröse, D. Hause-Reitner, A. Giese, M. H. Groschup and H. A. Kretzschmar (2000). "The Paraffin-Embedded Tissue Blot Detects PrPSc Early in the Incubation Time in Prion Diseases." Am. J. Pathol. 156: 51-56.
- Shaper, N. L., J. A. Meurer, D. H. Joziase, T. D. D. Chou, E. J. Smith, R. L. Schnaar and J. H. Shaper (1997). "The Chicken Genome Contains Two Functional Nonallelic beta 1,4-Galactosyltransferase Genes. CHROMOSOMAL ASSIGNMENT TO SYNTENIC REGIONS TRACKS FATE OF THE TWO GENE LINEAGES IN THE HUMAN GENOME." J. Biol. Chem. 272(50): 31389-31399.
- Sigurdson, C. J. and M. W. Miller (2003). "Other animal prion diseases." Br. Med. Bull. 66(1): 199-212.
- Silveira, J. R., B. Caughey and G. S. Baron (2004). "Prion protein and the Molecular Features of Transmissible Spongiform Encephalopathy Agents." CTMI 284: 1-50.
- Silveira, J. R., G. J. Raymond, A. G. Hughson, R. E. Race, V. L. Sim, S. F. Hayes and B. Caughey (2005). "The most infectious prion protein particles." Nature 437(7056): 257.
- Sokolowski, F., A. J. Modler, R. Masuch, D. Zirwer, M. Baier, G. Lutsch, D. A. Moss, K. Gast and D. Naumann (2003). "Formation of Critical Oligomers Is a Key Event during Conformational Transition of Recombinant Syrian Hamster Prion Protein." J. Biol. Chem. 278(42): 40481-40492.
- Sokolowski, F. and D. Naumann (2005). "FTIR study on thermal denaturation and aggregation of recombinant hamster prion protein SHaPrP90-232." Vibr. Spectr. 38: 39-44.
- Somerville, R., L. Ritchie and P. Gibson (1989). "Structural and biochemical evidence that scrapie-associated fibrils assemble in vivo." J Gen Virol 70: 25-35.
- Somerville, R. A. (2002). "TSE agent strains and PrP: reconciling structure and function." Trends Biochem. Sci. 27: 606-611.
- Somerville, R. A., A. Chong and O. U. Mulqueen (1997). "Biochemical typing of scrapie strains." Nature 386: 564.
- Somerville, R. A., R. C. Oberthur, U. Havekost, F. MacDonald, D. M. Taylor and A. G. Dickinson (2002). "Characterization of Thermodynamic Diversity between Transmissible Spongiform Encephalopathy Agent Strains and Its Theoretical Implications." J. Biol. Chem. 277(13): 11084-11089.
- Stahl, N., M. A. Baldwin, D. B. Teplow, L. Hood, B. W. Gibson, A. L. Burlingame and S. B. Prusiner (1993). "Structural Studies of Scrapie Prion Protein Using Mass Spectrometry and Amino Acid Sequencing." Biochemistry 32: 1991-2002.
- Stuard, B. (1997). Biological Applications of Infrared Spectroscopy, John Wiley & Sons, Ltd, Chichester.
- Sunde, M. and C. Balke (1997a). "The structure of amyloid fibrils by electron microscopy and X-ray diffraction." Adv. Protein. Chem. 50: 123-159.
- Sunde, M., L. C. Serpell, M. Bartlam, P. E. Fraser, M. B. Pepys and C. C. F. Blake (1997b). "Common core structure of amyloid fibrils by synchrotron X-ray diffraction." J. Mol. Biol. 273(3): 729-739.
- Surewicz, W. K., H. H. Mantsch and D. Chapman (1993). "Determination of protein secondary structure by Fourier transform infrared spectroscopy: a critical assessment." Biochemistry 32: 389-394.
- Susi, H. and M. Byler (1983). "Protein Structure by Fourier Transform Infrared Spectroscopy: second derivative spectra." Biochem. Biophys. Res. Commun. 115: 391-397.
- Tamm, L. K. and S. A. Tatulian (1997). "Infrared spectroscopy of proteins and peptides in lipid bilayers." Q. Rev. Biophys. 30: 365-429.

