

# **PRION STRAINS: ORIGINS, MECHANISMS AND IMPLICATIONS FOR DISEASE**

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The Banbury Center, Cold Spring Harbor Laboratory, 11-14 May 2008

Organized By: B. Caughey, NIAID Rocky Mountain Laboratories,  
C. Weissmann, Scripps Florida,  
J. Collinge, University College London,  
C. Soto, University of Texas Medical Branch

## **PROGRAM**

### **Sunday, 11 May**

Afternoon Arrival at Robertson House, Banbury Center, for registration and room assignment

6:00 pm Reception at Robertson House

7:30 pm Dinner at Robertson House

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### **Monday, 12 May**

7:15-8:15 am Breakfast at Robertson House

8:30-8:35 am J. A. Witkowski, Banbury Center, Cold Spring Harbor Laboratory, Cold Spring Harbor, New York:  
Introductory remarks.

8:35-9:30 am **SESSION 1: Strains - Definition, Concepts**  
Chair: J. Collinge, University College London, United Kingdom

8:35-9:00 am J. Collinge, University College London, United Kingdom:  
Prion strains, transmission barriers and neurotoxicity.

9:00-9:30 am Discussion

9:30-12:00 pm **SESSION 2: Strain Typing - Biochemical**  
Chair: J. Collinge, University College London, United Kingdom

9:30-9:50 am M. H. Groschup, Federal Research Institute for Animal Health, Greifswald-Insel Riems, Germany:  
Strain typing in animal TSE cases: criteria for strain definitions.

- 9:50-10:00 am Discussion
- 10:00-10:20 am J. Wadsworth, University College London, London, United Kingdom:  
Problems in defining human prion strains.
- 10:20-10:30 am Discussion
- 10:30-11:00 am Break
- 11:00-11:20 am A. Hill, University of Melbourne, Australia:  
Role of PrP post-translational modifications as markers of prion strain type.
- 11:20-11:30 am Discussion
- 11:30-11:50 am P. Gambetti, Case Western Reserve University, Cleveland, Ohio:  
Prion strains in human prion diseases.
- 11:50-12:00 pm Discussion
- 12:15 pm Luncheon at Robertson House
- 1:30-4:00 pm **SESSION 3: Strain Typing In Vivo and in Cell Culture**  
Chair: Byron Caughey, NIAID Rocky Mountain Laboratories, Hamilton,  
Montana
- 1:30-1:50 pm G. Telling, University of Kentucky, Lexington:  
Transgenic analysis of CWD strains.
- 1:50-2:00 pm Discussion
- 2:00-2:20 pm U. Agrimi, Istituto Superiore di Sanita, Rome, Italy:  
Strain typing of animal prions in natural hosts and by transmission to bank  
voles (*Myodes glareolus*).
- 2:20-2:30 pm Discussion
- 2:30-2:50 pm H. Laude, Institut National de la Recherche Agronomique, Jouy-en-Josas,  
France:  
Transmission of ruminant TSE to transgenic mice: further insight about prion  
strain diversity in naturally infected hosts.
- 2:50-3:00 pm Discussion
- 3:00-3:20 pm G. Zanusso, University of Verona, Italy:  
Intraspecies transmission of bovine amyloidotic spongiform encephalopathy.
- 3:20-3:30 pm Discussion
- 3:30-4:00 pm Break
- 4:00-6:00 pm **SESSION 4: Structural Basis of Strain-ness**  
Chair: B. Caughey, NIAID Rocky Mountain Laboratories, Hamilton, Montana

- 4:00-4:20 pm W. K. Surewicz, Case Western Reserve University, Cleveland, Ohio:  
Recombinant prion protein amyloid: molecular structure, strains and infectivity.
- 4:20-4:30 pm Discussion
- 4:30-4:50 pm I. Baskakov, University of Maryland Biotechnology Institute, Baltimore:  
Generating multiple strains of amyloid fibrils from a single polypeptide chain.
- 4:50-5:00 pm Discussion
- 5:00-5:20 pm D. S. Eisenberg, University of California, Los Angeles:  
Structural studies of amyloids, prions, and strains.
- 5:20-5:30 pm Discussion
- 5:30-5:50 pm R. B. Wickner, NIDDK, Bethesda, Maryland:  
Yeast prion amyloid structure explains heritability of 'strain' information.
- 5:50-6:00 pm Discussion
- 6:15 pm Reception at Robertson House
- 7:00 pm Dinner at Robertson House
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## Tuesday, 13 May

- 7:15-8:15 am Breakfast at Robertson House
- 8:30-9:00 am **SESSION 4: Structural Basis of Strain-ness (Cont'd.)**  
Chair: S. B. Prusiner, University of California, San Francisco
- 8:30-8:50 am B. Caughey, NIAID Rocky Mountain Laboratories, Hamilton, Montana:  
Ultrastructure and strain comparison of underglycosylated, anchorless scrapie  
prion protein fibrils.
- 8:50-9:00 am Discussion
- 9:00-12:00 pm **SESSION 5: Strain Biology *In Vivo***  
Chair: Stanley B. Prusiner, University of California, San Francisco
- 9:00-9:20 am B. W. Chesebro, NIH, Rocky Mountain Laboratories, Hamilton, Montana:  
Role of anchorless prion protein in pathogenesis induced by different scrapie  
strains.
- 9:20-9:30 am Discussion
- 9:30-9:50 am I. Vorberg, Technical University of Munich, Germany:  
Yeast prion aggregation propensities in mammalian cells.
- 9:50-10:00 am Discussion

- 10:00-10:20 am J. C. Bartz, Creighton University, Omaha, Nebraska:  
Prion strain interference.
- 10:20-10:30 am Discussion
- 10:30-11:00 am Break
- 11:00-11:20 am D. Westaway, University of Alberta, Edmonton, Canada:  
The PrP-like Shadoo protein: misfolding and *in vivo* variants.
- 11:20-11:30 am Discussion
- 11:30-11:50 am M. Jeffrey, Veterinary Laboratories Agency, Midlothian, United Kingdom:  
Species, strain and cell associated changes in abnormal PrP processing  
following prion infection.
- 11:50-12:00 pm Discussion
- 12:15 pm Luncheon at Robertson House
- 1:30-4:00 pm **SESSION 5: Strain Biology *In Vivo* (Cont'd.)**  
Chair: C. Soto, University of Texas Medical Branch, Galveston
- 1:30-1:50 pm J. Manson, Roslin Institute, Edinburgh, United Kingdom:  
Prion strains and host susceptibility.
- 1:50-2:00 pm Discussion
- 2:00-2:20 pm S. Priola, Rocky Mountain Laboratories, NIAID, NIH, Hamilton, Montana:  
Factors influencing *in vitro* and *in vivo* scrapie strain phenotypes.
- 2:20-2:30 pm Discussion
- 2:30-2:50 pm C. Weissmann, Scripps Florida, Jupiter:  
Do PrP-linked glycans contribute to prion strain determination?
- 2:50-3:00 pm Discussion
- 3:00-3:20 pm J. G. Safar, University of California, San Francisco:  
Conformational intermediates, clearance and species barrier of natural prion  
strains.
- 3:20-3:30 pm Discussion
- 3:30-4:00 pm Break
- 4:00-6:00 pm **SESSION 6: Strain Generation and propagation *In Vitro***  
Chair: C. Soto, University of Texas Medical Branch, Galveston
- 4:00-4:20 pm S.B. Prusiner, University of California, San Francisco:  
Protease sensitive and resistant strains of synthetic prions.
- 4:20-4:30 pm Discussion

- 4:30-4:50 pm C. Soto, University of Texas Medical Branch, Galveston:  
Generation of multiple new prion strains by *in vitro* PrP replication.
- 4:50-5:00 pm Discussion
- 5:00-5:20 pm J. Castilla, Scripps Florida, Jupiter:  
*In vitro* studies of the transmission barrier.
- 5:20-5:30 pm Discussion
- 5:30-5:50 pm S. W. Liebman, University of Illinois at Chicago:  
The birth of a foreign prion in yeast.
- 5:50-6:00 pm Discussion
- 6:15 pm Reception at Robertson House
- 7:15 pm Dinner at Robertson House
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### Wednesday, 14 May

- 7:15-8:15 am Breakfast at Robertson House
- 9:00-12:00 pm **SESSION 7: Varia**  
Chair: S.B. Prusiner, University of California, San Francisco
- 9:00-9:20 am R. A. Bessen, Montana State University, Bozeman:  
Using lymphoreticular system replication-deficient prion strains to determine  
routes of prion neuroinvasion.
- 9:20-9:30 am Discussion
- 9:30-9:50 am
- 9:50-10:00 am Discussion
- 10:00-12:00 pm **SESSION 8: General Discussion and Concluding Remarks**  
Chair: S. B. Prusiner, University of California, San Francisco
- 12:15 pm Luncheon at Robertson House
- Afternoon departure
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