- Taraboulos, A., D. Serban and S. Prusiner (1990). "Scrapie prion proteins accumulate in the cytoplasm of persistently infected cultured cells." J. Cell Biol. 110: 2117-2132.
- Taylor, D. M., K. Fernie, I. McConnell and P. J. Steele (1998). "Observations on thermostable subpopulations of the unconventional agents that cause transmissible degenerative encephalopathies." Veterinary Microbiology 64(1): 33-38.
- Thomzig, A., C. Kratzel, G. Lenz, D. Kruger and M. Beekes (2003). "Widespread PrPSc accumulation in muscles of hamsters orally infected with scrapie." EMBO Rep. 4: 530-533.
- Thomzig, A., S. Spassov, M. Friedrich, D. Naumann and M. Beekes (2004). "Discriminating scrapie and BSE isolates by infrared spectroscopy of pathological prion protein." J. Biol. Chem. 279: 33847-33854.
- Tjernberg, L. O., D. J. E. Callaway, A. Tjernberg, S. Hahne, C. Lilliehook, L. Terenius, J. Thyberg and C. Nordstedt (1999). "A Molecular Model of Alzheimer Amyloid beta - Peptide Fibril Formation." J. Biol. Chem. 274(18): 12619-12625.
- Tovey, E. R. and B. A. Baldo (1987). "Comparison by electroblotting of IgE-binding components in extracts of house dust mite bodies and spent mite culture." J. Allergy Clin. Immunol. 79(1): 93-102.
- Turk, E., D. Teplow, L. Hood and S. Prusiner (1988). "Purification and properties of the cellular and scrapie hamster prion proteins." Eur. J. Biochem. 176: 21-30.
- Tycko, R. (2004). "Progress towards a molecular-level structural understanding of amyloid fibrils." Curr. Opin. Struct. Biol. 14(1): 96-103.
- Unterberger, U., T. Voigtlander and H. Budka (2005). "Pathogenesis of prion diseases." Acta Neuropathol.
- Wadsworth, J., A. Hill, S. Joiner, G. Jackson, A. Clarke and Collinge J. (1999). "Strain-specific prion-protein conformation determined by metal ions." Nat Cell Biol. 1: 55-59.
- Wadsworth, J. D., A. F. Hill, J. A. Beck and J. Collinge (2003). "Molecular and clinical classification of human prion disease." Br. Med. Bull. 66(1): 241-254.
- Wang, Q., A. Kretlow, M. Beekes, D. Naumann and L. Miller (2005). "In situ characterization of prion protein structure and metal accumulation in scrapie-infected cells by synchrotron infrared and X-ray imaging." Vibrational Spectroscopy 38(1-2): 61.
- Wechselberger, C., S. Wurm, W. Pfarr and O. Hoglinger (2002). "The Physiological Functions of Prion Protein." Exp. Cell Res. 281(1): 1-8.
- Weissmann, C. (1990). "A "unified theory" of prion propagation." Nature 352: 679 - 683.
- Weissmann, C. (2002). "Molecular genetics of transmissible spongiform encephalopathies: an introduction." J Toxicol Sci 27: 69-77.
- Weissmann, C. (2004). "THE STATE OF THE PRION." Nat. Rev. Micro. 2(11): 861-871.
- Wells, G. A. H., A. C. Scott, C. T. Johnson, R. F. Gunning, R. D. Hancock, M. Jeffrey, M. Dawson and R. Bradley (1987). "A novel progressive spongiform encephalopathy in cattle." The Veterinari Record 121: 419-420.
- Westaway, D., S. J. DeArmond, J. Cayetano-Canlas, D. Groth, d. Fostwer, S.-L. Yang, M. Torchia, G. A. Carlson and S. B. Prusiner (1994). "Degeneration of skeletal muscle, peripheral nerves, and the central nervous system in transgenic mice overexpressing wild-type prion proteins." Cell 79: 117-129.
- Wickner, R. B. (2005). "Scrapie in Ancient China?" Science 309(5736): 874b-.
- Wilesmith, J. W., G. A. Wells, M. P. Cranwell and J. B. Ryan (1988). "Bovine spongiform encephalopathy: epidemiological studies." Vet. Rec. 17: 638-644.
- Will, R., J. Ironside, Zeidler M., Cousens SN., Estibeiro K., Alperovitch A., Poser S., Pocchiari M., Hofman A. and S. PG. (1996). "A new variant of Creutzfeldt-Jakob disease in the UK." Lancet 347: 921-925.

References

- Will, R. G. (2003). "Acquired prion disease: iatrogenic CJD, variant CJD, kuru." Br. Med. Bull. 66(1): 255-265.
- Wille, H., M. D. Michelitsch, V. Guenebaut, S. Supattapone, A. Serban, F. E. Cohen, D. A. Agard and S. B. Prusiner (2002). "Structural studies of the scrapie prion protein by electron crystallography." Proc. Natl Acad. Sci. U.S.A. 99(6): 3563-3568.
- Williams, A., P. J. Lucassen, D. Ritchie and M. Bruce (1997). "PrP Deposition, Microglial Activation, and Neuronal Apoptosis in Murine Scrapie." Exp. Neurol. 144: 433-438.
- Williams, E. and S. Young (1980). "Chronic wasting disease of captive mule deer: a spongiform encephalopathy." J Wildl Dis 16: 89-98.
- Wood, J. L., L. J. Lund and S. H. Done (1992). "The natural occurrence of scrapie in moufflon." Vet. Rec. 130(2): 25-27.
- Zanusso, G., A. Farinazzo, F. Prelli, M. Fiorini, M. Gelati, S. Ferrari, P. G. Righetti, N. Rizzuto, B. Frangione and S. Monaco (2004). "Identification of Distinct N-terminal Truncated Forms of Prion Protein in Different Creutzfeldt-Jakob Disease Subtypes." J. Biol. Chem. 279(37): 38936-38942.
- Zanusso, G., P. G. Righetti, Ferrari S, Terrin L, Farinazzo A, Cardone F, Pocchiari M, Rizzuto N and M. S. (2002). "Two-dimensional mapping of three phenotype-associated isoforms of the prion protein in sporadic Creutzfeldt-Jakob disease." Electrophoresis 23: 347-355.
- Zou, W.-Q. and P. Gambetti (2005). "From Microbes to Prions: The Final Proof of the Prion Hypothesis." Cell 121(2): 155.